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EDITOR'S NOTE

REVIEW ARTICLE

ARTICLES

CASE REPORTS

EDITOR'S NOTE :

FIVE YEARS OF JSP

The Journal of Surgery Pakistan (JSP) is in its fifth year of publication. The idea to start the Journal was conceived in 1996 by the present members of Editorial Board. Comments and blessings were requested from Editorial Consultants within the country and abroad. Their views were very encouraging and the Journal of Surgery Pakistan (International) was launched in 1996 under the patronage of Prof. F.U. Baqai and the efforts of Prof. Abdul Aziz, President and Secretary respectively of Pakistan Chapters of International College of Surgeons and SAARC Association of Surgeons.

Our deep gratitude is due to Prof. Muhammad Sultan Farooqui, President College of Physicians & Suregeons Pakistan, for providing us initial loan and the CPSP facilities to start this Journal. We are also thankful to the Pharmaceutical industry for providing us advertisements, which enabled us to print and distribute the Journal to the maximum number of surgeons within the country and abroad. The Journal of Surgery Pakistan is being sent to Bangladesh, India, Nepal, Sri Lanka, Maldives, Australia, Egypt, UAE, Jordan and U.K., as well as to the Royal Colleges of the United Kingdom and Ireland.The JSP was approved by the Pakistan Medical & Dental Council in October, 1997 and Index Medicus, WHO (EMRO) in April, 1999.

We will be failing in our duty if we do not appreciate and thank the doctors contributing original articles and case reports. Their efforts have enabled us to bring out the Journal regularly every three months. As a matter of fact, we are now receiving more articles than we can accommodate in any one issue. We want to assure our contributors that their articles are being reviewed by a minimum of three members of the Editoral Board and any corrections/changes made only where necessary.

We would like to particularly mention the following foreign Professors, whose contributions have encouraged us to continue with our efforts and maintain the high standard of JSP :

Australia:	Prof. T.C.K. Brown, 1998, 3,(1),1
Bangladesh:	Prof. Humayun Kabir Chowdhry, 1997, 2,(3),2-3
Egypt:	Prof. Reffat Kamel, 200, 5(3)2-8
Nepal:	Prof. A.K. Sharma, 1998, 4(1)2-4
United Kingdom:	Prof. John Hadfield, 1999, 4(3)37

EDITORS

SURGERY OF THE SPLEEN: SPLENIC CONSERVATION

REFFAT KAMEL* ; MANAR EL-TONSY**AND REFAAT R. KAMEL*

INTRODUCTION

The spleen has always been considered an organ of mystery. Although Hippocrates described the anatomy of the spleen remarkably accurately, the exact physiology of the spleen continued to baffle people for more than one thousand years after Hippocrates. Many primitive Mediterranean people believed that spleen was the cause of excessive mirth. It is also believed that ancient Greeks performed splenectomy to improve their prowess at the Olympic games. Even 600 years after Hippocrates, it was believed that spleen extracted melancholia from the blood and liver and purified it before excreting into the stomach. For this reason spleen is called the organ of laughter by Viaticus whereas the liver causes acrimony. Ancient Egyptians believed that spleen is the site of envy and malice.[1]

Spleen is a soft, fleshy, dark blue coloured organ situated in the upper left hypochondrium, 10 cm long and weighing about 200 gms. It forms a part of the lympho-reticuloendothelial system. In addition to its extensive lymphatic tissue, it has a complicated vascular system which, because of its tiny orifices and large macrophage population, acts as the optimal biological filter and constitutes a considerable hazard even for healthy red cells. Added to this, the spleen receives a disproportionate amount of the circulating blood volume for its relatively small size. Hence, it becomes involved secondarily in a wide range of haematological disorders.[2]

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FUNCTIONS OF THE SPLEEN:

The spleen, till the first half of the twentieth century, was considered to represent an organ of no special significance. Nowadays, its clearance function and position in the immunity system has been thoroughly investigated.[3] It has many important functions: haematological, immunological and phagocytic.

Haematological Functions of the Spleen:

Because of the peculiar anatomical arrangement of its blood vessels, the spleen is ideally suited as a site for fine control of the erythrocyte population. It removes fragmented, damaged or senescent red cells from the circulating blood, a process known as "Culling". It also plays a role in remodelling the surface of maturing erythrocytes and in preserving normal relationship between their membrane surface area and volume. Target cells, which have a relatively high ratio of membrane to intracellular content, appear in the peripheral blood soon after splenectomy.

A variety of intra-erythrocyte inclusions are removed by the spleen (through a process known as pitting), after which the red cells are returned to the circulation. Among the inclusions removed are Howell-Jolly bodies, which are probably nuclear remnants, siderotic granules, which are haemosiderin aggregates laid down during normal erythroid maturation and Heinz bodies, the pathological aggregates of denatured haemoglobin. Thus after splenectomy, Howell-Jolly bodies and siderotic granules may be seen in the peripheral blood and the red cells show striking changes in shape and size with

the appearance of acanthocytes, irregularly crenated cells and target forms.

The human spleen, unlike that of many animals, contains relatively little blood and hence has no important storage role. It appears to sequester a significant number of platelets; however after splenectomy there is nearly always a transient thrombocytosis.[2]

Immunological Functions of the Spleen:

The population of lymphocytes in spleen is in constant motion, a substantial proportion recirculating between lymph nodes and the spleen by way of the thoracic duct and blood stream. In the case of the spleen about half of the small lymphocytes recirculate fairly rapidly. Approximately a quarter of the body's total thymus-dependent (T) cells are in the spleen at any given time. The spleen helps in production of B-and T-lymphocytes through which many classes of immunoglobulins and cytokines are produced.

The main functions of spleen are summarized in Table I.

TABLE-I MAIN FUNCTIONS OF SPLEEN

IMMUNOLOGICAL:

1 Antibody production and cell mediated responses
2 Phagocytosis:
- Maturation of lymphoid cells
- Significant lymphopoiesis
- Source of suppressor T-cells
- Source of opsonin that promote neutrophil phagocytosis

HAEMATOLOGICAL:

- Filtration of particles from blood: non-specific or antibody coated
- Removal of red cell inclusions.
- Destruction of senescent or abnormal red cells.
- Compensatory haempoiesis, e.g. in myelofibrosis.
- Storage of platelets, iron and Factor VIII

Many studies have been carried out on the immunological and haematological changes in different clinical conditions associated with splenomegaly. Two groups of patients have been studied by Kamel R. et al[4] ; the first had schistosomal splenomegaly and the other group had undergone total splenectomy. In an attempt to assess splenic functions in these groups of patients, they found varying degrees of hypersplenism (anaemia, leukopenia and thrombocytopenia) in the first group. Abnormal red cells (acanthocytes, target cells, pitted cells and normoblasts) and inclusion bodies (Howell-Jolly bodies, argyrophilic inclusions and papenhiemer bodies) were detected in different values in the second group (Figs.1-5). Also they found modest reduction in the number of CD_4^+ cells (helper cells) in the first group while they were markedly decreased in the second group. The suppressor cells (CD_8^+) were elevated in the first group while they returned nearly to normal values in the second group.

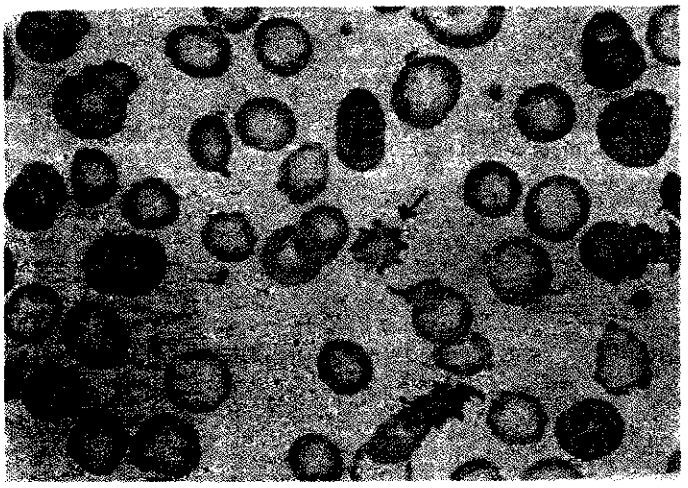

Figure 1. Acanthocytes in the blood film.

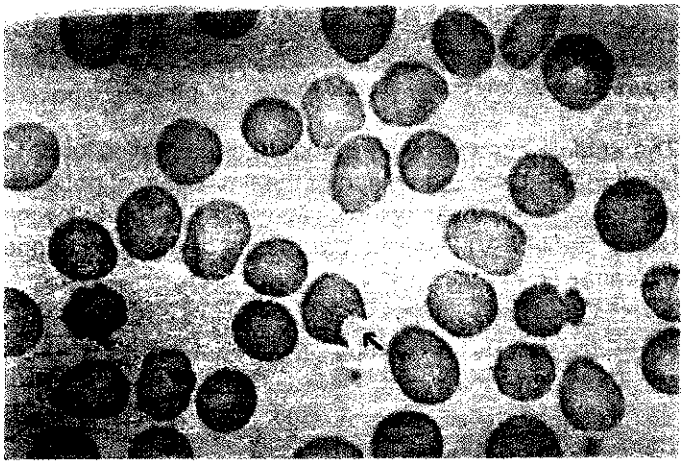

Figure 2. Pitted red blood cells.

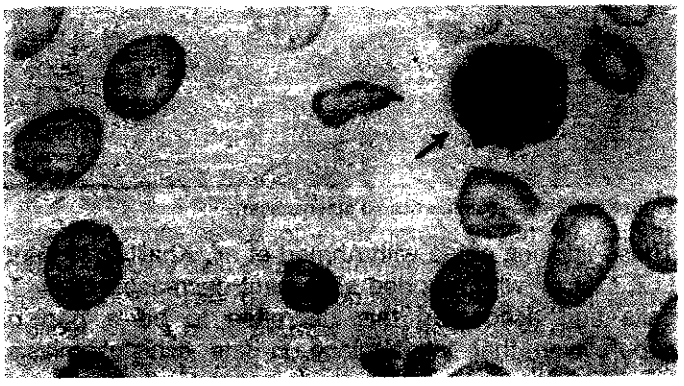

Figure 3. Normoblasts in the blood film.

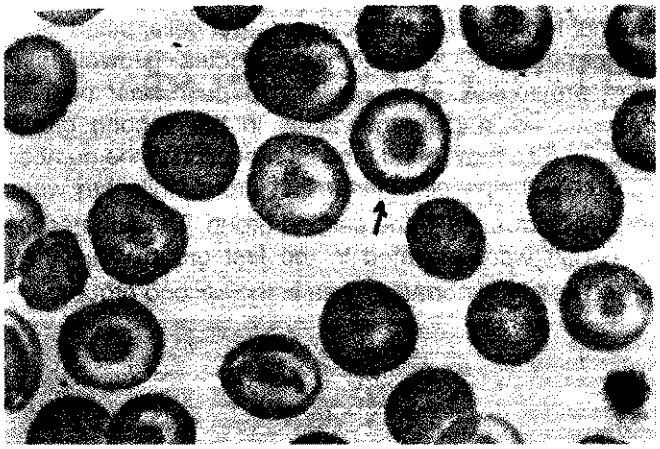

Figure 4. Target cells in the blood film.

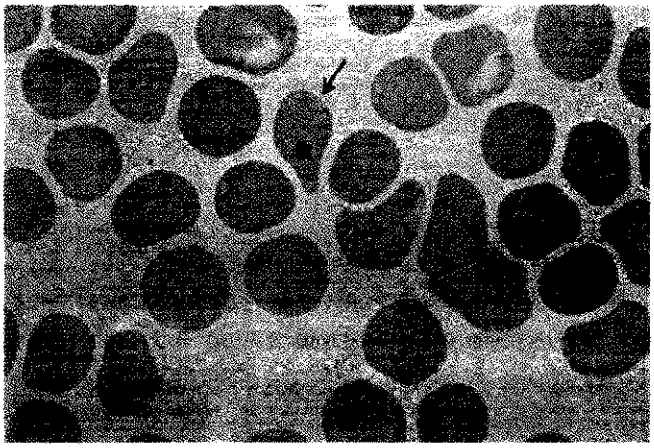

Figure 5. Howell Jolly bodies in blood film.

The splenic scan showed marked sequestration of ^{99m}Tc labelled R.B.Cs in the spleen of the first group of patients, with reversed hepatic/splenic ratio (Fig.6). The following tables (Table II-V) illustrate the results of this study done by Kamel R. et al :-

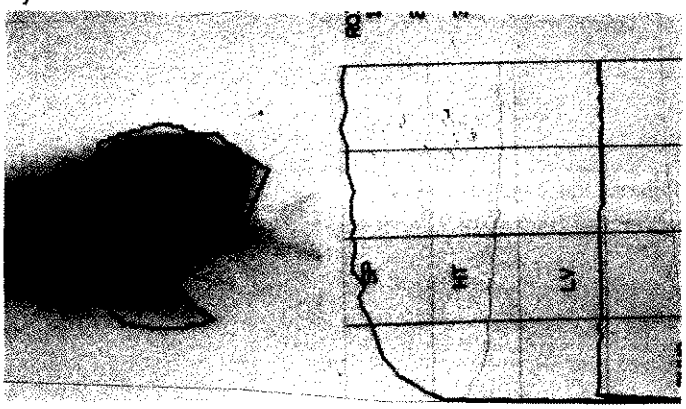

Figure 6. 99m Tc- sulphur colloid splenic scan.

These findings agree with Ahmed et al[5], Khalil et al[6] and Li,Z.J. et al[7] who reported significant decrease in CD_4 T-cells and accounted this decrease to be a part of suppression that takes place in late schistosomiasis. CD_4/CD_8 ratio was significantly decreased in the first group of patients, which is in agreement with Abdul Rahman et al.[8]

TABLE-II LABORATORY AND RADIOLOGICAL FINDINGS

Lab. & Rad. Data	Group II +ve	Group II %	Group III +ve	Group III %
B ova in stoo	I3	15%	0	0%
Rectal snip for B ova	8	40%	4	40%
IHAT for schistosomiasis	18	90%	5	50%
Hbs Ag.	4	20%	0	0%
HCV antibodies	12	60%	0	0%
Elevated transaminases	3	15%	1	10%
Abd. Ultrasonography:				
• Splenomegaly	20	100%	0	0%
• Enlarged liver	4	20%	4	40%
• Shrunken liver	6	30%	3	30%
• Normal sized liver	10	50%	3	30%

TABLE-III T-LYMPHOCYTES SUBPOPULATIONS IN ALL

T-lymphocytes	Group II Mean ± S.D.	P	Group III Mean± S.D.	P	Group I Mean ± S.D.
CD3+%	61.4±13.2	<.05	58.6±4.3	<0.01	78.4±6.5
CD4+%	43.3±8.6	<.05	36.4±4.8	<0.01	54.7±8.3
CD8+%	36.7±12.3	<.05	22.8±3.3	>0.05	25.3±4.7
CD4/CD8 ratio	1.2 ±0.5	<.05	1.6±0.2	<0.05	2.2±0.19

P>0.05=insignificant **P<0.01=significant**

TABLE-IV Haematological findings in all groups

Item	Group II Range	Group III Range	Group I Range
I-Complete bi. Picture			
Hb.conc.	8.1-11.3gm/dl	11.7-13.8gm/dl	12.5-14.6gm/dl
W.B.C.	1.7-7.2x10^3 /ul	4.8-12.3x10^3 /ul	7.0-11.2x10^3 /ul
R.B.C.	3.00-4.1x10^6 /ul	4.2-4.8x10^6 /ul	4.8-5.3x10^6 /ul
Platelets	52-291x10^3 /ul	365-458x10^3 /ul	250-290x10^3 /ul
Eosinophil	0-6%	0-2%	0-1%
II-Abnormal R.B.Cs.			
Normoblasts	0	0-8/100W.B.C.	0
Acanthocytes	80-142/500R.B.C.	52-150/500R.B.C.	0
Target cells	0	200-300/500 R.B.C.	0
Pitted cells	0	2-12/500 R.B.C.	0
III-Inclusion bodies			
Howell-Jolly bodies	0	7-15/500 R.B.C.	0
Argyrophil inclusion	0	0-6/500 R.B.C.	0
Pappenhiemer bodies	0	0-2.500 R.B.C.	0

TABLE-V ^{99m}Tc Sulphur colloid splenic scan of patients in group II

Time	Splenic/hepatic Mean ± S.D.	Splenic/ cardiac Mean ± S.D.
05 minutes	2.7±2.1	0.72±0.3
10 minutes	3.2±2.4	1.01±0.3
15 minutes	3.7±2.7	1.05±0.4
20 minutes	4.9±2.2	1.2±0.2

Group I : 10 healthy control subjects.
Group II : 20 patients with hepatosplenic schistosomiasis.
Group III: 10 bilharzial patients who underwent total Splenectomy.

TABLE-VI Causes of enlargement of the spleen:

Acute bacterial, viral and other infections
Subacute bacterial endocarditis
Chronic bacterial infection; T.B. and brucellosis
Chronic parastic infections; Malaria, Kala-azar, Schistosomiasis,
Toxoplasmosis and Babesisosis.
Idiopathic non-tropical splenomegaly

'Congestive'; portal hypertension
- Hereditary spherocytosis
- Symptomatic elliptocytosis
- Structural haemoglobinopathy, e.g. HbC disease
- Thalassaemia
- Red cell enzyme defects

Acquired haemolytic anaemia
- Warm antibody haemolytic anaemia
- Cryopathic haemolytic syndrome

Primary blood dyscrasia
- Acute leukemia
- Chronic myeloid leukemia
- Chronic lymphatic leukemia
- Polycythemia vera
- Myelosclerosis

Trauma
- Perisplenic haematoma

Reticulosis
- Hodgkin' disease
- Non-Hodgkin's lymphoma
- True histiocytic tumours

Miscellaneous
- Amyloid, sarcoidosis, tumours of the spleen

zConnective tissue disorders
SLE -Felty's syndrome

However, Khalil et al[6] reported that there was an increase in the number of pan T-cells and reduction in the number of T-suppressor cells, this indicates that T-helper/T-suppressor ratio improved after splenectomy with a concomitant improvement of the immunological response. Kamel R. et al[4] reported that CD_4/CD_8 ratio was also decreased in splenectomized patients inspite of returning the number of suppressor cells nearly to the normal values, which in agreement with another study, which evaluated segmental splenectomy versus total splenectomy.[9] Also M. Petroianu et al investigated the immunological profiles of patients with schistosomiasis, they studied the B- and T-lymphocyte counts in patients who underwent distal splenorenal shunt, preserving the spleen, showed significantly increased T-lymphocytes compared with healthy controls.[10]

Ma M.Y. et al studied the peripheral blood natural killer cell activity, phagocytosis of neutrophil and lymphoctye blastogenesis.[11] They suggested that splenectomy does not only reduce the portal hypertension but also improves the regulatory functions of cellular immunity which may enhance the patient's resistance against pathogens.

CAUSES OF SPLENOMEGALY

The spleen may enlarge transiently in a variety of acute infections, such as brucellosis and tuberculosis and in subacute bacterial endocarditis. Massive splenomegaly may result from several tropical disorders including malaria, leishmaniasis and schistosomiasis. Moderate enlargement may accompany any cause of portal hypertension. Table-VI summarizes the causes of splenomegaly:

The Clinical Assessment of Splenomegaly

The clinical significance of splenomegaly can be assessed by answering three questions: is a mass in the left upper quadrant really an enlarged spleen, if so, why it is enlarged and will splenectomy benefits the patient?

A mass in the left upper quadrant, which moves downwards and slightly medially on respiration, is usually an enlarged spleen. The most common difficulty experienced with masses in the left upper quadrant is mistaking for an enlarged left kidney or a neoplasm of the splenic flexure of the colon or stomach. Renal enlargement is usually fairly obvious because the mass feels like a kidney, tends to move directly downwards on respiration rather than downwards and medially and often has a band of resonance in front of it because of its relationship to the descending colon.

Investigations of the cause of Splenomegaly:

A palpable spleen is enlarged two to three times normal. A normal-sized spleen is never palpable. Splenomegaly should always be carefully investigated. With the aid of a good history and clinical examination, together with a few simple laboratory and radiological investigations, it is possible to determine the cause of splenomegaly in majority of the patients. In taking a history from patients with splenomegaly it is important to find out whether they have any symptoms suggestive of infection or malignant disease and where they have travelled over the preceding years. Clinical examination must include a very careful study of the superficial lymph nodes and the tonsillar and adenoid tissues, a search for stigmas of liver disease, a full examination of the skin for evidence or bruising, and a complete survey of the all systems.

A few simple laboratory investigations will provide the cause for most cases of splenomegaly. Almost all the splenomegalies associated with haematological disorders can be diagnosed by a complete blood examination together with a bone marrow aspiration. Liver disease is often associated with abnormal liver function tests or oesophageal varices on a barium swallow examination. Liver biopsy may be necessary to determine the type of

liver pathology. The diagnosis of lymphoma or other RE malignancies may be accomplished by biopsy of a suitable node, the liver or another involved organ, lymphangiography or bone marrow biopsy; in some cases it may be to go to a diagnostic splenectomy.

The various infections, which produce splenomegaly may be identified from appropriate haematological or serological studies. Infectious mononucleosis is diagnosed by the finding of atypical lymphocytes in the blood, a positive Paul-Bunnel test (or related screening procedures) and a rising anti-Epstein-Barr virus titre. Among those who have travelled to the tropics, malarial parasites should be looked for and the marrow examined for Leishman-Donovan bodies in case of Leishmaniasis (Kala-azar).

Assessment of Hypersplenism:

In assessing whether the removal of an enlarged spleen will benefit the patient there are two main factors to be considered; the cause of splenomegaly and whether there is a significant degree of hypersplenism. There are some conditions in which splenectomy is almost always associated with an improvement of the patient's clinical condition. However, there are many disorders including haemolytic anaemis, the myeloproliferative states, Felty's syndrome and some malignancies in which there may be a significant degree of splenomegaly and hypersplenism, but it is not clear to what degree the spleen is contributing to the clinical disability. How does one proceed to determine whether splenectomy is indicated?

Hypersplenism is usually associated with anaemia, neutropenia or thrombocytopenia. The neutropenia may be so severe to cause recurrent infections and the thrombocytopenia may be associated with purpura. It is vitaly important to obtain a good bone marrow sample as a part of the routine work-up of patients with suspected hypersplenism.

In the further assessment of splenic function three questions can be asked: what is the state of bone marrow function?, is there pooling or destruction of formed elements of the blood in the spleen? and is the spleen contributing significantly to red cell production? In assessing splenic pooling it is useful to compare the circulating red cell mass with the peripheral blood haematocrit; if there is a gross discrepancy and the haematocrit is much lower than might be expected from the circulating red cell mass, this suggests that part of the latter is being held in the enlarged spleen. A plasma volume estimation is also very useful because it is often markedly elevated in patients with hypersplenism. Finally, red cell survival is measured by ^{51}Cr labelling and the major sites of destruction assessed by external scanning.

The spleen can be visualized by scintillation counting following injection of labelled red cells after they have been heated at 49.5° C for 20 minutes, which causes them to be sequestered preferentially in the organ. If after heating they are labelled with ^{51}Cr or ^{99m}Tc, it is possible to obtain an accurate estimation of splenic size. In assessing the major sites of red cell destruction, the cells are labelled with ^{51}Cr and using a carefully calibrated external scanning system. A normal spleen contains less than 5% of the circulating red cell mass while in some disorders associated with splenomegaly and pooling as much as 40-60% of the circulating cell mass. Further investigations for the cause of splenomegaly include ultrasonography, C.T. Scan and magnetic resonance imaging techniques (MRI).

Indications for Splenectomy

Hypersplenism is the main indication for splenectomy. Removal of the spleen may be undertaken for those rare cases in which the spleen is suspected before operation, or found at operation, to suffer from an abscess confined to the organ from tuberculosis, from a cyst or from a splenic tumour. The operation is indicated in haemolytic jaundice, being in that condition more effective in the congenital than in the acquired variety.

Splenectomy is required in certain form of portal hypertension, either alone or as a preliminary to spleno-renal venous anastomosis. Rupture of the spleen of course requires splenectomy. It can be also done in Gaucher's disease and in purpura or as a part of another operation (Figs. 7,8,9).

Complications of splenectomy

Patients with hypersplenism are often at poor operative risks because of anaemia, thrombocytopenia, prolonged steroid therapy or advanced malignant disease.

- Overwhelming post-splenectomy infection (O.P.S.I.); it is the most important and serious complication and this directed the attention to the value of spleen and hence it leads to splenic conservation and segmental splenectomy
- Haemorrhage: [post-operative haemorrhage]
- Pulmonary atelectasis.
- Subphrenic abscess.
- Post-splenectomy fever:-
  - Chest or wound infection or the presence of a subphrenic abscess
  - Leucocyte agglutination antibodies, normally removed by the spleen
- Thrombocytosis.
- Pancreatic fistula.
- Gastric fistula.

Comparative Studies on Segmental Splenectomy Versus Total Splenectomy

Because of many complications that may occur after total splenectomy, the Surgeons modify the procedure of total splenectomy to another one with preserving some splenic tissue. Kamel R. et al, through their study on the splenic

Figure 7. Arterial supply of the spleen. Arrows indicate avascular planes coinciding with surface notches.(S, Splenic; ST, superior terminal; MT, middle terminal; IT, inferior terminal; LP, lower polar; SG, segmental.

Figure 8. Haemostatic compression of the spleen using manual pressure by the thumbs and forefingers of the second assistant. The cut on the diaphragmatic surface of the spleen is shown.

Figure 9. The splenic remnant with an omental patch in place.

functions in bilharzial patients, concluded that although total splenectomy improved the haematological pattern and the cytopenias, which are prominent features in hepatosplenic schistosomiasis, yet the immunological profile was still altered.[4] So, it is recommended to perform segmental splenectomy with retention of a normal mass of functioning residual spleen to preserve more immunological function and to protest against life-threatening occurrence of post-splenectomy sepsis.

Kamel R & Dunn MA studied segmental splenectomy in patients with hepatosplenic schistosomiasis.[12] The aim was to preserve a functional remnant comprising 20-30 per cent of the bulk of a greatly enlarged spleen. The operative technique involved devascularization of anatomic segments and suture of an omental patch to the residual spleen. They concluded that this procedure was simple, well tolerated and effective in relieving abdominal discomfort and cytopenias. The residual spleens showed normal uptake of ^{99m}Tc-sulphur colloid. They reported that if long term observation confirms the value of segmental

TABLE-VII **Haematological effects of segmental and total splenectomy**

Procedure	N	R.B.C. (106/mm3)	W.B.C. (10/mm3)	Platelets (103/mm3)
Segmental				
Pre-operative	51	3.2±0.3	5.5±1.6	140±50
Postoperative	45	3.9±0.4	4.8±1.3	363±44
Total				
Pre-operative	44	3.2±0.4	5.2±1.4	141±47
Postoperative	35	4.0±0.3	6.1±1.8	422±64

TABLE-VIII **Antibody titres 6 months after segmental and total splenectomy**

Vaccine	Procedure	1/Median titre	Range
Tetanus	Segmental	320	40-640
	Total	160	20-640
Pneumococcus	Segmental	320	80-640
	Total	320	40-1280
Streptokinase/	Segmental	640	80-1280
Streptodornase	Total	320	160-640
Measles	Segmental	80	20-160
	Total	80	40-160

splenectomy in hepatosplenic schistosomiasis, it might become appropriate therapy for those patients with symptomatic splenomegaly who do not required portal decompression.

Kamel R. et al[9] evaluated segmental splenectomy in 51 patients who required splenectomy to relieve the symptoms of schistosomal splenomegaly, and compared their clinical and immunological results with that of 44 patients who underwent total splenectomy in an

unrandamized study. They reported that patients having segmental splenectomy had a similar post-operative course to those having total splenectomy. Conversion of a segmental to a total splenectomy was required in two cases due to technical faults.

TABLE-IX **Peripheral blood T-helper and suppressor cells after segmental total splenectomy**

Cells	Pre-operative	Postoperative	Normal value
Total T cells %			
Segmental	65.9±14.4	80.4±6.6*	52.6±9.4
Total	50.7±4.2	54.9±3.8	
T-helper: T-suppressor Cell ratio			
Segmental	1.4±0.4	2.1±0.4+	1.6±0.6
Total	1.3±0.3	1.2±0.3	

Data are given as means ± S.D.
**Greater than pre-operative value with borderline statistical significance, 0.10>P>0.05 by "t" test.*
+Significantly greater than pre-operative, P<0.02 by "t" test.

No growth of the spleen has occurred in up to 4 years of observation. They noted an increased percentage of T lymphocytes with an increased ratio of T helper to T suppressor cells in patients having segmental splenectomy. The Tables VII-IX illustrate the results of the study done by Kamel R. et al[9] :-

Reciprocal titres were done in seven patients with segmental and five patients with total splenectomy vaccinated from 6 to 21 days before operation. There were no significant differences in antibody titres between patients with segmental and total splenectomics assessed by pooled estimates of variance.

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CARCINOMA STOMACH A THERAPEUTIC CHALLENGE

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ABSTRACT:
A five year retrospective study was carried out in Surgical Unit - IV Liaquat Medical College Hospital Jamshoro. The cases of carcinoma stomach admitted from January 1993 to December, 1997 were included in the study to determine the clinical presentation and treatment of gastric cancer. Duration of symptoms were less than six months in 70% cases. Abdominal pain (60%), vomiting (54%), peptic ulcer symptoms (50%), GIT bleeding (36), dysphagia (36%) weight loss (40%) palpable mass (48%), ascites (34.3%) and hepatomegaly (42.8%) were the main clinical presentations.

Out of 50 cases 35 were operated, 15 being unfit for the operation. Out of 35 cases no procedure could be performed in 15 cases due to advanced stage of disease. Palliative gastrectomy was performed in 10 and curative surgery in another 10 cases. Duodenal leakage (14.3%), bleeding (11.43%), wound infection (8.5%), UTI (17.14%) and small bowel obstruction (2.83%) were major complictions. Upper GIT endoscopy for early diagnosis of carcinoma of stomach is essential especially before starting treatment for peptic ulcer symptoms.

KEY WORDS: Carcinoma stomach, gastrectomy, Peptic ulcer, endoscopy.

INTRODUCTION

Carcinoma stomach is the second most common cause of death from malignant disease worldwide.[1] The highest incidence is in Japan. The prognosis of the patients with carcinoma stomach is poor due to late diagnosis and advanced disease at the time of diagnosis. Although operative mortality is high, surgery is the only option for cure. The aim of the study was to determine the clinical presentation and surgical treatment of carcinoma stomach at Liaquat Medical College Hospital, Jamshoro.

PATIENTS AND METHODS

From January 1993 to December 1997 (5 years) all the cases of carcinoma stomach admitted at Liaquat Medical College Hospital Jamshoro were studied. The cases were collected from hospital record, operation theatre and Radioisotope Centre Jamshoro.The clinical presentation, treatment and complications were recorded. Only histologically proven cases of carcinoma stomach were included. Barium meal and gastroscopy with biopsy was carried out in all cases. X-ray chest and ultrasound liver were done to detect secondaries. CT and MRI were not done because of the high cost.

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RESULTS

A total of 50 cases were managed in five years, out of which 35 were operated and 15 were inoperable. Majority of the patients were between 41-60 years of age, median age being 51 years. There were 10 female and 40 male patients.The duration of symptoms was less than six months in 35 patients (70%). The symptoms in decreasing order of frequency were abdominal pain (60%), vomiting (54%), peptic ulcer symptoms (50%), GIT bleeding (hematemesis / melena) (36%), dysphagia (36%), weight loss (40%). On examination mass was palpable in 48%, ascites 48%, hepatomegaly 40% and supraclavicular lymph nodes in 20% cases.

Out of the 35 operated cases, 15 (42.8%) were closed without any procedure due to advanced disease, palliative surgery was done in 10 patients (28.5%) and curative surgery in 10 patients. In palliative surgery group, gastrojejunostomy was done in 5 and palliative partial gastrectomy (Billroth II) in 5 cases. In curative surgery case, Billroth II was performed in 5 cases, Billroth I in one case and total gastrectomy in 4 cases.

Total gastrectomy was performed in tumours located in the proximal part of stomach or whole of the stomach and partial gasterectomy (Billroth I/II) were performed for tumours in the middle or distal part of the stomach. The perigastric lymph nodes along lesser and greater curvatures were removed and lesser and greater omentum resected. Location of the tumour was pyloric in 28 (56%), body 12 (24%), cardiac end 6 (12%) and whole of the stomach 4 (8%) cases.

Operative complications were duodenal leakage 5 (4 in Billroth II and one in total gastrectomy). Bleeding from G.I.T. occurred in 2 cases (one in Billroth II and one in total gastrectomy. Re-exploration was done in 2 cases in Billroth II because of leakage. Other complications were wound infection in 3, pneumonia in 8, urinary tract infection in 6 and small bowel obstruction in one case. Four patients died; three in Billroth II due to leakage and multiple organ failure and one in gastrectomy due to myocardial infarction. (Table-1):

TABLE-I POST OPERATIVE COMPLICATIONS

Complication	Total Series (n = 35)	Billroth-II (n = 10)	Gastrectomy (n = 4)
LEAKAGE (DUODENAL)	5 (14.3%)	4 (40%)	1 (25%)
BLEEDING (DUODENAL)	2 (5.7%)	1 (10%)	1 (25%)
BLEEDING GJ/OESOPHAGEAL	2 (5.7%)	1 (10%)	1 (25%)
RELAPAROTOMY	2 (5.7%)	2(20%)	0
WOUND INFECTION	3 (8.57%)	2 (20%)	1 (25%)
PNEUMONIA	8 (19.97%)	6 (60%)	2 (50%)
UTI	6 (17.14%)	3 (30%)	3 (8.57%)
SMALL BOWEL OBSTRUCTION	1 (2.83%)	1 (10%)	0
DEATH	4 (11.43%)	3 (30%)	1 (25%)

DISCUSSION

The incidence of carcinoma stomach has decreased worldwide, but it still remains a serious problem. It is not a common disease in Pakistan.[1,2,3] We have recorded 50 cases in 5 years.It is the disease of elderly people[4,5,6,7] but median age in our series was 51 years. It has also been reported in children. Our series showed high incidence in males, which is also supported by world literature.[6,7]

The symptoms of patients were less than six months duration in our series and other authors have also shown short duration.[9] Clinical presentation in our series was similar to that described in other series.[4,9,10,11] Symptoms are usually vague and treated like benign peptic ulcer till the tumour blocks the passage of food causing dysphagia or bleeding from the tumour. The delay in the diagnosis is due to symptomatic treatment with antacids or H_2 receptor blocker.

Our cases were diagnosed on barium meal and gastroscopy. We have not used laparoscopy with ultrasonograpy,[12] endoscopic ultrasonography, CT[13] and MRI for preoperative staging as suggested in literature. Laparotomy was done in all operable cases for staging and possibly palliative or curative resection. Some authors have suggested that unresectable carcinoma on scanning had potentially resectable lesion on laparotomy.[14]

Majority of our patients (56%) had carcinoma of the pyloric end of stomach, which is also shown in world literature.[6,15,16] But incidence of carcinoma of the upper end of stomach is gradually increasing, which has poor prognosis.[10,17,18] Our operative findings of involved lymph nodes, greater omentum, pancreas, transverse colon, ascites and liver metastasis is similar to other studies.[10]

Our cases were in advanced stage IV (40%) at the time of diagnosis, similar to other series.[10,15,19,20] We did not have a single case of early carcinoma stomach, while in Japan more than 50% of carcinoma stomach are detected early and in Europe the rate is 10-20%.[21] This is due to mass screening by radiography in Japan. In treatment of early carcinoma stomach 5 year survival is more than 90%.[22] In Pakistan we do not have screening programme for carcinoma stomach. We can detect cases by early referral and decrease the incidence by primary preventive measures, perhaps diet, antioxidants and eradication of Helicobacter pylori, as suggested by some authors.[1]

We have done total gastrectomy for tumour involving the proximal or whole of stomach and subtotal gastrectomy for distal part of stomach. This is also supported in other series.[4,10] Some surgeons advise total gastrectomy even for distal tumour but survival after total gastrectomy does not exceed that after partial gastrectomy.[23] We have done Billroth II gastrectomy for distal tumour and only in one case Billroth-I, because of fear of local recurrence due to compromise on resection margins. British stomach cancer group[24] have emphasised that microscopic disease of the resection line influenced long term survival and resection line must be clear; at least 5 cm margin clearance is required. If resection margins are not clear, then prognosis of stage-II falls to that of stage-IV.[25]

Billroth I has advantage over Billroth II that normal duodenal passage of food is preserved and the complications related to gastrojejunostomy are avoided. Recurrence is very common after Billroth I and some surgeons are reluctant to perform Billroth I for carcinoma stomach.[26]

We have not done any radical resection which is associated with high complication rate without any demonstrable survival advantage. Extended lymph node resection has been done by Japanese and some European surgeons, which is claimed to decrease the local recurrence.[27,28,29] But some surgeons have shown increased mortality in such resection.[30] In Japan the

radical operation has been done with mortality of only 0.4%. Less radical resection leads to resection line involvement and decreases the survival.[31] Palliative procedures have also been described, in literature and good control of symptom is reported in 50% of cases.[4,32,33]

Our operability rate of 70% and resectability is 30% and curative rate 20%. While Japanese[34] reported operability rate of 99.8%, resectability 93.1% and curative 58.6%. Other series report operability 74%, resectability 35.2% and curability 39.4%. Five year survival in Japanese study is 60.5% and in others 39.4%. We could not determine the 5 years survival because of lack of follow up.[34]

The operative mortality in our series (death within 30 days) is 11.43% while the literature shows 6-14%.[19,35,36] The frequency of post-operative complication was high in our series. There was duodenal leakage in five patients (14.3%), out of which three died. The duodenal leakage due to suture breakdown is described as the major cause of morbidity and mortality (1.2-7.2%)s in literature.[37,38,39] The other complications bleeding, pneumonia, wound infection and UTI have also been reported in literature.[39,40] Reoperation rate in our series is 5.7%, due to leakage while as reported in literature is 5% due to anastomotic leakage, abscess and dehiscence of wound.[26]

Recent dyspepsia in older patients should be investigated by endoscopy before treatment is started. Early gastroscopy should be done in patients with normal barium meal, who have persistent GIT symptoms after therapy.

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CORRIGENDUM

In the article "Olecranon Fractures Treated by Weber Vasey Tequnique" published in Journal of Surgery Pakistan (Vol. 5 No. 2 (April-June 2000).

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PREGNANCY WITH MITRAL VALVE DISEASE: AN EXPERIENCE WITH 44 CASES

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ABSTRACT:
Forty-four cases of pregnancy with mitral valve disease (MVD), admitted in the Department of Obstetrics and Gynecology, Jinnah Postgraduate Medical Center, Karachi from July, 1989 to December 1990, were studied and analysed for etiology and impact of the disease on pregnancy and vice versa. Management during pregnancy/labour and outcome of pregnancy were also studied. During this period 10,798 patients delivered including 44 cases of MVD (1:204). Forty-three cases had rheumatic valvular disease and one had mitral valve prolapse. Fourteen patients were in functional grade – I during pregnancy, 15 in grade – II, 10 in grade – III and 5 in grade – IV. Twenty cases were on medical treatment, while 24(54.5%) had undergone different type of surgeries on mitral valves. Out of 44 patients one had therapeutic abortion, one had an intrauterine death, 22 patients had outlet forceps delivery, 17 had spontaneous vaginal delivery, one had assisted breech, one twin delivery and one lower segment cesarean section. Five patients (11.3%) developed pulmonary oedema during labour. Mean weight of the babies was 2.6 kg. Perinatal death rate was 6.8%. No ill effects of drugs used during pregnancy were seen on babies.

KEY WORDS: Pregnancy, Mitral valve disease.

INTRODUCTION

Patients with heart disease are five times at risk than normal pregnant women and mitral valve disease is the most common (90%) of all cardiac diseases during pregnancy.[1] It reduces the response of patients to haemodynamic changes during pregnancy, resulting in increased risk of abortion, premature labour or fetal death and light for dates babies.[2] On the other hand, pregnancy imposes an augmented demand upon the heart by increasing blood volume, heart rate and cardiac output; 25% of the patients with mitral stenosis have cardiac failure first time during pregnancy.[3]

Mitral valve disease of child bearing age is almost always rheumatic in origin with minor contribution of mitral valve prolapse.[1] Prevalence of rheumatic heart disease in Pakistan is 7-11/1000 in rural and urban population.[4] In South East India it occurs in 44/1000 population,[5] while in Dhaka it comprises 35% of all cardiac cases,[6]

Proper understanding of haemodynamic changes during pregnancy and labour and their alteration by the disease help in early detection of the case so that it can be managed in such a way that extra demands of pregnancy and stress of labour are satisfactorily met and the patient would emerge from the experience of child bearing and child rearing without degradation of her condition.

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PATIENTS AND METHODS

An analysis of 44 cases of pregnancy with mitral valve disease is presented. These cases were admitted and managed in the Department of Obstetrics and Gynaecology, JPMC, Karachi over a period of one and half year from July 1989 to December, 1990.

After admission, detailed history of patients were recorded regarding their age , parity, booking status, socio-economic conditions, previous history of rheumatic fever and its medication, duration of mitral valve disease, is it medically treated or the patient has undergone cardiac surgical procedure. Her cardiac conditions in previous pregnancies were also noted. Later, a thorough physical examination with particular attention to cardiovascular system and obstetrical examination were performed.

Severity of the heart disease was graded from I-IV according to the classification of New York Heart

Association (NYHA, 1965).[7] This classification was taken as guide for management and was not wholly relied upon, as it is based on symptoms only.

- Grade-I: Patients with cardiac disease but no limitation on physical activity.
- Grade-II: Slight limitation of physical activity , comfortable at rest.
- Grade-III. Marked limitation of physical activity. Minimal physical exertion gives rise to discomfort.
- Grade-IV: Patients are unable to carry out any physical activity without discomfort and have orthoponea.

The patients were observed throughout their stay in hospital during pregnancy, labour and puerperium. Any deterioration in their cardiac status was observed or complications of pregnancy were noted. The mode of delivery, duration of labour and outcome of pregnancy were noted. Perinatal deaths were also observed . The patients were followed during puerperium and condition was noted at the time of discharge from the hospital which on average was 7-10 days post-natally.

RESULTS

Over a period of one and a half year from July 1989 to December 1990, the total number of obstetrical admissions were 12935 and total number of deliveries were 10798. Out of these 53 had heart disease . Among 53 cardiac patients 44 (83%) had mitral valve disease, 43 of whom had rheumatic valvular heart disease and one had mitral valve prolapse. The ratio of rheumatic to congenital heart disease was 43:1.

Out of 43 cases of post rheumatic mitral valve involvement 30 had mitral stenosis, 7 mitral regurgitation and 6 both mitral stenosis and regurgitation.Twenty cases were on medical treatment while the other 24 were treated surgically with 10 (22.8%) mitral valve replacements. Among them 2 had bivalve replacement, 7 (15.9%) had mitral valvotomy and 7 had mitral commisurotomy. Twenty-six (59%) were booked cases with regular antenatal and cardiologist checkups. The remaining 8 (41%) were non-booked, they were either referred from other clinics or had come in labour.Ages ranged from 18-32 years; 8 (18.8%) were below 20 years, 14 (31.8%) were between 20-24 years, 13 (29.5%) were between 25-30 years and 9 (20.45%) were above 30 years of age.

Out of 44 patients with mitral valve disease, 12 (27.3%) were primigravidas and 6 (13.6%) were grandmultiparas. Fourteen patients were in functional grade-I during pregnancy, 15 were in grade-II, 10 in grade-III and 5 in grade-IV. Out of these 5 in Grade-IV, three were booked cases and remained hospitalized for most of their antenatal period. Two cases came in labour. All were non-operated cases. One had therapeutic abortion at 12 weeks, who was in grade-IV and failed to respond to treatment. One had an induced abortion due to intrauterine death at 20 weeks of gestation, 22 had outlet forceps delivery, 17 had spontaneous vaginal delivery and one patient had assisted breech, one twin delivery and one had lower segment caesarean section for fetal distress.

Five patients (11.3%) developed pulmonary oedema during labour, among them 3 were in grade-III, one in grade-I and one in grade-IV.

The weight of the babies ranged between 2.2 – 3.4 k.g. with a mean of 2.6 k.g. Three babies had neonatal deaths and two developed jaundice on the 3rd postnatal day. Perinatal death rate was 6.8%. No ill effects of drugs used in pregnancy were seen on babies. Breast feeding was practised in 35 (79.5%) cases.

DISCUSSION

In this study the frequency of heart disease during pregnancy was 0.4% of all obstetrical admissions or 1:204 deliveries which is comparable to generally stated incidence of 0.3-3.5% .[4]

Mitral valve disease is the most common of all cardiac diseases during pregnancy, comprising approximately 90% of cases. It is almost always rheumatic in origin. The incidence of rheumatic heart is declining in developed world[1] but is still prevalent in developing countries due to poor socioeconomic conditions and densely residing population.[6] In this study out of 53 cardiac cases 83% (44 cases) had mitral valve disease. Majority of them (43) were rheumatic in origin.

In this study 59% patients were booked cases and minimum complications were seen in them, even in patients in grade – IV who remained hospitalized for most of the duration of their pregnancy. The general experience is that women with prosthetic heart valve tolerate very well the haemodynamic challenge posed by pregnancy and labour.[8] Ten cases had mitral valve replacement with minimum complications. Patients who developed cardiac failure or pulmonary oedema were non-booked and non-operated cases.

Chesley showed that there is no difference in the behaviour of the patients during pregnancy and labour with different parity.[9] Moreover, there is no difference in the ages of patients regarding onset of failure and death. However in this study, similar to other patients of mitral valve disease, majority belonged to young age and were of low parity. Termination of pregnancy is rarely to be considered in patients with mitral valve disease.[2] In the present study only one patient had undergone therapeutic abortion followed by sterilization due to uncontrolled cardiac failure right from conception.

The management of labour requires special care in patients with mitral valve disease. The duration of labour should be shortened. Relief of pain during labour is very important and particular care should be taken to avoid excessive effort on the part of the patients. The most dangerous time for developing congestive cardiac failure and pulmonary oedema is immediately after delivery.[7] In the present study, 5 developed pulmonary oedema during labour and 2 had congestive cardiac failure during early puerperium; among them one had cesarean section for fetal distress. There were only 3 perinatal death (6.8%) and no ill-effects of maternal medication was noticed in these babies.

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INTRACAVERNOSAL INJECTION PHARMACOTHERAPY IN MALE IMPOTENCE

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ABSTRACT:
Impotence is not an uncommon problem and is very distressing to the patients. Intra-penile injection of Aplrostadil is a novel approach with great diagnostic and therapeutic value. The aim of this prospective study was to confirm that intra-cavernosal pharmacotherapy is acceptable to our local community and that it is safe and effective. Patients with clinically suspected psychogenic impotence were offered a single intracavernosal injection of Alprostadil, 10 micrograms. The response was recorded. Two follow-up visits were arranged and a detailed questionnaire completed for every patient. Seventy-nine patients were clinically diagnosed to be suffering from psychogenic impotence. Five patients were lost to follow-up and were excluded. Four patients refused Intracavernosal injections. Of the patients receiving the drug, 60(86%) were satisfied with the results. Major complications included pain in 6(8.6%) that responded to conservative measures, a haematoma and an inadvertent urethral injection. Intracavernosal injection is a safe and effective tool in diagnosing and treating psyhogenic impotence. Time-consuming and expensive investigations are not required in majority of the cases clinically suspected of having a psychogenic origin. It is an acceptable treatment option. The complications are surgical in nature and we recommend that the injection be made by surgeons.

KEY WORDS: Impotence, Pharmacotherapy, intracavernosal injection

INTRODUCTION

Impotence is defined as the inability to initiate and sustain a penile erection for penetration and orgasm. Impotence may be vasculogenic, neugrogenic or psychogenic. The authorities seem to agree that in the vast majority of cases, some psychological element is present.[1] Most of the patients with impotence in our setting have psychologial factors responsible for impotence and furthermore, social factors prevent these young people from seeking appropriate medical advice in good time.

The management is difficult and includes an accurate clinical assessment and extensive laboratory investigations to diagnose the exact cause of impotence. Intracorporeal pharmacotherapy is a relatively new treatment modality for this problem. It was reported independently by Virag[2] and Brindley.[3] They were the first to suggest self injection. Papaverine was the first drug introduced, but because of the various risks, including prolonged erections and fibrosis, drugs like phentolamine, phenoxybenzamine and others were tried. In recent years, Alprostadil (Prostaglandin E1 (PGE1) has been reported to be an excellent agent. It is locally marketed by the trade name of Caverject. Since then, many researchers have demonstrated that this treatment modality is safe and effective. There is a growing trend towards using intracavernosal drug administration, both as a diagnostic as well as therapeutic measure. It is also recommended that the use of sophisticated tests to define the exact pathology is not indicated. This approach is particularly well suited to smaller centres like ours.[4]

The exact role of PGE1 is still under investigation. However in vitro it stimulates the normal corpora cavernosal function. In vivo, the response has been well-established by many authors with minimal side effects. It is metabolised in the lungs at a rapid pace and does not therefore produce systemic side effects. It, however, is also metabolised in the penile tissue and hence there is a minimal chance of local complications like priapism. Its side effects are infrequent when compared to other intracavernosal pharmacoagents.

PATIENTS AND METHODS

Seventynine patients with psychogenic impotence, attending our out-patient impotence clinic between Ist January 1998 and 3Ist December 1999 were included in this prospective study. The same team of consultants

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assessed all the patients. A thorough history was obtained and general and local physical examination was performed. Important points in the history including marital life, impotence, socio-economic conditions, education, diabetes, hypertension, heart disease, jaundice, mumps, tuberculosis, surgery, local trauma and smoking were noted. Symptoms suggestive of psychological elements include nocturnal emesis, early morning erections and abrupt onset as compared to gradual development of symptoms.[6] This history and examination findings were recorded on an elaborte proforma. Patients with evidence of organic disease were excluded from the study. Patients without any sign of organic disease and with suggestive symptoms were diagnosed as suffering from psychogenic impotence.

A single injection of Alprostadil (PGE1) 10 micrograms was offered to these patients. This was intended to confirm the cause of impotence as psychogenic and to treat the problem by demonstrating to the patients that they were fit. After injection with intrapenile Alprostadil 10 micrograms, erection was graded according to the following table.[7]

Grade 0	No Response.
Grade 1	Some degree of tumescence, insufficient for penetration.
Grade 2	Tumescence sufficient for penetrtion.
Grade 3	Full erection.

Two follow-up visits were routinely planned. The first at one week and the second at 6 weeks. From our earlier experience we knew that our patients were extremely reluctant to ensure direct questions like the number of times they had intercourse. However, they were willing to ensure indirect questions like whether they were satisfied or dissatisfied within their marital life.

RESULTS

A total of 79 patients attended the OPD, who were assessed and believed to fulfil the criteria of study. Out of these, 5 patients were lost to follow-up and were subsequently excluded from the study. Four patients refused to have intracavernosal injections, one for fear of pain and another for fear of disfigurement; two patients assigned no reasons.

Over a period of 2 years, 75 males were injected with postaglandin E1, 70 being available for follow-up. PGE1 is available by the trade name of Caverject. Mean age was 27.4 years (range 18-67 years), two of the patients were above 60. Most of them were from low socio-economic class and mostly uneducated. Only 4 were matriculate and 3 graduates. All of them had nocturnal emesis and early morning erection, which clearly established the cause of impotence as psychogenic.

Following were the results of injection therapy:

Grade 0	02 patients	2.8%
Grade 1	05 patients	7.14%
Grade 2	43 patients	61.4%
Grade 3	20 patients	28.5%

Both the patients above 60 years of age responded in grade 2. In our study, besides pain in 6 patients, we had only 2 complications, one accidental urethral injection and one haematoma which resolved spontaneously. Transient pain that settled with analgesics was felt by 14(20%) of the patients. Only 6(8.6%) had pain severe enough to warrant an extra visit and stay in hospital.

Seventy patients were available for follow-up: On the first visit, 60 out of total 70 patients (86%) were satisfied with the result. Ten patients (14%) including 7 (10%) who had not responded well, were "not happy" with the outcome. Another 4(6%), on their second visit, complained of a renewal of their original symptoms. These fourteen patients were offered a second shot/self injection. Six patients agreed for a second injection. All of them responded in grade 2/3. These six patients were reviewed in out patient clinic after one week and six weeks and they were "satisfied" with their sex life.

DISCUSSION

In our society, psychogenic impotence is a very common problem especially among the young and they are very reluctant to seek medical help. As the medical facilities and literacy rate are improving, awareness that impotence is a curable disease is spreading. Not before long, and still in many parts of the country, the majority would go to the quacks running clinics by glamorous names. Furthermore, as the general duty doctors are not fully trained to deal with these problems and patients keep on suffering.

The most important factor for psychogenic impotence is a lack of proper sex education. Sex education is non-existent in our society and uncommon in the civilised world.[8] Other factors include high self-expectations on the first night of marriage and misconcepts about masturbation. All these lead to first night failure and every ensuing night, the anxiety increases.

Most of our patients were young who recently got married with psychogenic impotence. Single injection therapy of Alprostadil was considered adequate treatment for them. The philosophy behind single injection is to show them that they are normal and once convinced, they had

normal sex life. Our main investigation tool is a thorough clinical evaluation, followed by intracavernosal injection of Alprostadil. In the absence of sophisticated equipment and monetary support, intracavernosal injection is a good diagnostic tool, as is clearly evident from our study.[4] It is also a good therapeutic procedure. It is well received and 94% of our patients agreed to this form of therapy. All the four patients who refused intracavernosal injections were young adults. A single dose of Alprostadil had very good results. Weiss et al noted significant improvement in upto 65% cases.[9]

Some of the laboratory investigations are very useful, albeit very expensive in terms of the cost involved of the sophisticated equipment and work hours of expert staff members. This has forced clinicians to adopt alternative pathways. We believe that it is not essential to pinpoint the exact cause of impotence, but to arrange for a method to improve the sex life of patients. Of course, this method needs to be safe and effective. Alprostadil is achieving this goal.[10]

Alprostadil was first used by Adaikan et al.[11] The response rate was considered very good. This is also supported by our small prospective study. In our series, 61.4% had excellent results, 28.5% good, 7.14% poor response, whereas 2.8% had no response. A 90% percent initial success rate, and an 80% continuing success is very promising in the management of a disease that is difficult to diagnose and treat. We used 10 micrograms of Alprostadil, but a lower (5 micrograms) or a higher (upto 65 micrograms) can be used.[12]

Pain is a very common problem with Alprostadil. Fourteen(20%), of our patients had transient local pain. This settled with conservative management. Pain is reported in similar percentages of patients in large international series.[10] Histologically as well, there is a minimal local inflammatory response.[13] We had only two other complications and they did not need any surgical intervention. However, these two patients were retained in the surgical unit for observation. As 80% of Alprostadil is metabolised by passing through the lungs, it rarely leads to circulatory side effects. Alprostadil is also metabolised by the penis, so a minimal risk of prolonged erection (Priapism).[10] The complications are not very common, but the complications are all surgical. It is therefore imperative that the injectin be first made by a surgeon or a urologist and the patient, if he is going to inject himself, should have liaision with the surgeon in case he gets complications.

Sex is paradoxical, the harder you try, the softer it gets and as is known the most important sex organ is brain. We should not be concentrating only below the belt, but we should also be doing psychotherapy of the patient. This led us to adopt the policy of a single injection therapy, thus demonstrating to the patients that they were perfectly normal. Our stdy confirms that this a valid form of therapy which is received very well even in conservative society like our.

We conclude that intracavernosal pharmacotherapy is a safe and effective tool in diagnosing and treating psychogenic impotence. Time-consuming and expensive investigations are not required in majority of cases clinically suspected of having psychogenic origin. It is an acceptable treatment option. The complications are surgical in nature and we recommend that the injection be made by surgeons.

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BRAIN ABSCESS WITH CYANOTIC HEART DISEASE

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ABSTRACT:
Twentyeight patients of congenital cyanotic heart disease associated with brain abscess , who underwent neurosurgical intervention over a period of 5 years from June 1995 to June 2000 were reviewed. The mean age was 6.32±3.2 years (range 3-14 years) with a male preponderance. Patients having the tetralogy of Fallot were 78.57%. Abscesses located supratentorially were 49.28% with 42.8% predominance in the frontal lobe. Eighteen out of 28 patients were polycythemic with a mean haemoglobin of 16.07±2.7gm% while their arterial oxygen saturation varied between 64%-92%. Twentyone patients underwent CT guided burrhole aspiration, under anaesthetist monitored local anaesthesia, with a repeat aspiration in 12 while aspiration was followed by craniotomy and excision of abscess in seven. Seven underwent primary excision. Mortality in patients undergoing craniotomy for excision of abscess was 35.7% as compared to burrhole aspiration group of 14.2% with an over all mortality of 25%. Morbidity in patients who survived surgery and were discharged was 57.42%.We found burrhole aspiration under anaesthetist monitored local anaesthesia relatively simple, effective and well tolerated in this particular group of patients.The overall prognosis of brain abscess associated with congenital cyanotic heart disease is poor, therefore early recognition of symptoms of brain abscess becomes mandatory.

KEY WORDS: Brain abscess, congenital cyanotic heart disease, burrhole aspiration.

INTRODUCTION

Brain abscess is a well known complication in patients with cyanotic congenital heart disease.[1] Although the first such case was reported in 1814 by Faree,[2] it was Ballet who pointed out the causal association of congenital heart disease with brain abscess.[3] In 1941 Hanna[4] reported seven of his own cases and reviewed 17 from literature. In 1950 Saneeta and Zimmerman[5] reviewed 42 cases from literature and added two of their own. In 1956 Newton[6] was able to review the data in 72 reported cases and seven of his own. In 1957 Campbell[7] reported another 15 cases. In 1983 Kagawa et al[8] reported 62 cases. In 1989 Yang Shu-Yuan[9] reviewed 25 cases and in the same year R.N. Chakrabarty et al[10]. reported 28 cases.

In this study 28 patients of brain abscess associated with congenital cyanotic heart disease, who underwent surgical intervention at Jinnah Postgraduate Medical Centre over a period of five years from June, 1995 to June, 2000 have been reviewed.

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PATIENTS AND METHODS

All patients with brain abscess and congenital cyanotic heart disease, who underwent surgical intervention, either Computed Tomography (CT) guided burr hole aspiration or craniotomy for excision of abscess, over a period of five years from June, 1995 to June, 2000 were analysed for age, sex, type of congenital cyanotic heart lesion, symptomatolgy, pre-operative investigations and location of abscess. Morbidity and mortality in this particular group was also analysed.

RESULTS

One hundred sixtyfive patients with brain abscess were admitted to our neurosurgical unit over a period of five years from June 1995 to June 2000.Thirty five patients (21.2%), had congenital cyanotic heart disease in association with brain abscess. Three were medically treated and discharged while four died within 48 hours of admission, without undergoing any surgical procedure while 28 underwent surgical intervention.

The mean age of the surgical patients was 6.32±3.2 years (range 3-14 years), while their mean weight was 14.21±5.8 Kg. There was a preponderance of males in this series (M:F = 20:8).The most common congenital cyanotic cardiac

lesion was the tetralogy of Fallot (78.57%). The distribution of other cardiac lesions was as shown in Table I.

TABLE-I CARDIAC MALFORMATIONS IN PATIENTS OF CYANOTIC HEART DISEASE WITH BRAIN ABSCESS

Cardiac Malformation	No. of Cases	%
Tetralogy of Fallot 3 with Blalock T. Shunt	22	78.57
Single ventricle	02	7.14
VSD with pulmonary stenosis	02	4.14
Pulmonary atresia	01	3.57
Tricuspid atresia with VSD and pulmonary stenosis	01	3.57

A total of 35 abscesses were found in 28 patients (Table II). Of these 94.28% were supratentorial, with a predominance in the frontal lobe (42.85%) followed by the parietal lobe (28.57%), while 10.7% patients had multiple abscesses.

TABLE-II LOCATION OF ABSCESSES

Location	No. of Abscesses	%
Supratentorial	33	94.28
Infratentorial	02	5.71
Frontal lobe	15	42.85
Parietal	10	28.5
Temporal	07	07
Occipital	01	2.85
Cerebellar	02	5.71
Multiple abscesses	03	10.71

The most common mode of presentation was headache associated with vomiting and altered sensorium in 64.2% while 57.14% had fever. History of effort intolerance and cyanosis was present in all the patients (Table III).

TABLE-III SYMPTOMATOLOGY

Cardiac	No. of Cases	%
Effort intolerance	28	100
Cyanosis	28	100
Cyanotic spell	03	10.71
Headache with vomiting and altered sensorium	18	64.2
Seizures	13	46.43
Hemiparesis	08	28.5
Aphasia	02	7.14
Fever	16	57.14

Investigations carried out in these patients revealed the following abnormalities: Eighteen patients had polycythemia i.e. Hb% above 15 gm%. Their mean haemoglobin was 16.07±2.71% (range 12-21.5 gm%), with a mean haematocrit of 49.15±8.3. Twelve had leucocytosis with a mean TLC of 12732±4,124/mm^3 Electrolyte abnormalities detected preoperatively were hyponatremia in 5 patients and hypokalemia in 4. Their arterial oxygen saturation varied between 64% - 92% (mean 84.7±6.9%). ECG abnormalities were detected in 16 patients while 10 had oligemic lung fields on X-ray. CT scans showed locations of abscesses in all the patients. Pre-operative medication consisted of b-blockers in 17 patients. Antibiotics, along with mannitol were given to practically all the patients.

Twenty one patients underwent CT guided burrhole aspiration under anaesthetist monitored local anaesthesia; repeat aspiration was necessary in 12 patients while aspiration was followed by craniotomy and excision of abscess in 7. Seven underwent primary excision under general anaesthesia.

Seven out of 28 patients died resulting in an overall mortality of 25%. Mortality in the patients undergoing craniotomy for excision of abscess was considerably higher (35.7%) as compared to CT guided burr hole aspiration group of 14.2%, while mortality in patients who did not undergo surgery was extremely high 57.1% (Table IV).

TABLE-IV MORTALITY

Treatment	Total	Died	%
Non surgical	07	04	57.14
Surgical	28		
CT guided burrhole aspiration	14	02	14.28
Primary excision	07	03	42.85
Excision following aspiration	07	02	28.57

Morbidity in patients who survived surgery and were discharged was 57.42% (Table V).

TABLE-V MORBIDITY PATIENTS WHO SURVIVED SURGERY (DISCHARGED)

Outcome	Burrhole aspiration	Primary excision	Excision after aspiration	Total
No neurological deficit	06	01	02	09
Mild to moderate focal dysfunction	06	03	02	11
Functionally incapacitated	00	0	01	01
Total	12	04	05	21

Mild to moderate neurological deficit was present in 52.58% while 4.76% became functionally incapacitated.

DISCUSSION

Although brain abscess is a known complication in patients with congenital cyanotic heart disease with a reported incidence varying between 2-6%[11,12,13] it is not commonly seen in neurosurgical series. Its incidence varies from 3.4% to 13.5% of brain abscesses.[14,15,16] This frequency was found to be relatively higher (21.1%) in our neurosurgical unit. The most common cardiac lesion associated with brain abscess is Tetralogy of Fallot. [3,5,9,17,18] In this series 78.6% of children had this anomaly.

A number of different theories have been postulated as to why congenital cyanotic heart disease has such a high association with brain abscess. Some authors postulate that because of right to left shunt venous blood passes into arterial system without being filtered through pulmonary circulation where bacteria are intercepted by phagocytes.[6,8,11,12,19] It is also thought that the brain itself may be predisposed to offer a focus of infection as a result of encephalomalacia secondary to embolism polycythemia and organisms may be seeded in this infarcted area forming a cerebral abscess.[8,12] The study of Fischbein et al.[11] suggested that low arterial oxygen saturation with polycythemia is the risk factor in development of brain abscess. In this series 66.2% patients had polycythemia with a mean Hb% of 16.07±2.7 gm% (range 12-21.5 gm%) and a mean haematocrit of 49.15±8.38% while the mean arterial oxygen saturation was 84.71±6.09 (range 64-92%), thus confirming the possibility of association of brain abscess with hypoxemia and polycythemia.

The mean age of 6.32±3.2 years with a range of 3-14 years at presentation in this series was similar to the experience of others.[8,20,21,22,23] Rarity of brain abscess in cyanotic congenital heart disease in early childhood may be related to the less frequent occurrence of bacteremia in infancy or dental infection before fully developed dentition.[24]

The predominant presenting complaint in our series was headache associated with vomiting and altered sensorium (64.28%) followed by fever (57.14%), seizure (46.13%) and motor deficit (28.5%). While in R.N. Chakrabarty et al[10]. series fever was the chief presenting complaint (42.86%) followed by headache vomiting and seizures in 32.14% patients.

The supratentorial abscesses in our series were 94.28% with predominance in the frontal lobe (42.85%), followed by the parietal (28.57%) which is in accordance to Balaji et al.[25] series where nine out of 10 abscesses were supratentorial and the most common location was the frontal lobe followed by the parietal lobes. While in Kagawa et al[8] and Yang Shu Yuan[9] series parietal lobe was the predominant location. In Kagawa et al. series 8% patients had multiple abscesses. Yang Shu Yuan reported an incidence of 12.9% while we had multiple abscesses in 10.7% of cases.

We found CT guided burrhole aspiration of abscess under anaesthetist monitored local anaesthesia relatively simple, effective and well-tolerated in patients with congenital cyanotic heart disease with brain abscess and it was offered to 21 out of 28 cases. This treatment has been advocated by a number of neurosurgeons as well.[21,26,27,28]

The prognosis of brain abscess associated with congenital cyanotic heart disease is poor with a mortality rate ranging between 27.5-66%.[3,7,8,11,12,21] Mortality in our patients undergoing craniotomy for excision of abscess was considerably high, 33.7% as compared to burrhole aspiration group of 14.2% with an overall mortality of 25%. Morbidity in patients who survived surgery and were discharged was 57.2%.

Brain abscess still carries a high mortality and morbidity inspite of antibiotic treatment, surgical aspiration, or excision in patients with congenital cyanotic heart disease.[24] Therefore early recognition of symptoms of brain abscess becomes mandatory.[29] Total surgical correction before the age of two years not only corrects cardiac disability but also prevents development of brain abscess.[11]

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CLINICAL PRESENTATION OF PARKINSON'S DISEASE

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ABSTRACT:
A hospital based cross sectional study was carried out at the Department of Neurology, Jinnah Postgraduate Medical Centre, Karachi to study the clinical presentation of Parkinson's disease. All cases presented at the Neurology clinic during the period September 1994 to February 1999 were studied. A total of 200 cases (158 males; 42 females) were studied. The highest number of cases were in the age group 50 years and above. All the cardinal manifestations (rigidity-70%, tremors-69% and bradykinesia-60%) were common. Majority of the cases were in Stage-I and -II of the Hoehn & Yahr scale. Various associated features and patient's occupation have also been studied. The age distribution and the frequency of cardinal manifestations are comparable to the other reported studies. Various associated features have also been noted which together may have functional and psychosocial impact on the life of the patients.

KEY WORDS: Parkinson's disease; cardinal manifestations; associated features; disability staging.

INTRODUCTION

Parkinson's disease (PD) has remained in the forefront of chronic neurodegenerative disorders since the discovery of levodopa replacement therapy in the late 1960s. The overall annual incidence of Parkinson's disease (PD) in the UK[1] is 20/100,000 and prevalence 165/100,000. Up to 80% reduction in nigral dopaminergic neurons have been argued to occur before symptoms appear; if diagnosed below the age of 40 years, it may run a course of over 30 years.[2] The average duration of disease from onset to death being about 11 years.[1] Early detection is not only useful for advising patients about their future but also for the recent claims that selegiline slows the progression of the underlying pathology,[3] James Parkinson in "An essay on the shaking palsy" in 1817 accurately described the features of PD.[4] Clinical criteria is still the main diagnostic tool. In the present study we have looked for the various clinical features of the disease including disability staging.

PATIENTS AND METHODS

All the patients with PD who presented at the Department of Neurology, Jinnah Postgraduate Medical Centre, Karachi, between September 1994 to February 1999, were included in the study. These patients either presented in the Movement Disorder Clinic or were admitted in the Department.

The diagnosis of PD was made on the basis of clinical criteria: two of the three cardinal motor signs of PD (tremor, rigidity, bradykinesia) and consistent response to levodopa.[5] Gibb's exclusion criteria were used for the clinical diagnosis of PD.[6] It included: more than one affected relative, a remitting course, history of encephalitis lethargica, oculogyric crisis, neuroleptic therapy within the preceding year, supranuclear down or lateral gaze palsy, cerebellar signs, autonomic neuropathy, Alzheimer's-like dementia from the onset of symptoms, pyramidal signs explained by other focal neurological disease and evidence of cerebrovascular disease. Autonomic neuropathy was considered as exclusion criteria only if it was the predominant presentation of the disease. Thus the diseases to be excluded include: Essential (Benign, familial) tremor, Shy Drager syndrome, progressive supranuclear palsy, normal pressure hydrocephalus, stroke and Alzheimer's disease.

The disability staging was done on the basis of Hoehn and Yahr Scale 1967[2]: Stage-I, unilateral involvement; Stage-II, Bilateral involvement but no postural imbalance; Stage-III, Bilateral involvement, mild postrural imbalance and independent life; Stage-IV, postural imbalance

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requiring support; Stage-V, severe disease, bed or chair bound. Stage-I and –II are considered mild (without postural reflex compromise) and Stage-IV and -V are considered severe (significant imbalance.)

Further confirmation of the diagnosis was supported by the "UK Parkinson's Disease Society Brain Bank diagnostic criteria™", as given below :-

Step: 1 Diagnosis of Parkinsonism Syndrome
Bradykinesia (slowness of initiation of voluntary movement with progressive education in speed and amplitude of repetitive actions). At least one of the following: muscular rigidity 4-6 Hz rest tremor and postural instability not caused by primary visual, cerebellar, or proprioceptive dysfunction.

Step: 2 Exclusion Criteria for Parkinson's Disease
Parkinsonism due to identifiable causes, such as stroke, head injury, encephalitis, neuroleptic exposure, hydrocephalus, or brain tumor. Oculogyric crises, sustained remissions and supranuclear gaze palsy.
Cerebellar signs: Early severe autonomic insufficiency , early severe dementia and poor response to large doses of levodopa.

Step: 3 Supportive Criteria for PD
Three or more required for diagnosis of definite PD unilateral onset, rest tremor present, progressive signs and symptoms, persistent asymmetry affective side of onset, excellent early response to L-dopa with persistence for > 5 years, L-dopa-induced dyskinesia and Clinical course of > 10 years.

RESULTS

In this study there were a total of 200 cases of PD; 158 males (79%) and 42 females (21%). The male to female ratio was 3.8:1. The frequency distribution according to the age shows that the highest number of the cases (64%) were seen in the age group 50 years and above. The lowest figure was found in the age group 20-29 years. (Table-I).

TABLE-I FREQUENCY DISTRIBUTION

Age	No	%
<30	08	(04.0%)
30-39	31	(15.5%)
40-49	33	(16.5%)
50-59	71	(35.5%)
=/>60	57	(28.5%)

All the cardinal features of PD (rigidity, tremor and bradykinesia) were common in the study. Rigidity (70%) and tremors (69%) were more common than bradykinesia (60%). The disability staging according to the Hoehn and Yahr scale showed that the majority of the cases were in stage-I and –II (27.5% and 30% respectively.) (Table-II).

TABLE-II HOEHN & YAHR DISABILITY SCALE

Stage	No	%
Stage I	55	(27.5%)
Stage II	60	(30.0%)
Stage III	35	(17.5%)
Stage IV	31	(15.5%)
Stage V	19	(9.5%)

A disturbed speech was noted in more than half of the cases. The common presentations were monotonous 121 (60.5%) and 133 low volume (66.5%) speech, followed by slurring in 55 cases (27.5%).

Features of intellectual impairment were also noted in this study. More than 60% of cases were suffering from disturbed attention and memory impairment. Ideomotor apraxia (27.3%), confusion (19.7%) and hallucinations (6.1%) were relatively less common features. (Table-III).

TABLE-III INTELLECTUAL IMPAIRMENT

Stage	No	%
Attention & Concentration	133	(66.5%)
Memory impairment	127	(63.5%)
Confusion	39	(19.5%)
Hallucinations	13	(6.5%)
Ideomotor Apraxia	55	(27.5%)

Associated problems were also noted in this study. Body stiffness, aches and pains, and constipation were reported by >80% of the cases. Osteoarthrosis, dysphagia, postural drop in B.P., depression and visual disturbances were noted in >30% of the patients. In about 20% of the cases there was evidence of urgency and frequency of urine, gastro-oesophagial reflux, hypertension, anxiety, sleep disturbance and dizziness. (Table-IV)

Patients were also asked about their occupation. In males, the highest number of cases were clerks (23.0%) followed by shopkeepers (15.0%.) In females, majority of the patients were housewives. (Table-V)

DISCUSSION

PD is a clinical syndrome defined by the presence of tremor, rigidity and bradykinesia (cardinal manifestations.) The presence of two-third of the cardinal

TABLE-IV ASSOCIATED PROBLEMS

Rheumatological Complaints		
Stiffness	164	(82.0%)
Aches & Pains	176	(87.8%)
Osteoarthrosis	67	(33.5%)
Frozen Shoulder	21	(10.5%)
G.I.T. Complaints:		
Dysphagia	60	(30.0%)
Gastro-oesophageal reflux	43	(21.2%)
Constipation	162	(81.0%)
C. V. S. Problems:		
Myocardial infarction	12	(6.0%)
Hypertension	44	(22.0%)
Postural drop	72	(36.0%)
Low Blood Pressure	18	(9.0%)
PSYCHOLOGICAL Problems:		
Anxiety	40	(20.0%)
Depression requiring treatment	80	(40.0%)
Sleep disturbance	40	(20.0%)
OTHER MANIFESTATIONS:		
Visual (blurring, double vision)	62	(31.0%)
Dizziness	40	(20.0%)
Urgency/frequency of urine	42	(21.0%)

TABLE-V OCCUPATION

	No	%
Males:		
Professionals	18	(9.0%)
Clerks	46	(23.0%)
Shop Keepers	30	(15.0%)
Farmers	24	(12.0%)
Drivers	20	(10.0%)
Laborers	20	(10.0%)
Females:		
Housewives	30	(15.0%)
Working women	12	(6.0%)

manifestations and a robust response to an adequate dose of L-dopa are considered by most experts to be essential for the diagnosis of PD and to distinguish it from other causes of parkinsonism. PD accounts for majority of cases of parkinsonism.[5] Accurate diagnosis is essential for clinical as well as academic reasons. Clinically, it is important to distinguish alternative causes of parkinsonism (specifically Parkinson's-plus syndromes), because treatment response is generally poor and the prognosis of the disease is more rapid. Additionally, some cases of drug-induced parkinsonism, if identified, can be fully reversible. Considering that the diagnosis of PD is still made purely on clinical grounds, it remains a challenging task even for the experts in the field.[7]

In the present study 200 cases of PD were examined in a period of about 5 years. Most cases were males (79%.) In the United States and western Europe it has an approximately equal sex distribution.[8] However, Hopkins[1] observed that PD may be slightly commoner in males at any age, but as women live longer and the incidence increases with age, there are more affected older women. The male preponderance in the present study could be due to selection bias because the study was hospital based.

As mentioned above, tremor, bradykinesia and cogwheel rigidity are considered cardinal manifestations of the disease. Some investigators also include postural reflex impairment in this category.[7] However, we did not include postural reflex impairment in our diagnostic criteria because this feature is also frequently found in Parkinson's-plus syndromes which were excluded from the study. In the present study rigidity (70%) and tremor (69%) were more common than bradykinesia (60%) In many studies tremor and bradykinesia are noted in more than 75% of the cases.[5,9]

Regarding frequency distribution according to age, majority of the cases were in the age group 50 and above. This is in accordance with the fact that the prevalence of the disease increases with the age and affects 1% of the population older that the age of 65 years.[10]

Majority of the cases were in the stage-I and –II (Hoehn and Yahr disability scale). In the DATATOP cohort patient were divided into benign and malignant depending upon the disability scale, duration of illness and involvement of (benign with stage-II or less, duration <4 years and late postural impairment.[11]) Thus majority of our cases were in the benign category.

Speech disturbance was also a common manifestation of the disease. Monotonous and low volume speech was commoner than slurring. Speech disturbance is commonly noted in literature.[1]

Among the features of intellectual impairment attention problem and memory impairment was the commonest manifestation in more than 60% cases. The incidence of cognitive impairment is variably noted in different studies from 7% to 85% with median prevalence of 20%.[2]

Various associated problems were observed in this study. Among them the commonest being body stiffness, aches and pains and constipation in more than 80% cases. The second common manifestations included osteoarthrosis, dysphagia, postural drop in B.P., depression and visual distrubance in about a third of cases. In about one-fifth of the cases there was evidence of urgency and frequency of urine, gastro-oesophageal reflux, hypertension, anxiety,

sleep disturbance and dizziness. In different studies sleep disturbance is noted in 74% to 98% cases.[13] Constipation and dysphagia are noted in more than 50% cases.[14,15] Urinary problems are noted in 58% to 71% cases.[7] Depressive symptoms are commonly noted in PD[16] and major depression is found to be up to 40%.[17] Anxiety is reported in up to 40%.[18] Sensory symptoms including pain are also reported in up to 40%.[19] Visual symptoms are also considered to be common.[7] Orthostatic hypertension is seen in PD without autonomic failure.[20]

We had also asked the patients about their occupations and the cases were common among clerks and shopkeepers. The relevant information was not found in literature.

In conclusion, Parkinson's disease is a commonly recognized disorder in a neurology setting. The age distribution and the frequency of the cardinal manifestations (tremor, rigidity and bradykinesia) are comparable to the other studies reported. Maximum number of the patients were in the stage-I and -II of Hoehn & Yahr scale, which shows a more benign course. Various associated features have also been noted which together may have functional and psychosocial impact on the life of the patients. Among them, the occurrence of aches and pains and constipation was comparable to the other studies but that of dysphagia, depression and sleep disturbances was less observed.

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RISK FACTORS OF CARCINOMA BREAST IN YOUNGER AGE GROUP FEMALES

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ABSTRACT:
A prospective study on carcinoma breast was carried out at Bahawal Victoria Hospital, Bahawalpur from July 1997 to June 2000 and a total of 75 cases were detected. Of these thirty patients (40%) work between age of 25-30 years, an uncommon age group. The common risk factors were absent; all belonged to poor socioeconomic group. Eighty percent of them were from rural area related to cultivation of cotton crop, where use of organophosphorus compounds as pesticides are common. Twenty (67%) presented with lump breast, 17% with ulceration and 13% had nipple discharge. Upper outer quadrant was involved in 23%, UIQ in 17%, IOQ in 13% and two quadrants in 27%. Duration of symptoms was less than six months in 60% of young patients. Most patients (90%) were in stage II and III at presentation. The tumor in these patients behaved aggressively. Intraductal carcinoma was found in 80% of patients.

KEY WORDS: Carcinoma breast, risk factors.

INTRODUCTION

In Pakistan 25% of cancer in females is carcinoma breast.[1] This disease is even more common in females in the West, as 7% of all females suffer from carcinoma breast in life.[2] This study aims at finding clinical presentation, risk factors staging and histopathology in the younger age group patients.

PATIENTS AND METHODS

The study was conducted at Bahawal Victoria Hospital Bahawalpur from July 1997 to June 2000. A total of 75 patients admitted from Surgical out-patient department were included in the study. The various clinical parameters: age, address, sign and symptoms, duration, age of menarche, menstrual history, site and size of tumor and involvement of axillary lymph nodes were noted. Ultrasonography of abdomen, X-ray chest and lumbosacral spine were done to find distant metastases. Bone scan was carried out when needed. Fine needle aspiration cytology was done in all patients before planning surgery. Staging of tumour was done. Treatment given was either modified radical mastectomy or simple mastectomy, radiotherapy and chemotherapy according to stage and need. The mastectomy specimen was sent for histopathology.

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RESULTS

The age distribution of patients is shown in Table I

TABLE-I AGE DISTRIBUTION

Age	No. of Patients	%
25-30	30	(40%)
31-40	09	(12%)
41-50	21	(28%)
51-60	12	(16%)
>60	03	(04%)
Total	75	(100%)

Forty percent patients were between the age of 25 and 30 years, which is a very young age group at presentation. These patients were analysed for risk factors. All the 30 patients were married at an early age (16-20) years. All the patients were multiparous having 4-6 children before the age of 30 years. Only 2 of 30 patients were obese. There was no family history of carcinoma breast. All patients were household females belonging to rural area, 24 (80%) were related to farming, working for cultivation of crops.

Clinical Features are shown in Table II.

TABLE-II CLINICAL FEATURES

Presentation	No. of Patients	%
Lump	20	(67%)
Ulceration	05	(17%)
Nipple discharge	04	(13%)
Backache	01	(03%)

Duration of symptoms is shown in Table III.

Right breast was involved in 14 (46%) and left in 16 (54%) out of 30 patients. Site of the breast involved in the carcinoma is shown in Table IV.

Stage of Disease at the time of presentation is shown in Table V.

Histopathology of the specimen is shown in Table VI.

TABLE-III DURATION OF SYMPTOMS

Duration	No. of Patients	%
06 months	18	(60%)
01 year	07	(24%)
1.5 year	05	(16%)

TABLE-IV SITE OF BREAST INVOLVED

Site	No. of Patients	%
UOQ	07	(23%)
UIQ	05	(17%)
LOQ	04	(13%)
LIQ	03	(10%)
Central	03	(10%)
Two or more	08	(27%)
Total	30	

TABLE-V DISEASE STAGE

Stage	No. of Patients	%
I	-	-
II	18	(60%)
III	09	(30%)
IV	03	(10%)
Total	30	

TABLE-VI HISTOPATHOLOGY

Histopathology	No. of Patients	%
Intraductal	24	(80%)
Intralobular	04	(14%)
Mixed	02	(06%)

DISCUSSION

Carcinoma of breast is the commonest malignant tumor among females. In the West 9.3% of women are at risk of developing carcinoma breast.[3] The incidence of carcinoma breast in Pakistan is 25% of all carcinomas in females, which is far less than that in the West.

In our series 40% of patients belonged to young age group, between the age of 25 to 30 years and 52% were below the age of 40 years, which is a very early presentation. In developing countries the reported incidence coincides with our study.[4] In western countries the reported peak incidence is between the age of 40 to 55 years. Early age carcinoma creates socio-economic problems as these women have younger children to look after and are also anxious to have more children.[5]

The usual presentation in the patients is a lump in breast (67%). Twenty to 25% of patients have lumps larger than 5cm[6] with poor prognosis.[7,8] In our study carcinoma breast was more common on the left side (54%) than the right (46%), as reported in literature.[9] Various reasons described are increase in breast tissue, breast feeding and trauma. In our study OUQ is the usual site (23%) followed by OIQ (17%) and both the regions in 27%, as is also reported in the literature. Skin involvement with oedema of 7.3 cm have poor prognosis.[10]

Most of the patients (60%) had a history of less than six months and were found in stage II or III. The patients are afraid to be examined themselves because of sociomedical habits, lack of education, fear and denial.[11,12] They therefore, have poor prognosis.[13] The short duration of the disease shows that the tumour is more aggressive in the younger age group.[14] Intraductal cell carcinoma was found in 80% and intralobular in 10% of patients. The disparities observed in incidence of the carcinoma breast in different races needs further investigations for the risk factors.[15,16,17]

Younger age group patients tolerate the treatment very well.[18] Poor prognosis is attributed to aggressive tumor, refusal of examination at early stage, breast density in younger age group, breast consistency due to lactation and lactation itself, associated with poor prognosis.[19] The risk factors listed in literature are not relevant in the young patients in our study. The risk factors which lead to sustained high oestrogen level as causative agent are late marriage, late pregnancy, avoidance of breast feeding, family history, use of oral contraceptives and use of alcohol. These are absent in our patients. The factors noted are poor socio-economic conditions and diet factors.[20]

We conclude that carcinoma breast is very common (40%) in early age group patients in our area. Incidence in this age group denies the usual risk factors. Because patients are married early, age of menarche is earlier, early pregnancy, multiparity, non-obesity, no family history and non-use of oral contraceptives are all protective factors. Poor socioeconomc conditions, diet and environmental factors may be involved. The patients in the study belong to rural area related to farming, cultivating cotton crop. This area needs exploration in

future studies.

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A CLINICOPATHOLOGICAL STUDY OF TESTICULAR CANCER

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ABSTRACT:

Seventynine cases of testicular tumour were studied at Chandka Medical College Hospital, Larkana. Their ages ranged from 18 months to 60 years with an average age of 26.85 years. Seminoma patients were older than those with non-seminomatous tumours. The peak age group was third decade of life. Two most common presenting complaints were testicular swelling and abdominal lump. The median time period between onset of symptoms and visit to physician was 6 months. The involved testis was undescended in 05(6.32%) cases. Inguinal and para aotic lymphadenopathy was present in 10(12.65%) and 13(16.45%) cases respectively. Metastasis to liver was present in 6.32% and to lung in 5.01% cases . Average diameter of testicular cancer was 7.49cm . Teratomas were largest and yolk sac tumours the smallest.The two most common histological variants were seminoma (56.98%) and embryonal carcinoma (17.72%). When compared to the West , evidence of metastasis was more frequent in our patients. Median period between onset of symptoms and visit to physician was also longer in our patients.

KEY WORDS: Testicular cancer, clinicopathological, pattern

INTRODUCTION

An overwhelming majority (over 90%) of testicular tumours arise from germ cell.[1] Although the incidence of germ cell tumours is increasing world over with effective treatment, majority of the patients who enter complete remission, can lead a normal life and regain fertility.[1,2] About half of malignant germ cell tumours are seminomas, which are sensitive to radiotherapy. The non-seminomatous tumours also respond well to combined radiotherapy, chemotherapy and surgery even in the presence of distant metastasis. Non-seminomatous germ cell cancers show a wide range of histological appearances. These may present as combination of different histological types.[1] Percentage of embryonal component and vascular invasion appear to be the most significant prognostic features determining the aggressive biological behaviour of germ cell testicular cancer.[3,4] The purpose of present study was to evaluate testicular cancer according to its clinicopathological presentation in patients who were operated at Chandka Medical College Hospital Larkana.

PATIENTS AND METHODS

The study was conducted at the Department of Pathology between January 1989 to December 1999. Cases with incomplete data and those with incisional biopsy alone were dropped from the study. Orchidectomy specimens of all cancer cases were fixed in 10% formalin and representative blocks of tissues were routinely processed for preparation of haemtoxylin and eosin (H & E) stained sections. Histological diagnosis was made on light microscopic examination according to criteria laid down by WHO.[5] Relevant clinical data including X-ray chest and bones and ultrasound (abdomen) were obtained with the biopsy request forms and from the hospital record. The cases were analysed according to age, clinical presentation and histological findings.

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RESULTS

A total 79 cases of testicular cancer were analysed during the stipulated period. Ages of the patients are as shown in Tables-I and II.

TABLE-I HISTOLOGICAL TYPE, FREQUENCY AND AGE GROUPS (YEARS)

Type	0-10	11-20	21-30	31-40	41-45	51-60	No.	%
Seminoma	-	1	18	12	12	2	45	56.98
Embryonal Carcinoma	3	5	2	3	1	-	14	17.72
Malignant Teratoma	-	4	1	2	-	-	7	8.86
Mixed germ cell tumour	-	1	4	-	-	-	5	6.32
Yolk sac tumour	3	-	-	1	-	-	4	5.06
Undifferentiated tumour	-	-	1	1	2	-	4	5.06
No.	6	11	26	19	15	2	79	
(%)	(7.59)	(13.92)	(13.93)	(24.05)	(18.98)	(2.53)	(100)	

TABLE-II INVOLVED SIDE, MEDIAN AGE AND AVERAGE AGE

	Median Age (years)	Average age Years (range)	Right	Left	No	%
Seminoma	35	36 (20-60)	25	20	45	56.98
Embryonal Carcinoma	20	22.67(1.5-45)	5	9	14	17.72
Malignant teratoma	20	24.0(15035)	2	5	7	8.86
Mixed germ cell tumour	26	26.2(19-30)	2	3	5	6.32
Yolk Sac tumour	7	14.0(5-37)	2	2	4	5.06

Their ages ranged from 18 months to 60 years with median and average ages of 29 and 26.85 years respectively. Patients with seminoma were older (average age: 36 years) than those with non-seminomatous tumours (average age : 21.71 years). More than half of the patients were under the age of 40 years. The yolk sac tumours were more common under the age of 10 years. Embryonal carcinoma and malignant teratoma were more frequent in teenagers. The peak age of seminoma, a most common testicular tumour was the third decade of life. Left testis was found to be more commonly involved (53.17%) than the right (46.83%). However seminoma was somewhat more common on the right side. Almost all patients complained of testicular swelling (except one who complained of testicular heaviness), which was painful in majority (49 cases) of cases. (Table-III):

TABLE-III PRESENTING COMPLAINTS

Complaint	No	%
Testicular swelling	78	98.73
Testicular swelling with pain	49	62.02
Abdominal lump	23	29.11
Abdominal lump with pain	5	6.32
Body aches	4	5.06
Cough /haemoptysis	3	3.79
Loss of appetite	3	3.79
Fever	2	2.53
Generalized weakness	2	3.53
Testicular heaviness	1	1.26

The second most common complaint was lower abdominal lump (23 cases) which was painful in a few of them (5). The time period between onset of symptoms and visit to physician ranged from 45 days to 08 years with median period of 6 months. The series included 5 cases of undescended testis and one case of inguinal hernia, all on the affected side. History of testicular trauma , also on the affected side was present in 5 cases.

Clinical notes revealed inguinal and para-aortic lymphadenopathy in 13 and 10 cases respectively. Ultrasound and X-ray findings were suggestive of metastasis to liver 5 cases, lung in 4 cases and bone one case.

Gross features are given in table-IV:

TABLE-IV GROSS FEATURES:

	Cystic component	Cystic softening (necrosis)	Haemorrhage
Seminoma	-	7	4
Embryonal carcinoma	-	4	1
Malignant teratoma	10	2	3
Mixed germ cell tumour	2	3	1
Yolk sac tumour	-	2	1
Undifferentiated tumour	-	2	1
	12 (15.18%)	20 (25.31%)	11 (13.92%)

The diameter of tumours ranged from 2 to 16 cm with average of 7.49 cm. Malignant teratomas were the largest (average diameter: 12.25cm) and the yolk sac tumours were smallest tumours (average diameter: 3.8 cm). Cut surfaces of most of the tumours were grayish yellow in colour. Cystic spaces were present in 12 case, areas of cystic softening (necorsis) in 20 and dark brown heamorrhagic foci in 11. Histological types are given in tables-I, II & IV.

More than half of tumours (56.98%) were seminomas. The second in order of frequency was embryonal carcinoma (17.72%). Foci of embryonal carcinoma were present in 3 out of a total 5 mixed germ cell tumours.

DISCUSSION

Majority of our patients were below the age of 40. Seminoma patients were much older (average age : 36 years) than the non-seminomatous malignancy patients (average age 21.71 years). These figures are not much different from those reported from other parts of the country,[6,7] and from abroad.[8,9]

Almost all patients complained of testicular swelling which was painful in majority of cases. Similar observations are made by Bosl et al.[9] However frequency of metastatic disease (abdominal mass and/or para-aortic lymphadenopathy) was much lower in that study than in the present one. This difference is obviously due to delayed diagnosis in our patients. Frequency of cryptorchidism was lower in our series than that reported in the above mentioned study.[9] According to histological break up the most common tumour in our series was seminoma. The frequency is less in comparison to that of JPMC (Karachi)[6] and a Japanese study,[8] but is higher than that reported from Rawalpindi (38%)[7] and USA (24.6%).[9] The disparity may be due to a sample error or it may even reflect the actual low incidence of seminomatous cancer in those regions.

The percentage of embryonal cells is the most significant prognostic feature of the germ cell testicular cancer.[3,4]This

histological variant was the second most frequent tumour in the present series (17.74%). Foci of this variant were also noticed in 3 out of total 5 mixed germ cell tumours. These observations are more or less comparable to a JPMC experience.[6] Keeping in view the poor state of socio-economic conditions and lack of education, we also analysed the cases according to time period between onset of clinical symptoms and presentation of patients to the physician. The median value of this period was much higher in our patients (6 months) than that of the western group (36 days).[9]

We observed that the clinicopathological pattern of testicular cancer in the present study was more or less comparable to that of other studies in many respects. But it was different from that of western study[9] in at least two aspects: Firstly the clinical evidence of metastatic disease was more common in our patients and secondly the time period between onset of the symptoms and the arrival of patient in hospital was much longer in our cases than those of the west. Since both of these factors are indicative of poor socioeconomic conditions and lack of education, our recommendations are similar to those of J.Talati[10] and Bosl et al[9] that the logical approach to testicular cancer should include testicular self examination, early visit to doctor, ultrasound screening of testis and abdomen and the histopathological examination of testicular swelling. These objectives can be achieved by better education and highten awareness of the disease by both patients and physicians.

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CLINICAL EXPERIENCE OF MOTOR NEURONE DISEASE

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ABSTRACT:
Motor Neurone Disease (MND) is an adult onset, idiopathic, progressive and ultimately fatal degenerative neurological disorder, involving both upper and lower motor neurons in the cerebral cortex, brain stem and spinal cord. Recent studies revealed additional degeneration of some other areas of brain. No definite cause or treatment is known. Clinically it has been shown to have different onset forms, which frequently evolve into a common clinical entity known as A.L.S. This study, describes clinical pattern of MND in a portion of Pakistani population who presented at Neurology Department of Civil Hospital Karachi during the period 1992 - 1998. Total of 29 cases fulfilled the diagnostic criteria; all the cases were sporadic, except an elderly male patient with pure lower motor neuron illness. Twentyeight cases presented with combination of upper and lower motor neuron signs. Of these, 8 had bulbar onset and later spinal features. Twenty had spinal onset, out of them 7 developed additional bulbar symptoms and 13 retained the features of onset at the time of presentation. Overall mean age of onset was 43±9 years, about a decade younger than European population and nearer to South Asian studies. Spinal onset form affected mostly young males (male: female ratio 19:1) and bulbar onset form affected elderly patients with equal sex ratio. However male to female ratio, considering all cases, was 4.8:1. Females were observed to be relatively spared during the reproductive age period. Rate of progression was slow to moderately rapid. Only two patients had fulminant course.

KEY WORDS: Clinical Pattern, Motor Neuron Disease.

INTRODUCTION

History of MND dates back to more than 100 years when it was first described by Charcot who worked with Joffroy and Gambault during the period 1869 to 1871. In the ongoing years, it has been observed that motor neurons may undergo degeneration in a variety of conditions, which may be acquired or familial with onset in infancy, childhood, young adulthood or late adult life. But the term Motor Neuron Disease (MND) has been used as a generic term for a more homogenous condition that is idiopathic and progressive, has its onset in adult life and is almost always inevitably fatal. Depending upon its presenting features it has been classified as Progressive Bulbar Palsy, Progressive Muscular Atrophy, Amyotrophic Lateral Sclerosis.

On epidemiologic and genetic factors it has been grouped as sporadic 90-95%, familial 5-10% (Mariana island form). Recently authors have been using ALS as a generic term for adult onset MND because it has been observed that whatever may be the mode of onset, there is a clear tendency to evolve into a common entity with features of ALS. Hence a new concept of classification is to name ALS according to the mode of onset, viz bulbar onset ALS and spinal onset ALS. This study also follows this concept of nomenclature.

Clinicopathological studies reveal that motor neuron disease is associated with degeneration of motor neurons in the spinal cord, brain stem and motor cortex in varying combination in individual cases. Characteristically, extraocular muscles, sphincters and sensations remain preserved. Some studies based on necropsy findings report the degenerative changes were also found in amygdala and parahippocampus in more than 30% of cases without any significant history of cognitive impairment. There are also reports of similar changes in anterior thalamus and medial frontal region; such patients showed selective cognitive impairment. These findings broaden the scope of degenerative process of MND.

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No single concept of cause or pathogenesis of MND has been accepted, however suspected causes include: slow virus infection, heavy metal intoxication, decrease in neuronal androgen receptors, trauma, radiation and electric shock. Glutamate excitotoxicity, free radical oxidative stress, neurofilament accumulation and autoimmunity have been proposed as possible pathogenetic mechanisms.

Five to ten percent cases have been found to be familial with autosomal dominant or autosomal recessive inheritance. Mutation in the genes for SOD –I on chromosome 21q have been found in dominant cases. No definite treatment is available. Recently a benzothiazol derivation Riluzole was claimed to have neuroprotective effect; but no significant improvement in morbidity has been reported The clinical presentation of MND has changed very little. However in the last 50 years slight regional variations has been reported in literature. The purpose of the present study is an attempt to define the clinical pattern of MND in a portion of Pakistani population with special reference to mode of onset, age of onset, sex incidence, rate of progression and to find out any variation from data presented in international literature.

PATIENTS AND METHODS

This is a hospital based study and includes cases admitted in the Department of Neurology, Civil Hospital Karachi during the period 1992 - 1998. Among a total of 29 cases, 24 were males and 5 females. Cases were selected according to a standard diagnostic criteria fulfilling clinical and electro-neurophysiological features of MND.

Routine hematological and biochemical screening was done in all cases. Suspected cases were additionally investigated by MRI Brain and Cervical Cord, and Serum CPK, to exclude other clinically simulating diseases. Studied cases were classified according to recent concept of nomenclature of MND. Result analysis was compared with other studies mentioned in literature.

RESULTS

Out of 29 cases, 8 had bulbar onset and 20 had spinal onset. One patient had pure lower motor neuron illness with features of progressive muscular atrophy. Overall male to female ratio was 4.8:1. Sex ratio in different onset forms is shown in Table I.

TABLE-I SEX RATIO IN DIFFERENT ONSET FORMS (N= 29*)

Onset form	Males	Females	Total
Bulbar onset	4	4	8
Spinal onset	19	1	20

***One male patient had pure L.M.N illness; overall mean age of onset was 46.3 years.**

Age in spinal onset form was 41.8 years and in bulbar onset 55.5 years. Age distribution in males and females is shown in Table II.

TABLE-II MEAN AGE OF ONSET AND SEX RATIO (N= 29*)

Onset form	Male (Years)	Female (Years)
Bulbar onset	54.6	41.5
Spinal onset	56.5	48

***One male patient with pure LMN illness was 65 years old.**

Total duration of illness from onset to the time of presentation at OPD, as shown in Table III, indicates that most patients presented between 1-2 years of onset of illness.

TABLE-III DURATION BETWEEN ONSET OF ILLNESS AND PRESENTATION IN OPD

Duration	No. of patients
1-6 months	8
7-12 months	8
13-24 months	10
25-36 months	3

Irrespective of mode of onset, analysis of symptoms at first presentation in OPD as shown in Table IV, indicates that quadreparesis was the most common spinal symptom in males, while dysarthria and dysphagia were equally distributed in both sexes.

TABLE-IV CLINICAL SYMPTOMS AT THE TIME OF PRESENTATION

Symptoms	Male	Female
Dysphagia	10	5
Dyrarthria	10	5
Emotional Lability	3	1
Weakness of one arm only.	1	0
Weakness of both arms only.	2	0
Weakness of one leg only.	1	0
Weakness of both legs only.	1	0
Weakness of one side of body.	1	0
Weakness of three limbs.	4	0
Weakness of all four limbs.	10	1
Pain in muscles (stretching).	3	0
Heaviness in limbs.	1	0
Twitching of muscles.	13	1
Wasting of one or more limbs.	16	1
Breathlessness	1	0

Possible important events in the past history are mentioned in Table V. Their possible etiologic importance has been discussed in literature.

TABLE-V POSSIBLE IMPORTANT EVENTS IN PAST HISTORY

Past history.	Number of patients.
Exposure to pesticide	4
Exposure to Insecticide (accidental drinking)	1
Exposure to spray paint.	1
Trauma to head and spine.	4
Electric shock.	1

The salient clinical sign characteristic of illness observed in studied patients are shown in Table VI.

TABLE-VI CLINICAL SIGNS CHARACTERISTIC OF MND

Past history.	Number of patients.
Mood	
Normal.	23
Depressed.	2
Labile.	4
Speech	
Normal.	13
Dysarthric.	12
Mute.	4
Tongue	
Normal.	13
Only Fasciculation	9
Only wasting.	0
Wasting + fasciculation.	4
Difficulty in protrusion.	3
Muscle bulk	
Normal.	10
Wasting of limbs.	19
Muscle Tone	
Normal.	11
Increased.	11
Decreased.	7
Visible Fasciculation	
Absent.	10
Present.	19
Power	
Normal	2
Decreased in limb/limbs.	27
Deep tendon reflexes	
Normal.	4
Brisk.	24
Depressed.	1
Plantars	
Both Flexors	14
Both Extensors	10
Both Equivocal	3
One sided extensor.	1
No response.	1

A correlation of mood changes with bulbar reflexes and of deep tendon reflexes with flexor plantars is shown in Table VII.

TABLE-VII MOOD CHANGES AND BULBAR REFLEXES

Jaw jerk and Gag Reflex		Normal mood	Depressed Mood	Labile Mood.
Normal	23	23	---	---
Brisk	6	1	1	4

DISCUSSION

Overall male to female ratio was 4.8:1. Raib and Jolly reported male to female ratio of 6:1.[1] Sahadevan reported an equal incidence.[2] Overall mean age of onset in present study was 46.3 years. Brain et al reported 55 years.[3] Gamel et al mentioned 52 years[4] and Brown RH. Jr. reported mean age of onset as 55 years.[5] J.S.Chopra from India, reported mean age of onset as 46.9 years.[6] Therefore it was about a decade less than European population but nearer to Asian study. It is important to note that more than half of the patients in present study had onset of illness between 31 to 50 years. Considering specific onset forms, the mean age of onset in case of spinal form was 41.8 years and in case of bulbar onset form 55.5 years. It was observed that bulbar onset form was the disease of elderly patients with equal sex ratio. Conversely spinal onset form was observed in males. Regarding specific onset forms, in case of spinal onset form (also labelled in literature as ALS), male to female ratio in present study was 19:1 which was dramatically different from Europian study[7] (i.e.) 1.6:1. But sex ratio in case of bulbar onset form (also labelled in literature as Progressive bulbar palsy) was 1:1 which was similar to western studies.

Only one male patient presented with pure lower motor neuron illness suggestive of progressive muscular atrophy. In this study the youngest patient was 15 1/2 years and eldest 70 years of age. From USA Mulder has noted that his youngest patient was of 20 years.[8] Sahadevan's youngest patient was 14 years old.[2] The youngest patient of Brain et al was of 17 years.[3] There was no patient below 20 in the study report of Mortara et al.[9] Hence the younger age of onset in present study was nearer to that of South Asian studies.

The most interesting observation appeared to be the distinct sparing of females during their reproductive age.This study shows that between the age of menarche and menopause only two females were affected. Most females affected by bulbar onset ALS were above 50 years and most males who were affected by spinal onset form were between 31 – 50 years. It has been postulated that loss of motor neurons in MND took place from a combination of an age related decrease in androgen level and diminished number of neuronal receptors.

Regarding physical signs at the time of presentation, 51.7% patients showed bulbar signs, 75.8% patients presented with asymmetrically evolving weakness and 58.6% with wasting of one or more limbs. Fasciculations were presented in 48.2% cases. Regarding upper motor neuron signs, it was found that spasticty and brisk reflexes were more commonly present than extensor plantars. Heterogenity of signs, almost similar to that presented in literature, was also noted. Most of those patients who had mood changes also had brisk jaw jerk and gag reflex. Among the cranial nerves, most commonly affected were VII and XII nerves. One had V nerve palsy and none had XI palsy. No patient showed external ocular muscle weakness or sphincter disturbance; sensations were intact in all patients. One patient presented with dyspnoea in whom no other cause of respiratory difficulty was apparent, except diphragmatic weakness. Motor and sensory nerve conduction studies were normal in all cases. The overall course of illness and mortality could not be ascertained because of lack of follow up in most cases after they were explained the prognosis. Most of the patients presented between 1 to 2 years of onset of illness. Two patients had very fulminant course and became bed bound within 6 months of onset; one of them had respiratory muscle involvement. Among the total number of cases, only four were non ambulant, others were able to walk with support. Referring the concept of nomenclature of MND, the present study supported the recent view of using ALS as generic term, because 28 out of 29 patients presented with combined features of upper and lower motor neuron lesions. Most of the patients included in this study showed clear tendency to evolve into common entity of ALS, regardless of mode of onset. However spinal form of onset was more common than bulbar onset forms and involved mostly young males as compared to elderly males and females in case of bulbar onset form. Pure lower motor neuron illness was found in a single elderly male but no case was found with isolated upper motor neuron illness. Among lower motor neuron signs, weakness was more common then wasting and fasciculations. Similarly, spasticity and brisk tendon reflexes were more common upper motor signs as compared to extensor plantars. Mood changes were found in association with brisk jaw jerk and gag reflex. Rate of progression varied: slow, moderately rapid and very rapid.

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CIRRHOSIS OF LIVER IN ASYMPTOMATIC PATIENTS

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ABSTRACT:

Autopsy experience shows that cirrhosis may be entirely asymptomatic and in life it may be found incidentally at surgery.[1] A study was carried out on 100 patients, selected at random, who underwent laparotomy. Purpose of study was to find the incidence of liver cirrhosis in asymtomatic patients, diagnosed on laparotomy and identify the risk factors (i. Viral hepatitis B and C, ii. Alcohol, iii. Toxins and drugs e.g., methotrexate.) Out of 100 patients, 54 patients were females and 46 males, with a mean age of 33.8 years. Seven patients had asymtomatic cirrhosis, 2 were hepatitis B positive, 4 were hepatitis C positive and one was alcoholic. Main cause of asymtomatic cirrhosis was Hepatitis B and C.

KEY WORDS: Asymptomatic cirrhosis, Laparotomy, Hepatitis B and C.

INTRODUCTION

Hepatic cirrhosis is necrosis of liver followed by fibrosis and regeneration of liver cells involving the whole organ.[2] Chronic liver disease resulting in liver cirrhosis is fairly common in our country.[3] The disease may be discovered at routine examination or biochemical screen or at operations undertaken for some other condition.[4] Hepatitis B virus, alcohol and some drugs are most common causes of cirrhosis. Alcoholic cirrhosis may also be clinically silent. In fact 10% cases are discovered incidentally at laparotomy or autopsy.[5]

Viral hepatitis is the most common liver disease today and constitutes a world wide problem. Hepatitis B virus is endemic in the entire human population and hyper-endemic in many parts of the world.[6] HBV is 100 times more infectious than HIV.[7] It has caused the disease in surgeons and nurses as a result of accidental prick by contaminated needles. The amount of blood or serum necessary to transmit the infection is as little as 0.01 ml.[8] Contaminated needles and medical equipment can be the cause of HBV transmission even months after being soiled by the virus.[9] Hepatitis C virus acquired from donor is a common complication of unscreened blood transfusions.[10] It is responsible for 80% of cases of post transfusion hepatitis.[11]

We studied the incidence of cirrhosis and its risk factors in asymptomatic patients with liver biopsy who underwent laparotomy.

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PATIENTS AND METHODS

This study was carried out in Surgical Unit-1, Rawalpindi General Hospital from June 1997 to December, 1998. A total of 100 patients were selected at random irrespective of their age, sex, diagnosis and mode of admission. History of jaundice, alcohol intake, drug intake, weight loss, decrease in appetite, melena, abdominal distention, breast swelling, loss of libido and amenorrhoea were recorded. Previous history of blood transfusions, operations, injections, vaccinations and contact with hepatitis patients was taken. Relevant information regarding obstetric history, ear or nose piercing in females, circumcisions in males by quacks, regular visits to barbers or any other surgical or dental procedures were recorded. Patients were also enquired about vaccination against hepatitis B.

These patients were further investigated for risk factors i.e. Viral hepatitis B and viral hepatitis C, alcohol, toxins and drugs e.g. methotrexate and their liver function tests were done.They were also examined for palmar erythema, clubbing, dupuytren's contracture, spider naevi, gynaecomastia, hepatomegally, ascites, oedema, dilated veins and testicular atrophy. Such patients with any manifestation of cirrhosis were excluded.

Liver biopsy was taken in those patients whose abdomen was opened through midline or right paramedian or right subcostal incision as liver was easily approachable.

RESULTS

Out of one hundred patients included in this study, 54 were female and 46 male with mean age of 33.8 years (range 5 to 62 year), with a peak observed in the third decade of life (Table I).

TABLE-I AGE AND SEX DISTRIBUTIONS OF THE PATIENTS

Age	Male	Female	Total
0-9	5	3	08
10-11	7	8	15
20-29	15	12	27
30-39	6	6	12
40-49	7	8	15
50-59	9	7	16
60-69	3	4	07

Sixty patients were admitted for emergency surgery while 40 patients underwent elective operation. None of the patients gave history of jaundice. On gross examination of liver, out of these 100 patients five had macro nodular cirrhosis, their biopsy reports also confirmed cirrhosis. Two patients were declared cirrhotic by histo-pathologist. A total of 7 patients (7%) were found to be cirrhotic who were totally asymptomatic and did not show any sign or symptom of cirrhosis. Out of these 7 patients, 4 were males and 3 females. These patients were further investigated for risk factors. Two patients were positive for hepatitis B surface antigen, 4 patients had positive hepatitis C virus antibodies (by ELISA Method). One patient was alcoholic.

Out of these 7 asymptomatic cirrhotic patients, 2 had history of surgical operation before admission. Five patients had undergone dental treatment from various hospitals and quacks (3 patients were anti HCV positive and 2 Patients were HBsAg positive). All the three female patients gave history of ear piercing. All these asymptomatic cirrhotic patients had history of injection or accidental needle stick injuries.

All 4 males were regular visitors to barbers (3 patients were anti HVC positive and one was HBsAg positive). None of the patients gave history of toxins and drugs e.g. Methotrexate and Amiodarone or blood transfusions. Liver function tests were done in these 7 patients: 3 patients had normal levels, 2 patients had elevated Gamma G.T. and Alkaline phosphatase, one patient had raised A.L.T. (SGPT). One patient had elevated level of all these enzymes (ALT, ASTand Alk Phos).

DISCUSSION

The death rate from cirrhosis of the liver exceeds 23000 per year in the USA and the incidence of the disease is increasing. At present it is the third most common cause of death in men in fifth decade of life.[12]

Cirrhotic patients may be asymptomatic and are diagnosed incidentally. Not uncommonly the condition will be unsuspected and comes to light at laparotomy. Merican et al, University Department of Medicine, Royal Free Hospital London saw cases with chronic active hepatitis C and declared one quarter had no symptoms of liver diseases and were generally asymptomatic or had presented with no specific complaints.[13]

Manna and Lessof have reported that cirrhosis may be far advanced before it produces any symptoms. It is frequently found at post mortem in patients dying from other conditions who have complained of no symptoms referable to the liver during life.[14] An incidental finding in many liver biopsy and autopsy specimens from tropical centers, especially in Africa, the frequency of the finding of liver cirrhosis is considerably greater than it is in non-tropical regions.[15]

According to Cuschieri et al, "Not uncommonly the condition will be unsuspected and comes to light because of a routine estimation of liver function tests or found incidentally at laparotomy".[6]

In our study out of 100 patients five patients had macro nodular cirrhosis, confirmed by biopsy reports. Two patients were declared cirrhotic by histo-pathologist. So total 7 patients (7%) were found to be cirrhotic and they were totally asymptomatic. Preoperatively they did not show any sign and symptom of cirrhosis.

Considering the causative factors of cirrhosis over the past two decades various studies have been published on hepatitis B and hepatitis C. The reported carrier rate has increased over the past decades is partly due to improved laboratory techniques applied for their detection and partly due to wide spread of the disease. The carrier rate of HBsAg in Pakistani adults are 0-14%.[16] In a study of 789 surgical patients in Japan, 129 (16.3%) patients were positive for HCV.[17] In another study conducted on 100 surgical patients by Azam et al, 23% patients were HBS Ag positive.[9] In our study these 7 asymptomatic cirrhotic patient were investigated for HCV and HBV. Four were anti HCV positive and two were HBs Ag positive.

Occurrence of cirrhosis in asymptomatic chronic hepatitis B virus carrier has been studied in Taiwan between November, 1980 and May, 1990. A total of 1506 cases of HBs Ag positive were followed and 89 of liver cirrhosis were seen. In these cases 16 patients developed hepatocellular carcinoma. The significant risk factors found for liver cirrhosis in HBs Ag carrier were age, hepatitis B antigen carrier status, chronic hepatitis, cigarette smoking, non-A blood types and low educational status.[18] In our study two out of 7 asymptomatic cirrhotic patients were HBs Ag +ve.

In another study in Italy, 15% of asymptomatic subjects had histological evidence of cirrhosis and among them 91.4% had viral aetiology. In a second study which enrolled cirrhotic patients from 13 centers from all regions of Italy, viral infections were detected in 82.6% of patients, the large majority of whom (71.2%) were positive for hepatitis C virus. Alcohol abuse was present in 8.17% cases.[19]

Similarly, a study was conducted in Korea on asymptomatic chronic HBs Ag carriers and it was seen that out of a total 110 asymptomatic carriers, 27 had histologically normal liver, 51 had chronic liver disease and 32 had non-specific histological abnormalities. Out of 51 patients with chronic liver disease only 3 had liver cirrhosis.[20] The most common routes of transmission of diseases are through contaminated needles used for parenteral medications and immunization, blood and its products, sharp surgical instruments, surgical and gynaecological procedures, dental treatment, regular visits to barber, sexual contact with hepatitis patients.

Ramos et al published a study of 4278 patients of parenteral exposures to blood or body fluids and among them dental students and dental assistants had the highest rates of exposure. In Pakistan dentistry staff had a prevalence rate of HBs Ag 16.0%[21] and 17%[22] in two different studies. In our study 5 patients had undergone dental treatment. (3 patients were anti HCV +ve and 2 were HBs Ag +ve).This shows a close relationship of dental treatment and frequency of hepatitis virus and mostly the syringe needle injuries while giving injections, cleaning instruments after procedures and drilling are the activities most frequently associated with exposures.[23]

Use of one needle to inject drugs in many patients can initiate a chain of infections. In Pakistan reuse of disposable syringes is a common practice and recent reports in press of repacking of disposable syringes is frightening.[9] In our study all of these asymptomatic cirrhotic patients had a past history of more than 10 injections per year. This suggests that sources could be either injections or some other factors.

In a study conducted in Bangladesh for post transfusion hepatitis 29.1% patients were HBV positive and 6.8% case were anti HCV positive.[24] In another study 35.71% patients were HBs Ag positive after blood transfusion.[9] This shows the close relationship of acquiring hepatitis B and hepatitis C after blood transfusion. Wherever voluntary blood donor and proper screening have been followed, incidence of post transfusion hepatitis had declined.[25] In our study, none of the patients had history of transfusion.

Cirrhosis of liver is a common problem. Some of the cirrhotic patients remain asymptomatic and process of necrosis and fibrosis continues without any manifestation. In these patients liver cirrhosis is diagnosed incidentally when liver is biopsied or discovered on postmortem examination. Hepatitis "B" and hepatitis "C" are the commonest causative agents in our society.

It is recommended that:-

- All patients who are undergoing surgery should be screened for HBs Ag and anti HCV antibody.
- Sterilization of surgical instruments and endoscopes should be done properly.
- Injection with disposable syringes should be encouraged and their reuse should be condemned and banned.
- Screening for hepatitis "B" and hepatitis "C" should be done in patients undergoing surgery or dental treatment and in blood donors before transfusion.
- Surgeons should adopt preventive methods in order to limit transmission of these viruses.
- All surgeons should be vaccinated against Hepatitis "B" and should know their sero status.
- Alcoholism should be discouraged.

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MALROTATION OF THE INTESTINE

SIKANDAR MUGHAL, UBEDULLAH MAGAN

ABSTRACT:
This is a studyof 15 patients of malrotation operated in the Department of Paediatric Surgery, Chandka Medical College Hospital Larkana, between January 1996 to June 2000. The patients were divided into three age groups: newborns(3), infants(7) and older children(5) because of the difference in their clinical presentations. Associated anomalies were present in 2 patients (13.3%). Upper GI study was the main diagnostic investigation. All patients underwent laparotomy. Resection and anastomosis was done in one newborn with volvulus and necrosis of bowel; Ladd's procedure was carried out in all other patients without appendectomy. Postoperative recovery was quicker in older children. Postoperative complications occurred in 4 patients (27%) and 14 patients got total relief of symptoms; only one patient had persistent GI symptoms even after surgery. There was no mortalty in the series. Follow up ranged from 3 months to 3 years.

KEY WORDS: Intestine Malrotation, Midgut Volvulus, Ladd's procedure

INTRODUCTION

Malrotation is the term used for a series of abnormalities of intestinal rotation and fixation. It ranges from a mobile caecum with the duodenojejunal junction to the right of the spine, to a complete nonrotation with an associated midgut volvulus. Regarding embroylgical evolution of malrotation, many theories have been suggested.[1-3] Normal development of the midgut results in rapid growth and umbilical herniation during the sixth week. During weeks 10 through 11, the gut begins its 270° counterclockwise rotation about the superior mesenteric artery and re-enters the abdomen. By the 12th week, fixation occurs. Malrotation occurs when this normal rotation fails.[4] Ladd's procedure, as described in 1936,[5] is still the basis for the surgical treatment of malrotation today. The aim of the present study was to find the difference in clinical presentation of malrotation in various age groups, the results of surgery and postoperative course and complications.

PATIENTS AND METHODS

In this study we only included cases of isolated malrotation. Malrotation associated with omphalocele, gastroschisis or congenital diaphragmatic hernhia was excluded to obtain a uniform patient pattern. Fifteen patients operated for malrotation between January 1996 to June 2000 in the Department of Paediatric Surgery, Chandka Medical College Children Hospital Larkana, were included in this study.

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Patients were divided into three groups: newborns (group-I,3), infants (group-II, 7), and patients older than 1 year (group-III,5). The age range in the series was from 1 day to 2 years. Mean age of patients in group-I was 8.7 days, in Group-II, 3.5 months and in Group-III, 1.9 years. Of 15 patients, there were 3 girls and 12 boys. Associated anomalies were seen in 2 patients, which were duodenal atresia in one and imperforate anus and duodenal atresia in another patient.

The presenting features in three groups were different; newborns presented with signs and symptoms of acute intestinal obstruction, infants presented with complaint of repeated episodes of bilious vomiting and failure to thrive and the older children presented with complaint of bilious vomiting and subacute intestinal obstruction. Group-II and III patients were malnourished. Only plain Xray abdomen was done in newborn babies; in other patients (group-II and III) upper gastrointestinal contrast study was done, which was diagnostic in all cases.

All patients were operated upon through right supraumbilical transverse incision. Ladd's procedure was performed in all patients, comprising of evisceration and inspection of the mesenteric root, anticlockwise derotation

in case of midgut volvulus, lysis of th Ladd's bands with straightening of the duodenum along the right paracolic gutter and placement of the caecum into the left lower quadrant alongside the sigmoid. However, appendicectomy was not performed in any patient. Out of 15 patients, 2 had duodenal atresia type 1, which was surgically corrected. A pelvic colostomy was done in patient with imperforate anus. One patient had multiple ileoileal intussusception which were reduced manually at the time of operation. Followup of patients ranged from 3 months to 3 years.

RESULTS

Postoperatively nasogastric tube decompression by free gravity drainage was necessary for an average of 7 days in group I, for 6 days in group II and 4 days in group III. Resumptin of bowel activity as shown by time of gastric tube decompression was faster in the group of older children. Of group I & II patients had midgut volvulus; one required resection and anastomosis due to gut necrosis. Of group II & III, all patients had Ladd's bands causing duodenal obstruction. Mean hospital stay in group I was 9 days, in group II, 6 days and in group III, 5 days. Postoperative complications were seen in 4 patients, which were adhesive bowel obstruction in 2 patients, wound infection in one patient and persistent gastrointestinal sysmptoms (of abdominal pain, vomiting and feeding difficulty) in one patient. Intestinal obstruction due to adhesions was relieved on conservative treatment in one patient but required operation in the other patient. There was no mortality in this series.

DISCUSSION

Malrotation is a serious and frequent cause of intestinal obstruction in the nursery. It is a major cause of obstruction throughout infancy and the early childhood period. The embryology of malrotation was described by Mall[6] in 1898 and the first understanding between anatomy and clinical outome was described by Dott[7] in 1923. Two series of case reports precede the classic paper written by Ladd in 1936 describing treatment of malrotation.[5] The diagnosis of malrotation is not a problem in newborns as majority of them present in acute condition. However, in infants and older children, much time is lost before diagnosing the disease. Bilious vomiting and bloody stools are said to be the most common clinical presentation in neonates, while bilious vomiting, recurrent abdominal pain and failure to thrive are the most common symptoms after the newborn period.[7] The associated anomalies are seen in 30-62% of reported series of malrotation[8] and in the present study, the associated anomalies were detected in 2 patients (13.3%). Malrotation is a major diagnostic challenge in children,[9] which historically is made using a barium enema to describe the position of the caecum. However, the variation of the normal caecal position makes the examination difficult to interpret. Therefore, an upper gastrointestinal series (UGI) has become a more reliable examination and still remais the standard modality for diagnosis. The UGI may demonstrate equivocal findings with an abnormal duodenal sweep, which may fail to cross the midline or follow a wandering path.[10] Ultrasound scan can be used to support the diagnosis of malrotation by describing an inverse relationship between superior mesenteric artery and vein,[11] the so called "whirl pool" pattern of SMV, but is not yet a sufficiently reliable test to screen for malrotation.[12]

If the ligament of treitz (LOT) is in an equivocal position and not to the left of the spine at L1 or L2, it is important to determine the position and fixation of the caecum to assess the breadth of the mesenteric pedicle. Laparoscopy can determine the position of LOT and whether the caecum is fixed in the right lower quadrant.[13] Laparoscopy may help to verify the diagnosis in patients who do not have classic radiographic findings.[14] The factors associated with an increased risk of mortality are: presence of necrosis, presence of other abnormalities and younger age.[15] The four steps of the Ladd's procedure require much handling and manipulation of the bowel. The consequent adhesions resulting from the operations are thought to be advantageous because they are felt to stabilize the bowel in its new position. Malrotation in the newborn is an anomaly for which there are clear indication for surgery; however the management of the older patient with this entity is not well defined.[16] At the time of operation, the situation most feared is volvulus with necrotic bowel.[17] In such a case, resection of bowel is associated with a higher incidence of mortality and complications. Presence of volvulus is said to delay postoperative return of bowel function and many gastrointestinal symptoms remain in one quarter of patients postoperatively. In many cases malrotation is associated with intestinal motility dysfunction. This intestinal dysmotility as a postoperative complication has been reported by many surgeons.[18-20] Therefore, performing Ladd's procedure does not guarantee a cured child.[17] Hence the need for a prolonged and careful followup in these patients. Division of patients into three age groups in our study was based on difference of their clinical presentations. This is in contrast to other studies which speak of only two age groups i.e. patients yunger than one year and patients older than one year. Subacute intestinal obstruction was a new clinical presentation observed only in our older children. Appendicectomy, as part of classical Ladd's procedure, was not performed in any case because of poor condition of health in these children. Postoperative recovery in our study was quicker in oldr children as compared to newborns and infants; this is in conformity with other studies.[17] Total relief of symptoms occurred in 64% of patients in one series.[16] In our study, total relief of symptoms occurred in 14 patients (93.3%); only one patient has persistent gastrointestinal symptoms even after surgery.

In conclusion, malrotation should always be considered in differential diagnosis of infants and children who present with bilious vomiting and failure to thrive. Upper GI contrast study still remains the mainstay of diagnosis and surgery has good results in simple malrotation.

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SPINAL A-V MALFORMATION

A CASE REPORT

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AMANULLAH, HUSSAIN HAROON, SABEEN NAZEER

ABSTRACT:
Spinal A-V malformation is rare disease, but if diagnosed earlier, surgery or chemoembolization or both can prevent the neurological deficit (paraplegia). A young lady presented with a history of sudden loss of power and impaired sensation in both lower limbs for 3 hours. Routine investigations were normal, X-ray dorsal spine did not reveal any pathology, however MRI spine showed an A-V malformation at D-8 level and the spinal angiography further confirmed the diagnosis. Patient was managed surgically and neurological rehabilitation was advised.

KEY WORDS: Paraplegia, Spinal A-V malformation, Spinal Angiography

CASE REPORT

A 28 year old female was admitted with inability to move both the lower limbs along with impaired sensation, for three hours. Initially the patient developed severe backache, which radiated down to the lower limbs. Later on, she developed weakness in both lower limbs and was unable to stand from sitting position. She also developed retention of urine simultaneously. There was no significant past medical or surgical history.

Physical examination showed a young lady of average height and built, vitals within normal limits and rest of the general physical examinations was unremarkable. Similarly, the examination of cardiovascular, respiratory and abdominal systems was normal. Regarding neurological examination, the higher mental function, speech and cranial nerves were intact and there was no abnormal finding in the upper limb examinations. In the lower limb examination, bulk was normal, tone was decreased bilaterally, power grade 0 on right side and grade 3 on the left side. Reflexes in the right lower limb were grade 4 and grade 1 on the left side. On the right side, clonus was also present. Sensory examination of lower limbs showed all sensations diminished especially on right side up to D-8 level.Cerebellar system found intact, SLR was found normal and there were no signs of meningeal irritation. Gait could not be assessed.

Investigations showed a normal blood profile. X-ray dorsal spine was unremarkable. MRI spine showed a mass within the spinal cord at D-8 level, (Fig.1) which was hypointense in both T1 and T2 images.There was evidence of expansion of spinal cord at these level (8x6mm cranio-caudal direction) and there was further evidence of multiple "flow whites" within the

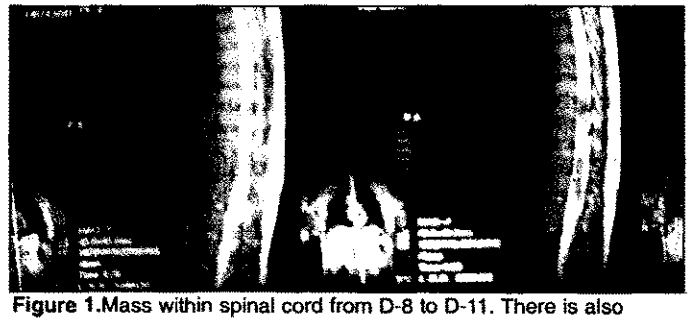

Figure 1. Mass within spinal cord from D-8 to D-11. There is also evidence of expansion of spinal cord

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intramedullary regions, extending from D-8 level to D-11 level, (Fig.2).These findings are most probably due to intramedullary A-V malformation.

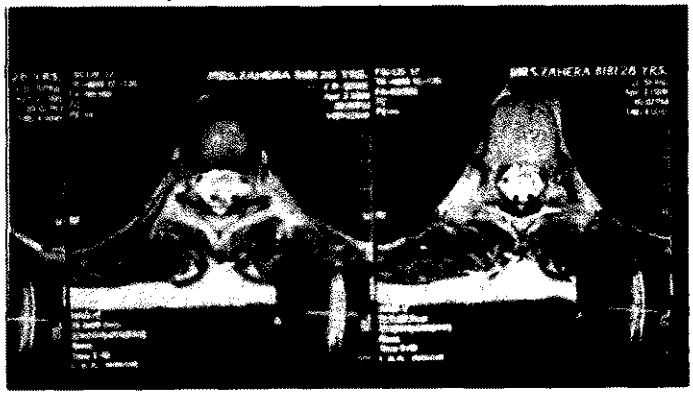

Figure 2. Mass within spinal cord. There are multiple intramedualllary "Flow Whites" suggestive of intramedualllary A-V malformation

To confirm the diagnosis, selective angiogram was performed. It showed spinal A-V malformation at the level of D-8 & D-9, which appeared to be intra-medurally glomus variety (Type II) (Fig.3).The perimedullary or pial draining veins were situated intradurally and posteriorly and extending upto D-11 washed out into the right intercostal viens.The major supply was from "artery of Adamkiewicz" originating from left 10th intercostal artery. There was a minor supply from the radicular branch of right 8th intercostal artery.

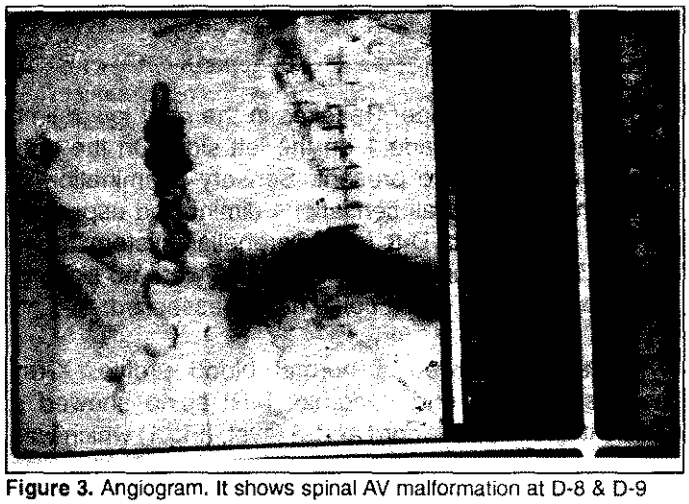

Figure 3. Angiogram. It shows spinal AV malformation at D-8 & D-9 level which is intramedullary glomus variety (Type II). Major supply is from "Artery of Admakiewic 2" originating from left 10th intercostal artery.

Her neurological rehabilitation was started and the condition began to improve. After two weeks of illness, power in the right lower limb became grade 3 (initially it was 0) and the tone increased bilaterally. Reflexes in the left lower limb became brisk (initially grade1). Ankle clonus, which was present on right side, developed on the left side as well.

The definitive treatment of AVM is surgery or embolization or both. Intramedullary AVM is very difficult to be corrected surgically. Considering the young age of the patient, surgical treatment was planned. Resection was done, but her condition did not improve any further. She was discharged for neurological rehabilitation and care.

DISCUSSION:

Arteriovenous Malformation is a direct fistula between an artery and vein without capillary beds[1]. Mostly they involve the brain but in rare cases there is spinal AVM. Although these lesions are congenital, only rarely they are familial. During the periods between 7th and 12th weeks of development, congenital AVM are mostly likely to form[2].

AVM are typically asymptomatic at birth. However, as time progresses the size and complexity of the lesion increases. Hemodynamic forces cause vessels to elongate and become tortuous. Feeding arteries dilate and occasionally develop aneurysms[3]. Draining veins also show characteristic changes as high flow and intraluminal pressure cause them to dilate and thicken[4].

The first description of spinal AVM was given in 1888 by Gaupp, who said that, spinal cord lesions are "Haemorrhoids of piamater[5]". The recognition and proper categorization of spinal AVM are crucial aspects of treatment planning. The AVM are classified in four types according to the angiographic pattern of arterial supply and venous drainage.

- *Type I: Spinal AVM [85-90%]:* They are dural and the most common type, found in lower thoracic region. This type has got a feeding artery.
- *Type II: [Glomus Variety]:* This type represents an intramedullary variety and has multiple feeding arteries.
- *Type III: or Juvenile Malformation:* This type consists of a large fistula with a rapid flow of blood with multiple feeding vessels. It is predominantly intramedullary but also has extramedullary extensions.
- *Type IV:* This type is extramedullary intradural in nature and fed by anterior spinal artery.

Apart from causing subarachnoid and intramedullary hemorrhages, it may lead to necrosis of the cord. The chronic effects of AVM leads to raised venous pressure[1] resulting in decreased perfusion and ischemia[6].

The clinical picture is either apoplectic, intermittent or progressive[7], presenting in the middle aged people with a progressive myelopathy. The myelopathy may worsen slowly or rapidly or may have periods of remission. Acute deterioration due to hemorrhage into the spinal cord or subarachnoid space may occur, but is uncommon. At presentation, most patients have sensory, motor and bladder disturbances. The motor disorders may predominate and produce a mixed picture of UMN and

LMN signs. There may be pain and intermittent claudication.

Diagnostic tools are : C.T scan, myelography[8], MRI, MR angiography[9] and selective spinal angiography[10]. Prognosis is poor[11]; 19% were disabled within six months and 50% within 3 years of onset.

Therapeutic alternatives for management of spinal AVM include surgery, endovascular embolization and a combination of both[12]. Most of the type II and type III AVM are treated by endovascular embolization. Other lesions are treated by surgery or preoperative embolization plus surgery[12]. Embolization is hazardous if anterior spinal artery or artery of Adamkiewicz is the major supply. Spinal AVM affects all age groups from neonates to elderly in different entities[13].

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RETROPERITONEAL TERATOMA

A CASE REPORT

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ABSTRACT:
A five-month old female baby presented with huge abdominal mass on right side that on investigations found to be arising from retroperitoneal area. At operation, an 18 cm x 16 cm mass was removed. Both kidneys were normal. The mass contained solid and cystic areas. Biopsy suggested benign teratoma.

KEY WORDS: Retroperitoneal teratoma, Infant

INTRODUCTION

Retroperitoneum is an uncommon site for the teratoma[1]. Most of the lesions occur in infancy[2]. In this case report we describe our experience of one such tumor.

CASE REPORT

A five-month old female baby weighing 6.5 kg presented with abdominal distension that was noticed one week earlier. Patient also developed high grade fever for which she was admitted to Medical unit. On abdominal examination a huge firm, non-tender, fixed mass was palpable on right side of abdomen. Patient was investigated and found anaemic with haemoglobin of 8.9 gm%. Biochemical investigations including urea, LFTs and bleeding profile were within normal limits. Ultrasound abdomen showed a complex mass measuring 16 x 11 x 10 cm on right side displacing the kidney and liver. CT scan abdomen revealed a 17 cm x 11 cm size mass with heterogeneously enhancing solid and cystic components on right side in retroperitoneum displacing adjacent structures. Right kidney could not be visualized. Left kidney functioned normally (Fig. I). With suspicion of neuroblastoma FNAC was done which was inconclusive.

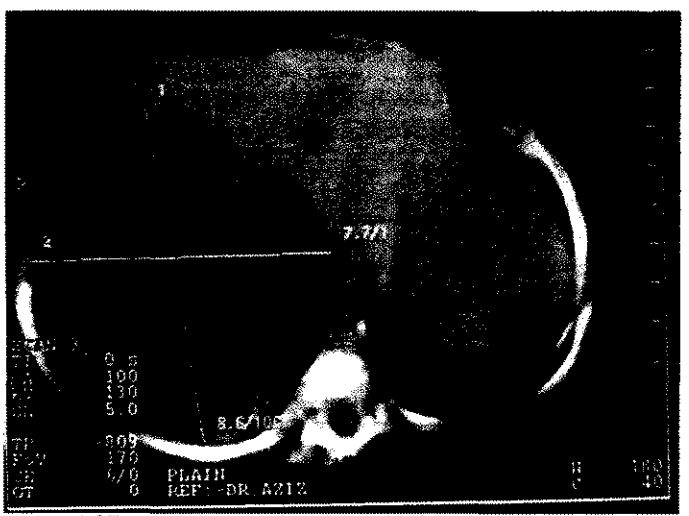

Figure I CT scan showing a large complex heterogeneous tumor

Initially patient was put on parenteral antibiotics and blood was also transfused. Following improvement in general condition patient was operated. A large retroperitoneal tumor found with flimsy blood supply from adjacent tissue pushing the right kidney anteriorly and inferiorly. Mass was excised and drain kept. Post-operative recovery was uneventful. The mass weighed 800 gm. Cut section revealed solid and cystic areas with cartilagenous tissue. Cystic areas were filled with gelatinous substance (Fig.II). On first post-operative day patient developed haematemesis and maelena. Urine was of normal color and drain contained few cc of straw colored fluid. Bleeding profile revealed deranged PT and APTT. Patient was transfused fresh blood and plasma. Her general

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Figure II Cut section of the tumor showing solid and cystic areas

condition did not improve and she died in spite of all resuscitative measures. The cause of death could not be ascertained. Biopsy showed benign teratoma.

DISCUSSION

Teratomas are tumors of germ cell origin. These cells are totipotential and can form many different types of tissue. They are initially found in the yolk sac endoderm from where they migrate to embryo and finally reach gonads. During this migration these cells may lodge in various places and later on develop into tumor. Retroperitoneum is an uncommon site for teratoma. In one of the reviews of reported cases of teratomas, only 4% (29/741) occurred in this region[3].

Girls are more frequently affected as compared to males and almost 50% manifest during infancy. Most are found on left side. They have to be differentiated from more common tumors in this age group like neuroblastoma. In our patient no calcification was noted on x-ray, which is one of the clues as to the nature of tumor. Ultrasound and CT scan can help in arriving at diagnosis by revealing solid and cystic nature of the tumor and by delineating normal anatomy of kidney[4]. FNAC failed to reveal any diagnosis in our patient

Teratomas are huge tumors with blood supply derived from all adjacent structures. In spite of this excision of tumor is usually easy as tumor is loosely attached in retroperitoneum[5]. In our patient renal pedicle and ureter were stretched over the tumor but a plane of dissection found easily. Haemostasis was easily secured and post-operative (PO) recovery was uneventful. Haematemesis with maelena in our patient on first PO day with deranged bleeding profile points to DIC the cause of which could not be ascertained. This condition can occur with malignancy (due to release of tissue thromboplastin into circulation) and septicaemia (due to endo or exo toxemia) and one of these probably is the cause of mortality in our case.

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AGENESIS OF PENIS: A RARE ANOMALY

A CASE REPORT

DAULAT KHAN

ABSTRACT:
A case of penile agenesis, with retention of urine, is reported in a three-day old baby. Scrotum was normally developed and contained testis. A small sac was found anterior to normally placed anus through which urine came out in drops from a small opening. Patient was catheterized to empty bladder but baby again developed retention following removal of catheter; so vesicostomy was performed for temporary urinary diversion. Patient was discharged and never brought for follow-up.

KEY WORDS: Agenesis, Penile agenesis, Management

INTRODUCTION

Penile agenesis is a rare anomaly of male genitalia, the management of which is a difficult issue[1]. Herein we report our experience of one such lesion with brief literature review.

CASE REPORT

A 3 day old, male baby weighing 3.0 kg, presented with non-passage of urine and absence of penis. Patient was brought from a remote area of the Lorali district in Baluchistan. Patient passed meconium within first 24 hours and was on breast feed, which he had tolerated. On examination a healthy baby with normal physical appearance except for external genitalia was found. There was no phallus and scrotum contained well-developed testis. Just anterior to normally placed anus a small sac was found on squeezing, urine in drops came out through a small pin- hole opening (Fig. I). No gross erectile tissue found. Ultrasound abdomen revealed no abnormality of urinary system. Blood urea, serum creatinine and electrolytes were within normal range. Patient was catheterized and bag was filled with adequate quantity of urine. Catheter was removed after two days but again patient went into retention. It was then decided to perform urinary diversion at this stage to avoid consequences of obstructive uropathy. Suprapubic vesicostomy was performed (Fig II). Counseling with regard to gender assignment was done but parents were not mentally prepared for the issue and requested for some time to look into this matter. The baby was never brought for follow-up.

Figure I A well developed sac lying anterior to anus containing urinary opening.

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DISCUSSION

Penis develops from genital tubercle during fourth week of intrauterine life[2]. Failure of development of genital tubercle results in penile agenesis that is reported to occur 1 in 10 to 30 million births[3]. Urethral opening can be found anywhere from inside rectum to perineum. In our patient it was found anterior to the anus opening into a sac like structure. Our patient did not have any associated anomalies that are reported in more than 50 % of the cases in literature[4].

These patients are usually managed by converting their

Figure II Supra-pubic cystostomy performed in a patient with penile agenesis

external genitalia into female type[5]. The same was offered to the parents in our case. This option is difficult to accept in primitive society like ours where male gender without phallus probably is easy to manage rather than facing a dilemma throughout the life of having sex conversion with no prospect of children in future if such a female gets married. Our patient was not brought for follow-up so it is difficult to comment what parents really would have thought.

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TREATMENT OF LONG TUBULAR DUPLICATIONS BY SUB-SEROMUSCULAR EXCISION OF THE MUCOSAL TUBE: MODIFICATION OF AN OLD TECHNIQUE

A CASE REPORT

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ABSTRACT:
An 8-month-old child at the time of operation was found to have an 85-cm long tubular duplication of small intestine, along the antimesenteric border closely adherent to the normal intestine. A sub-seromuscular mucosectomy of the duplicated bowel was performed. Whole 85 cm of the resected mucosa was removed intact, not causing even a single tear in the mucosa. The sero-muscular layers were closed over each other so that no raw area was left. The child had an uneventful recovery.

KEY WORDS: Long tubular duplications, treatment

CASE REPORT

An 8-month-old female child presented with episodes of profuse bleeding per rectum since 4 months of age. The child received symptomatic treatment only during this period and was never investigated for her symptoms. On admission, except for the extreme pallor, no other pathology could be identified. Barium follow through was inconclusive. Child was anemic with hemoglobin of 3 grams percent. She required several packed cells transfusions to correct the hemoglobin status. A provisional diagnosis of bleeding Meckel's diverticulum was made. After stabilization, exploratory laparotomy was performed. The child had a long tubular duplication of small intestine. It started about 60 cm from the duodeno-jejunal flexure and was 85 cm long. Both the normal and duplicated bowel loops were closely adherent and no clear line of cleavage was present. Proximal end of the duplicated bowel was blind and distal end was opening into the normal intestine where a 4-cm long ulcer was present in the normal intestine, which was responsible for the bleeding (Figs.1). Resection of this duplicated bowel along with normal bowel would not have been compatible with normal bowel function, therefore it was decided to preserve the normal intestine. A whole length longitudinal incision was made along the antimesenteric border of the duplicated bowel in the sero-muscular layer but sparing the mucosa. A sub sero-muscular plan of cleavage was made in the duplicated bowel and whole 85 cm of duplicated bowel mucosa was excised alongwith the ulcerated segment of normal bowel (Fig.2). Bipolar diathermy was used liberally to coagulate small blood

Figure 1. Long tubular jejuno-ileal duplication: Blind proximal end with distal end opening in to the normal intestine

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Figure 2. Mucosal tube along with the resected normal bowel having a deep ulcer

vessels in the sub-muscosal plane. Wet peanut swabs helped in the creation of a plane of cleavage. Not a single tear occurred during the dissection. An end to end anastomosis was performed between two bowel segments. The raw sero-muscular sleeve was repaired (Fig.3). The child had an uneventful recovery. She was discharged after five days of hospitalization. At one-year follow-up she remained perfectly well and symptoms free.

DISCUSSION

Small intestinal duplications are not uncommon in children. The duplications may be cystic or tubular.[1] Cystic duplications often present as a mobile abdominal mass, intermittent abdominal pain and acute intestinal obstruction by twisting and volvulus[2,3,4]. Tubular duplications however may remain silent for a long time. They often present with intermittent or profuse bleeing per-rectum[4]. The patients may become severely anemic if not treated in time. Initially a lot of time is wasted in investigating these children with endoscopies for polyps, radioisotope scan and contrast studies. Often no cause is found for the severe anemia and bleeding per rectum.

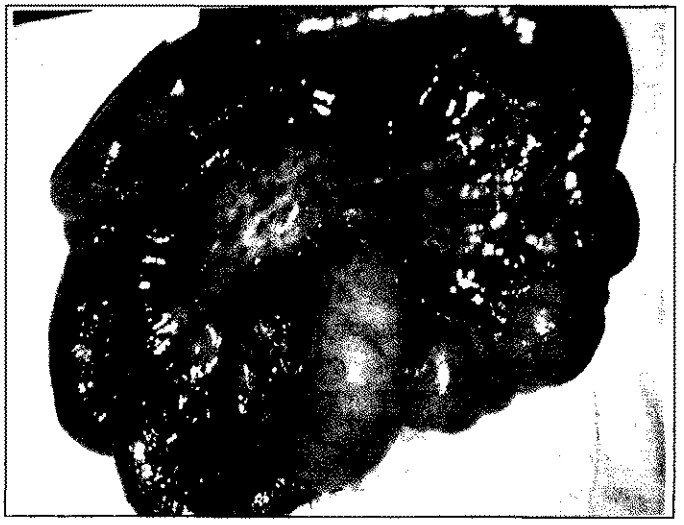

Figure 3. Result after excision of duplicated bowel mucosa and ulcerated area of normal intestine and end-to-end anastomosis

Most cases of small intestinal duplications are found at operations for other diagnosis[5,6]. The cause of bleeding in these patients is ulceration of the duplicated or normal intestine by the acid produced by ectopic gastric mucosa[7].

The duplications occur mostly along the mesenteric border of the intestine and therefore share a common blood supply with the normal intestine[8]. A plane of cleavage between the normal and duplicated bowel may be present, making resection easy[8,9]. However often both walls are closely adherent and resection is not possible without damaging the blood supply to the normal intestine. There are several ways of tackling this problem. Internal drainage of the duplication into the normal bowel has been successfully done in few cases[10,11] , however the high incidence of heterotopic mucosa negates this procedure[4]. If a plane of cleavage can be found between the duplicated and normal intestine then resection of the duplication can be performed without jeopardizing the blood supply to the normal intestine[12,13,14]. Norris has used the Bianchi principle for excision of duplications, which share a common blood supply, with good results.[15] However often duplication and the native bowel share a common wall. In these cases the treatment has been by series of multiple incisions in the duplicated bowel and mucosal stripping[1]. In our patient the same principle had been used but in a different way. Instead of making several incisions, we made a long incision along the whole length of the duplicated bowel because duplicated segment was found on antimesentric border, which is an unusual finding.

To summarise, it is possible to treat long tubular duplications present on antimescenteric border by excising the mucosa in sub sero-muscular plane. This not only preserves the vascularity of normal intestine, but also allows a clean per-operative field with minimal chance of post-operative complications.

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