VOL. 1 NO. 1 APRIL - JUNE 1996 QUARTERLY

JSP JOURNAL OF SURGERY PAKISTAN

Published by: Dr.Abdul Aziz for Prof.F.U.Baqai, Baqai Postgraduate Medical Institute, IIIC, 1/12, Nazimabad, Karachi
Address for correspondence: Col. (Retd.) Masudul Haque, Executive Secretary, Surgical Publications, Baqai Postgraduate Medical Institute, IIIC, 1/12, Nazimabad, karachi, Tel & Fax:5887333
Subscribeption rates: Per copy-in Pakistan Rs.100/= in SAARC countries U.S. $20, in other countries U.S.$30, Annual- in Pakistan Rs.300/= in SARRC countries US $ 60, in other countries U.S.$90.
Layout by : Aleemuddin Siddqiui.

EDITOR'S NOTE

ORIGINAL ARTICLES

REVIEW ARTICLE

CASE REPORTS

E D I T O R ' S N O T E

Remarkable changes have taken place in the health delivery systems in the 20th Century and we expect to live much longer and healthier than our grand parents. Yet despite this progress, scores of health problems remain to be solved. Efforts are being made all over the world to find answers to these unsolved problems by carrying out research for the benefit of the ailing millions. The fruits of this research and experiences are being shared by researchers through medical journals and publications. A large number of journal are published all over the world and scores of new ones are added each year, yet no journal exists to share the wide experiences of researchers in the SAARC region, which housed almost one quarter of the world population with common medical problems and similar diseases and disease patterns.

An ocean of scientific and clinical material exists in the SAARC region and the Journal of Surgery Pakistan is our attempt to tap this information. It has a wider scope as the SAARC Association of Surgeons (Pakistan) has been joined by International College of Surgeons (Pakistan Chapter) in this Venture. The purpose of this publication is to bring together all the members of theses two societies on a common platform. As this journal is to be distributed to all surgeons and Heads of Surgery in Pakistan, internationally through International College of Surgeons (Pakistan), as well as the SAARC countries, it will serve to bring the surgeons of these societies and region closer and will encourage exchange of knowledge and useful interaction.

This first issue which is in your hands, contains articles on topics of great importance by Pakistani surgeons and we hope to have the benefit of the experiences and studies of surgeons in SAARC countries in our future issues. We also plan to introduce a "Letters to the Editor" portion where surgeons will be encouraged to express views on similar studies and give their comments and observations on published articles and reviews.

We may add that a journal of this nature cannot be published and distributed without the active support and collaboration of the pharmaceutical and other Industry business houses. It could provide a great opportunity to the pharmaceutical Industry to advertise their firm, services and products, besides promoting the cause of surgical profession. We would therefore welcome advertisements for our future issues.

We are grateful to the contributors to this first issue and the firms that advertised in it.

Editor

A 10 YEARS STUDY OF LYMPHANGIOMATOUS MALFORMAITON AT N.I.C.H.

JAMSHED AKHTAR, FARHAT MIRZA, SOOFIA AHMED, ASADULLAH KHAN, ABDUL AZIZ.

ABSTRACT

Between 1st January, 1984 and 31st December 1993, 126 cases of the Lymphangiomatous malformation were managed at National Institute of Child Health, Karachi. Most of the patients were under 6 months of age. Male domination was noted. Six neonates presented in emergency with respiratory distress. Cervico-facial region was involved in 61.9% of the cases. Surgery was treatment of choice. Over all results were satisfactory. There were only two deaths in this series which were in non operated cases.

KEY WORDS: Cystic hygroma, Lymphangioma.

INTRODUCTION

Lymphangiomatous malformation is the congenital maldevelopment of the lymphatic system. It may take the shape of cysts of variable sizes when it is known as cystic hygroma. (Fig 1) The capillary and cavernous types of lesions are termed as lymphangiomas. Many modes of treatment have been described in literature with variable results. We present our experience of this malformation over a period of ten years.

Figure 1 Massive Cystic Hygroma of Head and Neck Region

MATERIALS AND METHODS

Department of Pediatric Surgery at NICH provides services not only to people of Karachi but also to the rest of Sindh and lower Baluchistan. Few cases are referred from up country as well. 120 cases were admitted as elective cases and 6 cases were admitted in emergency due to variable degree of respiratory distress. Elective cases underwent routine investigations like complete blood count and X-ray chest, where indicated. Other investigations were done according to the site of involvement. When judged fit enough they were given appointment for surgery. No case was operated in emergency. In majority of the cases total excision was carried out. Conservative surgery was done in 10 cases of huge cystic hygromas of the face and neck region with infiltration of the deeper structures. Staged procedures were carried out in 4 cases-2 involved tongue and 2 involved trunk. Complications noted in post-operative period included infection, leakage of the lymph and recurrence. Two deaths were recorded in our series.

RESULTS

126 cases were studied during 10 years from 1984 to 1993. There were 78 (61.9%) male and 48 female (38.1%) patients. Age distribution is given in Table-I. Most of the cases were under six months of age. In the regional distribution cervico-facial region was the most common part involved. Table II. Total excision was performed in 110 cases (88.9%), partial excision was done in 10 cases (8.0%) and staged procedure was performed in 4 cases, (3.1%). Post-operative complications are given in Table III.

Dr. Abdul Aziz,
Dept. of Paedriatic Surgery,
National Institute of Child Health, Karachi

Table I

Age Distribution

Age Distribution	No. of cases	Percentage
Day 1 to 6 Months	42	33.3%
6 Months to 12 Months	14	11.2%
1 Year to 3 Years.	52	41.2%
5 Years and above	18	14.3%

Table II

Regional Distribution

Regional Distribution	No. of cases	Percentage
Cervical Region	68	53.9%
Face	10	8.0%
Tongue	2	1.6%
Chest Wall	12	9.6%
Abdominal Wall	4	3.2%
Upper Limb (including Axilla)	16	12.5%
Lower Limb	10	8.0%
Perineum	4	3.2%

Table III

Postoperative Complications

Early	No of cases	Percentage
Respiratory Distress	4	3.2%
Wound Infection	4	3.2%
Chest complications	6	4.8%
Transient Facial nerve paresis	2	1.6%
LATE		
Contracture	2	1.6%
Facial nerve Paralysis	2	1.6%
Recurrence	8	6.4%

DISCUSSION

Cystic hygroma and lymphangioma are benign, soft compressible malformations of the lymphatic system. Head and neck region is the most common site of involvement[1]. These lesions usually appear after birth but upto 40% can be seen in new born[2]. Cullen et al have reported a series of 30 cases of cervical cystic hygroma diagnosed in the first trimester of pregnancy. In the modern era of antenatal diagnosis with ultrasonography, it is now possible to arrange for appropriate means to save the life in case of huge infiltrating cystic hygroma at the time of birth[3,4]. This aspect of management is still lacking in under-developed countries where lack of co-ordination among various specialities leads to high morbidity and mortality.

The number of cases reported from under-developed countries is more than from developed countries. Ricciardelli et al has presented a series of 34 cases in 10 years[5]. In 10 years period we have studied 126 cases and a similar number of cases have been presented by Ninh and Ninh[6]. The reason for this difference is population which is much more in under-developed countries like ours with large drainage area and because of paucity of health services. Another point to be emphasized is the use of diagnostic techniques like ante-natal ultrasonographic detection of these cases. Complications like miscarriage in first trimester have been noted by Turnpenny et al in fetuses affected by this malformation[7]. Longo et al has described 5/1000 incidence of abortion with cystic hygroma[8]. Abortions on medical grounds with these malformations could be another reason for the lesser number of cases from western countries[9]. Shulman et al have emphasized an increased risk of fetal chromosomal abnormalities especially autosomal Trisomy[10]. Tanaka et al have reported more than 20% mortality in post-natal period following deliveries in such cases[11]. He advised cesarean section and in utero endo-tracheal intubation of the neonate to save life. Tricoire et al have reported 8 cases of familial cystic hygroma in 3 families[12]. The cystic hygroma has also been noted in the siblings.

Miyabara et al have mentioned in their study about the presence of cystic hygroma in twins with conjoined pregnancy[13]. Cystic hygroma has been associated with certain other abnormalities. Droste et al have pointed out its association with Turner syndrome[14]. Achiron et al have described its association with congenital heart defects[15]. Macleod et al have associated this condition with hydrops fetalis[16]. Bulas et al have reported a case of Fryn's syndrome presenting with cystic hygroma[17]. Gorenstein et al

have described the association of venous aneurysm with cystic hygroma[18].

Rare sites for cystic hygroma have been mentioned in the literature and have been noted by us as well, for example mediastinal cystic hygroma, a rare lesion reported by Muraskas et al[19]. Their lesion was in the anterior mediastinum causing airway obstruction, mediastinal shift and hypoplasia of the lung. We have noted lesion in maxillary part of the cheek with extension into retro-orbital and anterior intracranial fossa. Other rare sites mentioned in literature include parotid region[20] and bone[21] etc.

The diagnosis is usually made on history and physical examination. Ultrasonography, C.T. scanning, M.R.I. and lymphangiography are useful in delineating the extent of the lesion. Various modes of treatment have been described in addition to surgery. Dickerhoff et al have mentioned the use of chemotherapy, like cyclophosphamide in non-resectable cystic hygromas[22]. Bleomycin and OK-432 have been used by some[23,24]. Recurrence is a common problem especially in massive lesions. Scally has reported recurrence in adults[25]. In our study recurrence was noted in the case of tongue and trunk and in two cases involving cervico-facial region.

In conclusion, lymphangiomatous malformations are not infrequent. If facilities are available and a cystic hygroma is suspected in the ante-natal period, even if of small size, detailed ultrasonographic examination and karyotyping are recommended because of its frequent association with chromosomal defects.

Surgery, either total excision, or staged excision is the treatment recommended. Recurrence is known in massive lesions.

REFERENCES

1. Singh, S., Baboo, ML, Pathak IC.: Cystic lymphangioma in children. Report of 32 cases including lesions at rare sites. Surg. 1971: 69, 947-951.

2. Thompson DM., Kasperbauer JL. Congenital cystic hygroma involving larynx, presenting as an airway emergency. J Natl Med Assoc. 1994: 86, 629-32.

3. Langer JC., Fitzgerald PG., Desa D., et al. Cervical cystic hygroma in the fetus: Clinical spectrum and outcome. J Pediatr Surg. 1990: 21, 58-61.

4. Cullen MT., Gabrielli S., Green JJ. Diagnosis and significance of cystic hygroma in the first trimester. Prenat Diagn. 1990: 10, 643-51.

5. Riccardelli EJ., Richardson MA. Cervicofacial cystic hygroma: patterns of recurrence and management of difficult case. Arch Otolaryngol Head neck surg. 1991: 117, 546-53.

6. Ninh TN., Ninh TX. Cystic hygroma in children. A report of 1236 cases. J Pediatr Surg.:1974,:9, 191-95.

7. Turnpenny PD., Gonasegaran R., Smith CN., et al. Recurrent miscarriage, cystic hygroma and incontinentia. Br J Obstet Gynecol. 1992: 99, 920-1.

8. Longo M., Pecoraro G., Zarcone R., et al. Cystic hygroma : 2 case reports and review of literature. Minerva Ginecol.(Italian). 1994: 46, 285 93.

9. Christodoulou CN., Kotoulas IG., Agapitos EB. et al. Intrafetal prostaglandin F 2 Alpha administration for mid trimester pregnancy termination. A case report. Am J Obstet Gynecol. 1990: 162, 1245-6.

10. Shulman LP., Raafat NA., Mace PC. et al. Significance of septation in isolated fetal cystic hygroma detected in the first trimester. Prenat Diagn. 1994: 14, 223-6.

11. Tanaka M., Sato S., Naito H., et al. Anaesthesia management of a neonate with prenatally diagnosed cervical tumors and upper air way obstruction. Can J Anaesth. 1994: 41, 236-40.

12. Tricoire J., Sarraman MF., Rolland M. et al. Familial cystic hygroma. Report of 8 cases in 3 families. Genet Couns. 1993: 4, 265-9.

13. Miyabara S., Suzomori K., Uemura Y. et al. Fetal cystic hygroma in sibs: developmental analysis of co-existent cardiovascular malformation relevant to pathogenesis. Birth Defects. 1993: 29, 303-15.

14. Droste S., Hendricks SK., Von Alfrey H., et al. Cystic hygroma colli: perinatal outcome after pre-natal diagnosis. J Perinat Med. 1991: 19, 449-51.

15. Achiron R., Rotstein Z., Lipitz S., et al. First trimester diagnosis of fetal congenital heart dis-

ease by transvaginal ultrasonography. Obstet Gynecol. 1994; 84, 69-72.

16. Macleod AM., Mchugo JM. Prenatal diagnosis of nuchal cystic hygroma. Br J Radiol. 1991, 64 (765); 802-7.

17. Bulas HI., Saal HM., Allen JF., et al. Cystic hygroma and congenital diaphragmatic hernia; early prenatal sonographic evaluation of Fryn's syndrome. Prenat Diagn. 1992: 12, 867-75.

18. Gorenstein A., Katz S., Rein A., et al. Giant cystic hygroma associated with venous aneurysm J Pediatr Surg. 1992: 27, 1504-6.

I9. Muraskas JK., Gianopoulos JG., Husain A., et al. Mediastinal cystic hygroma, prenatal decompression with neonatal resection and recurrence at 19 month of age. J Perinatol. I993: 13, 381-4.

20. Goshen S., Ophir D. Cystic hygroma of the parotid gland. J Laryngol Otol. 1993: 107, 855-857.

21. Law PJ., Hall CM. The clavicular overgrowth in association with cystic hygroma. Skeletal Radiol. 1991: 20, 597-9.

22. Dickerhoff R., Bode VU. Cyclophosphamide in non resectable cystic hygroma. Lancet. 1990: 335, 1474-75.

23. Tanigawa N., Shimomatsuya T., Takahashin K., et al. Treatment of cystic hygroma and lymphangioma with the use of bleomycin fat emulsion. Cancer. 1987: 60, 741-9.

24. Ogita S., Tsuto T., Deguchi E., OK 432 therapy for unresectable lymphangioma in children. J Pediatr Surg. 1991: 26, 263 70.

25. Scally CM., Black JH. Cystic hygroma, massive recurrence in adult life. J Laryngol Otol. 1990: 104, 908-10.

PATTERN OF MALIGNANCY IN CHILDREN

JAMSHED AKHTAR & ABDUL AZIZ

ABSTRACT

Over a period of five years eighty one tumors were seen with varied presentation. No significant difference was observed as regard to sex distribution. More than 20% of tumors were observed in patients under one year of age. Wilm's tumor was the most common tumor in this study. Late presentation and high mortality were the significant features. We stress on coordinated efforts to collect relevant epidemiological data so that future planning could be made in order to reduce the morbidity and mortality.

KEY WORDS: Tumor Registry, childhood malignancy.

INTRODUCTION

Incidence of cancer is derived from population based statistics collected by tumor registries. In our country and infact in our part of the world such comprehensive data does not exist because of the non-existence of such organizations. Prevalence and incidence rates are frequently used in epidemiological studies as the basis for planning and evaluating health programs. As we do not know prevalence and incidence of tumors in our population, disease pattern can not be evaluated and no future projection can be made. It is for this reason we conducted a study in patients on our surgical service to look at the statistics and to evaluate our results and to make future recommendations in this regard.

MATERIALS AND METHODS

A prospective epidemiological study was planned and conducted between 1st. July 1989 and 30th. June 1994 in one of the two units of Paediatric Surgery at National Institute Of Child Health, Karachi to collect the relevant details on childhood malignancies upto the age of 12 years on a structured proforma. The subjects included were all those who attended our surgical service. Patients sent to us for lymph node biopsies following diagnosis of lymphoma on the basis of fine needle aspiration cytology were not included as they were managed in medical unit. Our department does not deal with neurosurgical or ophthalmologic malignancies so these cases were excluded from the study. A total of 81 cases were seen during this period with tumors involving almost every part of the body.

RESULTS

During five years study period no significant difference was seen as regards to the yearly distribution and sex involvement. There were 40 male and 42 female patients. More than 20% of tumors were seen in infants. (Table I). Wilm's tumor was the most common tumor in this study, (Table II). Follow up of four most common tumors is given in table.III.

Table-1
Age Distribution

upto 1 year	19
Between 1-5 years	25
Above 5 years	37

Table II
Types of Tumours

Tumour	Number
Nephroblastoma	12
Neuroblastoma	6
Ewing's sarcoma	6
Rhabdomyosarcoma	4
Sacrococcygeal Teratoma	9 (Malignant - 1)
Cervical Teratoma	4

Dept. of Paediatric Surgery, NICH, Karachi

Ovarian Tumor	9 (Malignant - 2)
Testicular Tumor	3
Lymphoma (Abdominal)	3
Adrenal Carcinoma	2
Squamous Cell Carcinoma	2
Ganglioneuroblastoma	1
Ganglioneuroma	1
Chondrosarcoma	1
Osteosarcoma	1
Osteoclastoma	1
Fibroliposarcoma	1
Hepatoblastoma	1
Adenolymphoma (parotid gland)	1
Others	13
Total	81

Table III
Followup chart

Tumor	NO.	On chemotherapy	On Follow up	Died	Lost to follow up
Wim's tumor	12	3	1	3	5
Neuroblastoma	6	1	-	4	1
Ewing's sarcoma	6	2	-	1	3
Rhabdomyosarcoma	4	2	2	-	-

DISCUSSION

In developed countries cancer is second only to trauma as the leading cause of death in children[1].According to the western studies incidence of major malignant neoplasm is 124.5 / 1,000,000 in Caucasian children under 15 years of age[2]. No such statistics are available for Pakistan. Sporadic reports from various centers can be found in different periodicals but coordinated studies are lacking. Zaidi and Jafery[3] collected 213 malignant tumors over a period of six years from southern part of Pakistan. Almost 50% of tumors were lymphomas, leukemias and retinoblastoma. Renal and bone tumors comprised 38 cases (18%). It has been suggested that as Wilm's tumor occurs with the least geographical variation it should be used as standard of comparison. In another study from Armed Forces Institute of Pathology, Rawalpindi, 840 malignant tumors in children under fifteen years of age were collected in twelve years from northern areas of Pakistan[4]. More than 2/3rd of the cases were leukemias, lymphomas, C.N.S. and eye tumors. Renal tumors comprised almost 5% of the total cases followed by soft tissue sarcomas. In a study from Pakistan Institute Of Medical Sciences Islamabad Wilm's tumor comprised l0.3% (49) of the 475 malignant tumors collected in 6.5 years[5]. In the above mentioned series significant male predominance was seen which is not borne out by our study. In our study a large number of tumors were seen in infants under one year of age which highlights the importance of their inclusion in differential diagnosis of masses. The most common tumor in our study was Wilm's tumor followed by bone tumors, neuroblastoma and soft tissue sarcomas. This order is also seen in other studies. We came across different tumors with varied presentations like Osteosarcoma involving proximal phalanx of finger, Liposarcoma involving clavicle and Chondrosarcoma in supra clavicular region. These sites are rarely involved and are rarely reported in literature[6,7]. Cervical teratoma, a rare lesion of which only 212 cases are reported in about 130 years[8]. We managed four such cases in a short span of time. An important observation made in this study was late presentation. Most of the tumors were either in stage III or IV. Looking at the follow up, a large number of patients either died or lost to follow up and are presumed dead. The treatment plan involves multidisciplinary programs that include surgery, chemotherapy, radiation and to limited extent immunotherapy. Chemotherapy has dramatically improved the prognosis of many paediatric neoplasma particularly Wilm's tumor and rhabdomyosarcoma[9]. These drugs are costly and treatment is also prolonged so, drop out rate is high and so is the mortality. Help from social workers and voluntary organizations in this regard is to be appreciated.

In conclusion, a paediatric oncology group should be established to compile the important statistics. The data collected will be of help in future planning including research. At present more funds should be generated both for diagnosis and treatment of childhood malignancies.

REFERENCES

1. Morgan ER, Baum ES. Tumors. in Raffensperger JG (ed.) Swenson's Paediatric Surgery, 5th. edition, section V, Appleton & Lange publishers. 1990: 345-346.

2. Young JL, Miller RW Incidence of malignant tumors in US. children. J Pediatr. 1975: 8, 254.

3. Zaidi SHM, Jafery NA. Childhood tumors in Karachi.JPMA. 1977: 27, 346-349. .

4. Ahmed M, Khan AH Mansoor A. The pattern of malignant tumors in northern Pakistan. JPMA. 1991: 41, 270-273.

5. Islam SS, Bukhshee MJ, Islam S, et al. Present status of Wilm's tumor at the children's hospital, PIMS. Pak J Pediatr Surg. 1994: 1, 60-70.

6. Price CGH, Zhuber K, Kuntschik MS, et al. Osteosarcoma in children. A study of 125 cases. J Bone Joint S urg. 1975: 57-B, 341-345.

7. Resnick D, Tumors and tumor like diseases in Resnick D, Niwayama G. (ed). Diagnosis of bone and joint disorders. 2nd. ed., vol. 6, chap. 91. WB Saunders Co. 1988: 3602-4011.

8. Jordan R, Gauderer M. Cervical teratoma: An analysis, literature review and proposed classification. J Pediatr Surg. 1988: 23, 583-591.

9. D'Angio GJ, Evans A, Breslow N, et al. The treatment of wilm's tumor: Results of second National wilm's tumor study. Cancer. 1981: 47, 2302.

NORMAL APPENDICES IN PATIENTS OPERATED FOR SUSPECTED ACUTE APPENDICITIS

SHAISTA KHAN, FARHAT ABBAS AND HIZBULLAH SHAIKH

Abstract

The morbidity and mortality associated with delays in diagnosis dictates that appendectomy be done early on strong clinical suspicion. No single laboratory or imaging technique has gained specificity for acute appendicitis. In the age group 15-25 years, specially in women, other clinical diseases may mimick appendicitis to such an extent that the number of appendectomies may rise to an un-acceptable level. In order to assess our "normal" appendices in appendectomies rates, we reviewed all the histology specimens of appendices removed between January 1986 - January 1987. Amongst 60 appendectomies performed during this period, 54 (90%) were for suspected acute appendicitis, where as 6 (10%) were incidental appendectomies. Histologically 14 (25%) appendectomies suspected for acute appendicitis were reported as normal. This is well within range of normal appendices removed, as reported in literature. 12 (85.7%) of these 14 patients were females, it emerges, therefore, that in young females a more intense effort to diagnose acute appendicitis preoperatively must be made. The use of laparoscopy could be exploited further.

Key words: Appendicectomy, Normal appendix.

INTRODUCTION

Acute appendicitis can be a difficult diagnostic problem. Frequently appendices removed on clinical suspicion are reported histologically as normal[1]. The morbidity and mortality associated with delays in diagnosis especially, at extremes of age, dictates that appendicectomy be done early on strong clinical suspicion. However, specially in women in the 15-25 years age group, other clinical syndromes mimic appendicitis[2,3] to such an extent that the incidence of unnecessary appendicectomy may rise to unacceptable levels. For these two reasons it is necessary that every effort be made to accurately diagnose acute appendicitis before surgery.

It is considered acceptable to remove 5-25%[4] of normal appendices as a safeguard against underdiagnosing a potentially lethal condition. An increase over this range may be interpreted as a sign of unnecessary explorations and review by individual hospitals can serve as a basis of quality assurance in health care.

Dr. Shaista Khan, Dept. of Surgery,
Aga Khan University Hospital, Karachi

In order to assess our 'normal appendix' appendicectomy rates, we undertook the study reviewing all appendectomies done in a 13 months period, from January 1986 to January 1987.

PATIENTS AND METHODS

The case notes of all patients who underwent appendicectomy between January 1986 - January 1987, were retrospectively reviewed. The case notes of patients with histopathologically normal appendix and in whom appendicectomy was not incidental to another operation, were scrutinised. Age, sex, the clinical parameters used for diagnosis (including duration of symptoms, nature and location of pain, abdominal tenderness and associated complaints like nausea, vomiting and pyrexia) complete blood count, urinalysis and the final diagnosis were noted.

RESULTS

Between January 1986 - January 1987, a total of 1784 patients were admitted to the department of Surgery (Table I). 1663 (93.2%) required surgical procedures. Of these 60, 3.3%, (30 males and 30 females) procedures were for removal of appendix.

Six patients (10%) required incidental appendicectomy and were excluded from the study, leaving 54 patients operated for suspected appendicitis. The age distribution was 3.5 to 61 years with mean age being 25 years. Fourteen patient (26%) were found to have histologically normal appendix. Twelve (85.7%) of fourteen patients were females. Their ages ranged from 15-25 years. There was no appreciable difference in the symptomatology between groups of patients with inflamed and normal appendix. Similarly, localized pain in the right lower quadrant with elicitable rebound tenderness was noted equally in both groups (Table II). Leucocytosis was noted to be a poor index in differentiating acute appendicitis from non inflamed appendix, similar to experience by other groups[5]. 86% of the latter revealing leucocytosis in excess of 10,000/cmm. Two of the 14 patients (14.2%) developed minor complications including one wound infection. Another patient with underlying Crohns disease later developed sub-acute intestinal obstruction felt to be related to underlying Crohns disease. Appendicectomy in this patient was carried out with uneventful recovery with no residual enterocutaneous fistula. There were no deaths. Among 14 patients, eleven (8.5%) patients revealed underlying diseases adequately explaining the cause of abdominal pain (Table III). The spectrum of disease in our patients was not too dissimilar to the experience in the West[4] (Table IV).

TABLE I

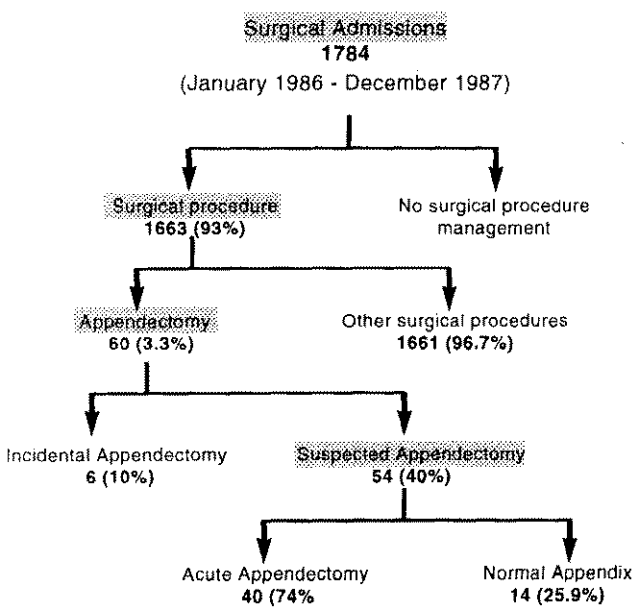


TABLE II
COMPARISON BETWEEN CLINICAL PRESENTATION OF NORMAL AND ACUTE APPENDICITIS

	ACUTE (40)	NORMAL (14)
PAIN		
Typical	26 (65%)	9 (64%)
RIF	8 (20%)	2 (14%)
Atypical	4 (10%)	3 (21%)
NAUSEA/VOMITING		
Nausea	7 (17%)	2 (14%)
Vomiting	27 (67%	7 (50%)
Nil	6 (15%)	5 (36%)
FEVER		
Above 39 C	5 (13%)	3 (21%)
Below 39 C	22 (55%)	5 (36%)
Normal Temp.	12 (30%)	6 (43%)
LOCAL TENDERNESS/REBOUND		
Tenderness	11 (28%)	2 (14%)
Tenderness & Rebound	28 (70%)	12 (86%)

TABLE III
PATHOLOGY OF 14 CASES WITH NORMAL APPENDIX AT THE AGA KHAN UNIVERSITY HOSPITAL

CECAL DIVERTICULUM	1
CROHN'S DISEASE	2
INFARCTED OMENTUM	1
PRIMARY PERITONITIS	2
SALPINGITIS	2
MESENTERIC ADENITIS	1
CORPUS LUTEUM RETENTION CYST	1
T.B. GREATER TROCHANTER	1
DIAGNOSIS NOT KNOWN	1
	14

TABLE IV
DISEASES MIMICKING ACUTE APPENDICITIS *

DISEASE	NO. OF PATIENTS
Diagnosis at operation	**32**
Surgery required	**13**
Meckel's diverticulitis	3
Perforated cecal diverticulitis	1
Perforated cecum by foreign body	1
Adenocarcinoma of appendix	1
Leiomyosarcoma of duodenum	1
Acute cholecystitis	1
Torsion of gallbladder	1
Torsion of omentum	1

Torsion of right ovarian cyst	2
Ruptured ectopic pregnancy	1
Surgery not required	**19**
Nonspecific mesenteric lymphadenitis	9
Bleeding follicular or luteal ovarian cysts	5
Pelvic inflammatory disease2	
Endometriosis	1
Tuberculous ileitis	1
Yersinia pseudotuberculosis	1
Diagnosis after surgery	**10**
By investigation	**3**
Salmonella infection	3
Gallstones	1
Acute pancreatitis	1
Urinary tract infection	1
Benign ovarian teratoma	1
Infectious monoculeosis	1
By second laparotomy	**1**
Ischemic colitis	1

* From Lau et al - The American Journal of Surgery, Volume 148 September 1984 (reproduced by permission).

It emerges, therefore, that young women in the age group of 15-25 stand a higher risk of misdiagnosis and hence require more intensive investigation and observation before exploration. In this cohort of patients, laparoscopy has proved helpful in visualizing 25% of normal appendices.

DISCUSSION

Debate has occurred for many years on the acceptable rate of unnecessary appendicectomy resulting from inaccurate diagnosis in acute abdomen. The major concerns of a surgeon managing a patient with acute abdominal pain are the risks associated with a negative exploration versus the hazards of conservative management, which may allow an appendix to perforate. Most studies report a low complication rate following unnecessary appendicectomy. The complications are usually minor: wound infections and haematomas. Additional financial losses, due to absence from work in patients with continuing symptoms, non appendicular in origin, remains a problem. However, Lau et al in 1984[4] reported an overall complication rate of 14%. On the other hand surgery following perforation of an inflamed appendix, continues to carry significant morbidity and mortality[2]. In this context the acceptable rate would vary with the hospital setting. In a district hospital with limited diagnostic services, the practice would be towards early exploration to avoid perforation of the viscus. Poor patient compliance may also influence the decision to manage conservatively. On the contrary, a well equipped tertiary care centre should allow a more accurate preoperative diagnosis and hence reduce the number of negative surgical exploration. In the former setting an acceptable rate may exceed 30%, whereas the latter should restrict the number to 20% or less. A small percentage of this group may require further investigations and hospitalization. Conversely, delayed exploration culminating in perforation of the organ, increases the morbidity and costs due to prolonged hospitalization. Charles McBurney's classical paper in 1899 quoted in[6], emphasizing the need for early appendicectomy to avoid perforation with its ensuing complications probably resulted in a spate of appendicectomies and instigated Harte[7] to state that 'the perfect man is the man without an appendix'. A review by Richardson[8] revealed acute inflammation in only 58% of the patients following appendicectomy. In a survey conducted in different hospitals in United States of America[9] between the years 1924 to 1963, of the 64,591 surgically removed appendices, 35,223 (54.5%) were normal and were labelled by pathologist as 'prophylactically removed' or 'appendicectomy for chronic appendicitis'. In a comparative study conducted in Northern Europe[10] the incidence of normal appendices was 40-50%, whereas, a similar study from Britain[11,12] reported 20% rate. This wide discrepancy is probably attributable to different attitudes towards surgical intervention and perhaps also due to availability of medical services in different countries. Recent surveys[4] have tended to give more conservative figures with a 5 to 27% incidence of normal appendices.

Looking at the various causes of abdominal pain suggestive of or mimicking acute appendicitis, a review of the cases at the Age Khan University Hospital reveals a similarity of experience to that in the West. The percentage of normal appendicectomies in our service (25%) is in the higher reaches of the acceptable range[13], but has effected a diagnostic approach to reduce this figure. The underlying diseases encountered in our experience display close similarity to those in other series (Table IV). As reported by others, in a small number of patients in our series, the final diagnosis remained unclear following exploration.

In these patients if symptoms persists, following surgery, further investigations including serological tests, abdominal ultrasound and radiological studies may be indicated. If these are inconclusive, a regular followup for several months may uncover an underlying disease process. However, measures to reduce the incidence of negative appendicectomies without risking an increased incidence of perforation should be evaluated further. The advantages of laparoscopy

in the last decade has emerged and perhaps needs to be exploited further, utilizing it, in order to arrive at an accurate diagnosis before surgery. Hospitals in Pakistan should identify the rates of histologically normal appendices, removed on clinical suspicion of acute appendicitis. This exercise would also bring about an awareness and improve the clinical acumen of junior staff, the group generally involved in first line management of these patients.

REFERENCES

1. Chang F.C., Hogle H.H. Welling R.D.: The fate of the negative appendix. American Journal of Surgery. 1973 126, 752-54.

2. Perr jess, B.Bjerregaard et al. Acute Appendicitis.: Prospective trial concerning diagnostic accuracy and complication. American Journal of Surgery. 1981 141, 232-34.

3. Silbermanv: Appendicectomy in a large metropolitan hospital. Retrospective analysis of 1013 cases. American Journal of Surgery 1981 142 No.8, 615-18.

4. Lau WL, Fans, Wong SH.: Negative findings at appendectomy American Journal of Surgery. 1984 148, 375-78.

5. Bomer RJ, Bell MJ, Teruberg FL.: Diagnostic value of the white blood count and Neutrophil percentage in the evaluation of abdominal pain in children. Surgery, Gynaecology and Obstetrics 1981 152, 424-26.

6. Harte RH.: In discussion of Richardson MH. Trans American Surgery Association. 17:22 1989.

7. Mcburney C.: Experience with early operative interference in cases of diseases of vermiform appendix. NY. Med J. Dec 21, 1889, 676.

8. Richardson MH.: Appendicitis. Trans American Surgery Association 17:22 1899.

9. Collins DC, 71,000.: Human appendix specimens. A final report, summarizing Forty years study. The American Journal of Proctology, 1963, 14 (6), 365-81.

10. Neutra R.: Indication for the Surgical treatment of suspected acute appendicitis: a cost effectiveness approach. In: Bunker JP Bariness BA, Mosteller F. eds. Costs, risks and benefits of Surgery New York - Oxford University Press 1977. 277-307.

11. de Dombal FT, leaper. DJ. Stamiland JR, McCann AP, Horrocks JC: Computer aided diagnosis of acute abdominal pain - BMJ. 1972 2, 9-13.

12. Gilmore OJA, Brodrib AJM, Bronett JP et al.: Appendicitis and mimicking conditions - Lancet 1975 2, 421-4.

13. Alexander A. Dentsch, Naphtali Shami and Raphael Reiss. Are some appendicectomies unnecessary. Journal of Royal College of Surgeons, Edinburgh, 28 (1), 35-40.

14. Lucian L. Max L. Ramensofsky.: Laparoscopy for questionable appendicitis. Anals of Surgery 1980 191 (4), 410-13.

IATROGENIC GASTROINTESTINAL PERFORATIONS

NEELAM JAVID; LUBNA ISHAQ BHATTI; JAWAD SHER, ABID QAZI . IFTIKHAR A JAN . NAEEM-UZ-ZAFAR KHAN .

ABSTRACT

Iatrogenic injuries are an unfortunate but significant aspect of any therapeutic intervention, contributing considerably to morbidity and mortality statistics. Although the element of human error may never be entirely eradicated, it can be reduced substantially by taking meticulous precautions to ensure mistakes are never repeated. We have analysed nine cases of unusual forms of gastrointestinal perforations in children. The causes of perforations. modes of presentation, methods of management and the subsequent outcome is discussed. Most of the cases were managed operatively but in selected cases a conservative approach was practiced. Only one death occurred in this series and that also due to the late presentation and extremely moribund state of the patient.

KEY WORDS: Perforation, Gastro-intestinal Perforation, Iatrogenic injury

INTRODUCTION

There are a host of reasons for gastrointestinal (GIT) perforations in the paediatric age group. The commonest cause nowadays may be due to the recent introduction of endoscopic instrumentation and surgical procedures. There are well known causes of spontaneous gastrointestinal perforations due to gangrene and congenital bowel obstruction such as midgut volvulus, intussusception, bowel atresia, Hirschsprung's disease and meconium ileus, where the perforation may be caused in the proximal dilated bowel.

In our country, there are several prevalent diseases like enteric fever, severe gastroenteritis and tuberculosis which are also responsible for gastrointestinal perforation. Recognized late and managed improperly, gastrointestinal perforation leads to severe morbidity and mortality.

In this paper however, only those GIT perforations in children are reported which were produced inadvertently and were iatrogenic in nature. A total number of nine cases are reported, whose management strategies and outcome is discussed. These cases underscore the importance of awareness of the possibilities of unusual causes of GIT perforations .

CASE SERIES

CASE NO.I

A two day old male, premature born at 32 weeks of gestation, was admitted with delay in passage of meconium and abdominal distension since birth. On physical examination the abdomen was moderately distended with normally situated anal orifice. On plain X-ray there was only gaseous distension of the bowel. A lubricated thermometer was introduced to check the patency of the anl canal and rectum. This procedure resulted in sudden deterioration of the patient's condition with a tympanitic abdomen. Plain X-Ray abdomen showed pneumoperitoneum (fig 1). It

Fig 1: Massive pneumoperitoneum with displacement of liver towards midline as a result of gastlic perforation from vigorous resuscitative efforts following birth asphyxia.

Prof. Naeem-uz-Zafar Khan, Head Dept. of Paedriatic Surgery
The Children Hospital, PIMS, Islamabad

was obvious that the perforation was caused inadvertently by the thermometer. The patient was taken to the operating room within 2 hours of the accident. At laparotomy, a sigmoid perforation was noted. Rest of the bowel was normal. After evacuation of meconium, the perforation was closed using 4.0 vicryl. Post operative recovery was uneventful. On discharge patient was tolerating oral feeds and had normal bowel habits. We assumed that delay in passage of meconium was due to prematurity.

CASE NO.2

A 6 month old infant was admitted with signs and symptoms of meningitis and seizures. Management included oxygen therapy through nasal tube and Nasogastric feeding. Inadvertently, the nasogastric tube was connected to the oxygen cylinder. resulting in what was presumed to be gastric perforation with massive pneumoperitoneum and surgical emphysema of the abdomen and chest wall. Because of his general poor condition from meningitis, the patient was initially managed conservatively with nasogastric suction, parental antibiotics and appropriate fluids. After initial improvement, the patient's condition deteriorated with signs of established peritonitis and thus the patient had to be surgically explored. The whole peritoneal cavity was full of bile and the bowel was matted with fibrinous exudates. A perforation was noted on the medial side of the junction of the second and third part of duodenum just above the opening of the ampulla of Vater and adjacent to the head of the pancreas. Because of technical difficulties, only a one layer closure of perforation with interrupted 4.0 vicryl was performed and a peritoneal lavage was done. The patient was treated with broad spectrum antibiotics. nasogastric suction and parenteral fluids. He made an uneventful and complete recovery, both from the accidental perforation and meningitis and was discharged after a fortnight from the paediatric intensive care unit.

CASE NO.3

A three day old girl, born after full term pregnancy, was admitted with a history of abdominal distension and bilious vomiting. Baby was breastfed since birth. On examination the patient was in shock with peripheral circulatory collapse and a tympanitic abdomen. Plain X-ray showed free subphrenic gas. A working diagnosis of bowel perforation due to some congenital anomaly was considered. After initial resuscitation, the patient was taken to the operating room for laparotomy. On opening the abdomen a moderate amount of seropurulent free fluid was drained from the peritoneal cavity. On examination of bowel. a flexible, wooden splinter with a sharp tip was noted which was 2.5 cms, long (fig 2) and was protruding half outside and half within the lumen of the anterior surface of the first part of duodenum; this was the cause of perforation with peritonitis. The perforation was closed with one stitch of 5.0 vicryl, which sufficed to seal the perforation. Postoperative period was unremarkable and the patient made a complete recovery. On further enquiry from the mother, it transpired that "ghutti" was given to the baby . It is assumed that this thorn was one of the components of the ghutti.

Ghutti is a form of herbal concoction used commonly in the subcontinent by illiterate people to clean the bowel and relieve colic of the neonate

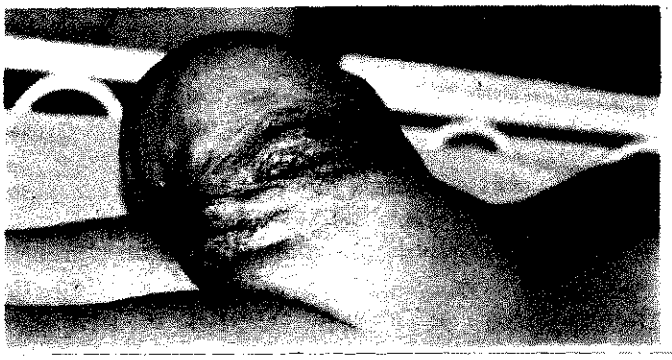


Fig.2: 4 years old child (Case 2) sought advice for the first time because of sudden enlargement in a sacrococcygeal teratoma present since birth

CASE NO 4

A new born male was transferred from a local hospital having had resuscitation for birth asphyxia, which involved nasogastric suction, use of ambu bag, endotracheal intubation and correction of acidosis. On admission he showed signs of peripheral circulatory collapse, tachycardia and dyspnea with a tympanitic abdomen. Plain X-ray chest and abdomen revealed free gas under the diaphragm indicating bowel perforation. After resuscitation and stabilizing the patient,a laparotomy revealed generalized peritonitis and a linear tear of about one cm. In the lesser curvature of the stomach which was closed in two layers. Decompressing gastrostomy was also instituted. He was treated post-operatively with a broad spectrum antibiotic and I/V fluids. Later on, the integrity of stomach repair was judged by a gastrograffin study before feeding was started.The patient was allowed home after a fortnight from the neonatal intensive care unit.

Case No. 5

A full term baby delivered by a Dai (traditional birth attendant) at home had what appeared to be a case of difficult labour.Vigorous abdominal massage was applied by a Dai to assist the delivery of the child. On admission, 24 hours after birth the baby was completely collapsed, with dusky cyanosis and abdominal distension,

marked tachypnea with shallow breathing and profound hypothermia. He was in shock and acidotic.
A plain X Ray abdomen showed massive pneumoperitoneum with displacement of liver and viscera towards the midline. After initial resuscitation laparotomy was performed and a linear tear of 1.5cms in the lesser curvature of the stomach was noted which had resulted in peritonitis with pneumoperitoneum. The perforation was closed and peritoneal lavage done. Because of the poor condition of the patient he was put on a ventilator for assisted breathing, but despite vigorous efforts the patient expired 12 hours after surgery.

The actual cause of perforation could not be established but it can be presumed that external abdominal trauma and asphyxia was responsible for the perforation and the delay in transfer of the patient led to irreversible shock.

Case No.6
A nine year old boy with a history of fresh rectal bleeding intermittently but normal bowel habits was suspected of having rectal/colonic polyps. Sigmoidoscopy was performed under general anaesthesia to examine and remove the polyp. A juvenile polyp was noted at the rectosigmoid junction and it was removed by using a cutting biopsy forcep. The patient was allowed home.

After four days of the procedure, the patient was admitted with fever, abdominal pain and distension of the abdomen. There was generalised tenderness, most marked in the left lower quadrant of the abdomen. A plain X-Ray showed free gas under the diaphragm and distension of loops of small intestine indicating ileus. An ultrasound was done which showed a small collection in the pelvis. The patient was treated conservatively with nasogastric suction, parenteral fluids and infusion of metronidazole and gentamicin.

His general condition continued to improve and therefore surgery was not considered. Seven days after admission he was passing stools normally and was taking oral fluids .Fever had subsided. He was allowed home on oral metronidazole and Zanitrin for anothel seven days. On follow up after a fortnight the patient had completely recovered .In this case the perforation which was caused by the endoscopic procedure was perhaps small and sealed by the adjacent viscera.

A lesson which we can draw is that conservative management has a definite role in the management of a perforated viscus especially if it is localised and detected late and on close monitoring the patient's condition continues to show improvement.

Case NO.7
A three year old girl presented with a history of having ingestion of floor cleaner four months earlier and had a 10 cms long irregular tight stricture of the proximal thoracic oesophagus (fig 3). On oesophagoscopy and attempted dilatation with bougies, the oesophagus was inadvertently perforated resulting in mediastinitis and right pneumothorax. She was treated conservatively in intensive care unit with pharyngeal suction, parenteral fluids and an under water seal chest drain. A feeding gastrostomy was placed after five days.

She was allowed home after a fortnight stay in the hospital. Eventually after four months a retrohilar reverse gastric tube through a separate laparotomy and right thoracotomy approach was constructed. She has remained well six years after her oesophageal replacement.

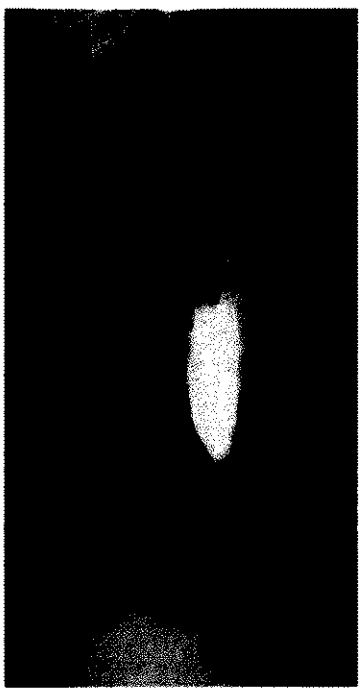


Fig 3: Esophageal stricture causing complete obstruction to passage of swallowed gastrograffin. These strictures are prone to perforations during attempted dilatations.

Case No 8
A two year's old boy had middle oesohageal perforation during an attempted dilatation of an oesophageal stricture caused by caustic injury. This resulted in pneumothorax and symptoms of mediastinitis (fig 4). He was treated conservatively with pharyngeal suction, feeding gastrostomy tube and parenteral fluid. Chest tube was not placed because of the small size of the pneumothorax.

Subsequent X-rays showed fluid in the right pleural cavity and insertion of an underwater seal chest tube drainage became necessary which yielded frank pus. The patient recovered without further problems and after three weeks of admission he was allowed home. Eventually an oesophageal replacement was carried out using right colonic conduit.

He is well for the last seven years.

Case No. 9

A two year old patient was admitted with a stricture of the lower third of the oesophagus secondary to untreated gastro-oesophageal reflux. An attempted oesophageal dilatation resulted in a perforation of the lower abdominal oesophagus. This was recognised because the patient was developing signs of peritonitis and an X-ray showed free gas under the diaphragm. Laparotomy was performed and a linear tear of 0.5 cm was noted on the anterior surface of the abdominal oesophagus at its junction with the stomach.

This was closed and reinforced with adjacent cardia of the stomach. A feeding gastrostomy was placed. The patient made an uneventful recovery and was allowed home on gastrostomy feeding after ten days. Eventually, a colonic conduit was placed as an oesophageal replacement.

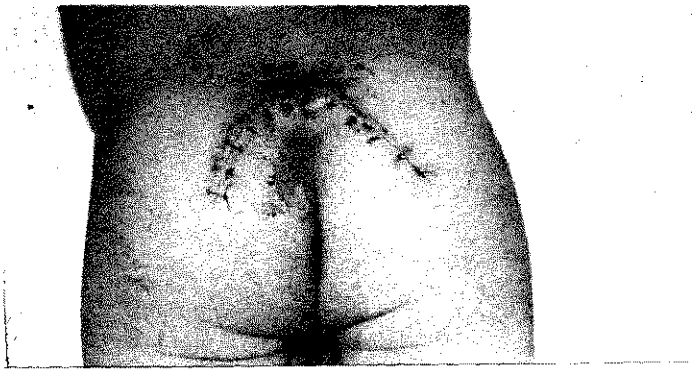


Fig. 4: Post-operative apparance of the patient in Fig. 2. Note the restoration of gluteal folds and reconstition of anorectal sling

DISCUSSION

One of the most common causes of gastrointestinal perforations today may be those due to invasive surgical procedures especially endoscopic instrumentation and the incidence of iatrogenic perforations has increased with the tremendous growth in the number of these procedures. Perforations of the oesophagus are no exception. but other etiologies do exist such as accidental caustic ingestion, spontaneous perforations, perforations resulting from foreign bodies and malignancies. In a report[2] by Ballesta Lopez C. et al from the School of Medicine, University of Barcelona ascertaining causes of iatrogenic oesophageal perforations, they found that 28.6%, of perforations followed hiatal surgical procedures, l9%, occurred after diagnostic endoscopies, 19%, after endoscopic dilatations and 19% during removal of foreign bodies.

Case 8 exemplifies this inherent hazard associated with oesophageal dilatations. In our previous study[3] this was the commonest complication we encountered. From our experience it came to light that various factors prevailing in our society militate against the success of frequent dilatations. Poor patients coming from remote areas with inadequate realisation of the fact that regular dilatations are an important therapeutic aspect of strictures, we feel that a definitive procedure of oesophageal replacement, although a major undertaking, should be offered more readily than the usual criteria laid down for this procedure. This applies especially for those patients where compliance is poor, and who return only when the stricture is so far advanced that attempts at dilatation readily produces perforation .

Early diagnosis with expedient management is the secret of reducing morbidity and mortality. After any surgical or endoscopic procedure, a high degree of suspicion and close monitoring will allow early detection of perforation. One should watch for signs of shock, pallor, tachycardia, tachypnea, subcutaneous emphysema, chest pain and fever.In cases of doubt, confirmation can be achieved by an oesophagogram using a water soluble contrast medium.

Outcome depends on the cause, location, size and the interval between the injuries and the initiation of treatment. The site of oesophageal perforation also determines the extent of sequelae. Prognosis is worse in thoracic lesions with significant mortality if treatment is delayed for more than 24 hrs. Selecting the most appropriate therapy also depends on the aforementioned factors. Although thoracotomy with reinforced primary repair and mediastinal drainage has been recommended for cases with manifestations of mediastinitis"[6]. with the advent of effective antibiotics and better intensive care facilities, conservative management consisting of antibiotics, gastric decompression, parenteral nutrition and thoracocentesis, if needed, can be appropriate especially in injuries[5] resulting from nasogastric tubes .

There may be several long term complications of oesophageal perforations such as external and internal fistular formation, mediastinal abscesses and empyema of thorax.[7] Gastroduodenal perforations may also be produced in the same manner as are mentioned for oesophageal perforations. In addition, in neonates gastroduodenal perforations are also produced by vigorous resuscitative efforts at birth, nasogastric tube injuries and congenital distal obstructive lesions such as atresias etc.

Perforation of the medial aspect of the second and third part of duodenum produced inadvertently by the rapid introduction of a large amount of oxygen through a nasogastric tube in case 2 in our series could

have been avoided if a strict colour coding of oxygen tubes and nasogastric tubes are followed. One should always confirm the length of nasal oxygen tube before connecting it to the oxygen supply. Ideally one should administer oxygen with specially designed oxygen delivery tubes which are fixed on the external nares.

Gastro- duodenal perforations caused by thorny material contained in "ghutti", such as in case 3 in our series is most unusual and should be taken note of as a possible complication of local concoctions given to children at birth as a cultural habit in our society. Perforations in the gastroduodenal area are easy to detect because of a sudden collapse of the patient and a plain X ray will reveal free gas under the diaphragm. The site of abdominal visceral perforation however, can only be detected at laparotomy. Management entails vigorous resuscitative measures and early laparotomy.A delay in diagnosis and concomitant congenital anomalies are associated with a high morbidity and mortality.

In neonatal and paediatric age group the most common cause of colonic perforation is distal obstruction secondary to atresias, Hirschsprung disease and anorectal agenesis. However in this series we have considered only iatrogenic perforations of the colon to highlight the importance of an avoidable injury.

Iatrogenic injuries of the colon can be due to barium enemas, thermometer and instrumentation[8]. In case 1 where sigmoid perforation was produced by a thermometer and recognised early, prompt surgical intervention prevented any serious sequelae. Despite its rarity, rectal/sigmoid perforation occurring during thermometer instillation is one hazard that one should be aware of and take measures to avoid, particularly because rectal temperature is a more sensitive indicator of an infant's temperature than an axillaly temperature measurement[9]. In contrast to the immediate action employed in case 1, in case 6 the colonic perforation went undetected for four days. Here the bodys defence mechanism sealed the perforation and conservative management by parenteral antibiotics and intravenous infusion was sufficient. Conservative management is advocated in those patients with good bowel preparation'°, for silent colonic perforation, those with localized symptoms and signs and those where diagnosis is made eight hours after colonoscopic injury". If there is a large perforation, generalised peritonitis or failure to improve on conservative treatment then surgical exploration is necessary.

In summary, gastrointestinal perforation can be largely prevented if early diagnosis of congenital anomalies producing luminal obstruction is entertained. Similarly iatrogenic perforations which are specifically highlighted in this article, could also be minimised with awareness and practice of certain precautions. Importance of early detection of visceral perforation and prompt action cannot be over emphasised. The mainstay of deciding which course of action to follow is frequent observation and accurate anticipation of development of symptoms, hence one can obviate the need for surgery in selected patients.

REFERENCES

1. Newcomer M.K; Brazer S.R; Complications of upper gastrointestinal endoscopy and their management. Gastrointest-Endosc-Clin-N-Am. 1994 Jul; 551-70.
2. Ballesta Lopez Z.;Vallet Fernandez J.; Catarci M; Bastida Vi X; Iatrogenic perforations of the esophagus. Int-Surg. 1993 Jan-March; 28-31.
3. Naeem-uz-Zafar Khan et al: Management aspects and associated problems of established esophageal strictures. Pakistan's J. Med. Sci. April-June 1993, Vol.9: No.3; 253-260.
4. Kim Deobald J; Kozarek R.A.; Eosophageal perforation: an 8 year review of a multispecialty clinic's experience. Am.J.Gastroenterol. 1992 Sep; 1112-9.
5. Weber TW. Esophageal rupture and perforation. In: Welch IJ? Randolph JG, Ravitch MM, et al,. (Eds). Pediatric Surgery (4th Ed) Year Book Medical Publishers. Inc, Chicago. pg.727
6. Rowe I.M.; O'Neill J.A.; Grosfield J.L.;Fonkulsrud E.W.; Coran A.G.; Essentials of Pediatric Surgery; 1995, pg 419.
7. Ohri S.K.; Liakakos T.A.; Pathi V.; Townsend E.R.; Fountain S.W.; Primaly repail of iatrogenic thoracic esophageal perforation and Boerhaave's syndrome. Ann. Thorac. Surg. 1993; pp 603-6.
8. Williams S.M.: Harned R.K.; Recognition and prevention of barium enema complications. Curr. Probl. Diagn. Radiol.; 1991; pp 123-151
9. Morley C.J.; Hewson P.H.; Thornton A.J.; Cole T.J.; Aillary and rectal temperature measlll-el11ents in infants. Arch.Dis.Child.; 1992; pp 122-5.
10. Hall-C.; Dorricott N.J.; Donovan I.A.; Neoptolemos J.P.; Colon perforation during colonoscopy; surgical versus conselvative management. Br. J.Surg. 1991 May: 542-4.
11. Kavin.H; Sinicrope.F.; Esker.A.H; Management of perforation of the colon at colonoscopy. Am.J.Gastroenterol. 1992 Feb; 161-7.

ROLE OF FINE NEEDLE ASPIRATION BIOPSY AND CYTOLOGY (FNABC) IN A COLD NODULE OF THYROID AND ITS COMPARISON WITH HISTOPATHOLOGY

SHAHIDA AFRIDI, ASADULLAH KHAN, S. WAQAR AHMED, IRSHAD WAHEED

ABSTRACT

This study evaluates the diagnostic importance of Fine Needle Aspiration Biopsy and Cytology (FNAC) in a palpable cold nodule of thyroid. Conducted at Jinnah Postgraduate Medical Centre, Karachi over a period of two years from January 1992 to December, 1993, this study included 62 patients; 57 were reported as benign and two as malignant on both FNABC and Histopathology. Two cases reported as "abnormal cells seen" and "follicular cells suggestive of malignancy" turned out to be malignant on Histopathology. One case was a Cyst which disappeared on aspiration and did not reoccur/refill on 6 months follow up. The accuracy of our result is 95% and 3.2% cases suggesting abnormality, proved to be malignant.

FNABC has an excellent patient compliance, is simple, economical and quick to perform in an out patient department, to differentiate between benign and malignant conditions.

KEY WORDS: Thyroid cold nodule, Fine needle aspiration biopsy, cytology, Biopsy, FNAC, Thyroid, Cytology, Aspiration Cytology.

INTRODUCTION

Fine needle aspiration biopsy and cytology of thyroid nodules is a sensitive and specific tool for detection of thyroid cancers, thus preventing unnecessary operations.

This technique has been in use since long. It was first introduced for thyroid tumors in America in 1926[1]. Later Martin and Ellis in the 1930's[2] studied the role of FNABC on thyroid, lymphnode and breast. Most of the thyroid nodules are benign and these can be differentiated from malignant by FNABC[3]. A solitary nodule is a common presentation of thyroid disease and may be cold, warm or hot. Solitary cold and solid nodules have greater chances of being maligant (5-10%)[3].

FNABC of a cold nodule of thyroid has been used in Sweden since the begining of 1950[4] and 8000 FNABC were carried out during the last twenty years. Lowhagen et al., (1979)[5] also prefered this method in the diagnosis of thyroid disease. Boey et al. (1979)[6] recommended FNABC in every thyroid nodule, regardless of its clinical nature, whether neoplastic or not. Silverman (1986) and Frable (1976) recommended pre-operative use of FNABC in a cold nodule of thyroid[7]. FNABC helps in the operative planning for a definitive procedure based on tissue diagnosis.

PATIENTS AND METHODS

Patients selected for this study were from Surgical Unit I and surgical Outpatient Department of JPMC. History was taken and general physical and local examinations were carried out. Thyroid scan was helpful in determining the functional status of the gland. Cases of cold nodule on thyroid scan were selected for this study.

Material required for FNABC for specimen collection:
1. Spirit swab
2. 22 guage needle with syringe
3. Glass slides

FNABC was performed with the patient in supine position with hyperextended neck. After examination and cleaning, negative pressure was created in the syringe, to aspirate the cells, with 0.2-0.3 cc air approximately[8]. The patients were asked not to swallow or talk during the procedure. The swelling was fixed with one hand and the other hand was used to introduce the needle into the swelling. The needle was then moved to and fro

DEPARTMENT OF SURGERY
JINNAH POSTGRADUATE MEDICAL CENTRE KARACHI

to get the tissue detached. The needle was then withdrawn and separated from the syringe; air 0.2-0.3 cc was sucked and the needle was reattached and the material was expressed on slides. Examination of slides was carried out in the Histopathology department, after special staining [with Hematoxylin[7], Eosin, Orange G-6, E-A-50, or Eosin Azure-50].

RESULTS

TABLE I
SEX DISTRIBUTION

Total No. of patients	62	Percentage
Female	52	83.8
Male	10	16.2
M:F (Ratio)	1:5.2	

TABLE I I
SEX DISTRIBUTION ACCORDING TO AGE

Age (Years)	Male	Female	Number	Percentage
0-10	-	-	-	-
11-20	2	3	5	8.0
21-30	2	15	17	27.5
31-40	3	25	28	45.2
41-50	1	8	9	14.5
51-60	2	1	3	4.8

TABLE I I I
DISTRIBUTION OF SWELLING ACCORDING TO SITE

Site	No. of patients	Percentage
Right Lobe	15	24.2
Left Lobe	11	17.2
Both Lobes	26	41.9
Isthmus	5	8.1
Right Lobe with isthmus	5	8.1

TABLE I V
DISTRIBUTION ACCORDING TO SIZE

Size (cm)	No. of patients	Percentage
1-3	5	8.1
3-6	28	45.2
7-9	21	33.9
10-12	6	9.6
13-15	2	3.2

TABLE V
RESULTS OF FNABC

No.	FNABC Results	No.	Percentage
1.	No malignant cell seen	57	92
2.	Malignant cell seen	2	3.2
3.	Abnormal cell seen	1	1.6
4.	Follicular cell seen Suggestive of growth	1	1.6
5.	Cyst - Fluid No abnormal cells seen	1	1.6

TABLE VI
COMPARISON BETWEEN FNABC AND HISTOPATHOLOGY

62 FNABC Result		61 Histopathology Results		
57	Benign	Multinodular goiter	31	57
		Solitary Nodule	21	
		Follicular Adenoma	5	
01	Abnormal Cells	Papillary Carcinoma	1	
01	Follicular Cells Suggestive of malignancy	Follicular Carcinoma	1	
02	Malignant	Papillary Carcinoma	2	
01	Cyst	Biopsy not done Because of complete regression and Cytology of fluid showed no abnormal cells.		

DISCUSSION

Thyroid diseases are quite common in Pakistan and present as a swelling in front of the neck, which may be due to goitre, inflammation, cyst or malignancy etc[17].There are no early clinical signs, symptoms or investigations which can differentiate between benign and malignant conditions preoperatively except FNABC which is the only reliable diagnostic test and which is helpful in the subsequent management[18] of these patients.

Single Non-functioning cold and solid nodule has a 5-10% chance of being malignant[19]. It is recommended by Lowhagen et al. that FNABC should be performed on every thyroid cold nodule, whether neoplastic or not[4,20,22].

FNABC is safe, simple, economical and time saving technique, requiring no anaesthesia and has no danger of dissemination. FNABC has high rate of accuracy in experienced hands. FNABC accuracy in different series of studies is shown in Table VII.

Thyroid pathology which can be diagnosed on FNABC alongwith clinical background includes goitre, inflamma-

tions, papillary carcinoma, medullary and carcinoma and anaplastic carcinoma[21].

TABLE VII
COMPARISON WITH OTHER STUDIES

Refferences	Year	No. of Cases	Accuracy
Collachio et al[9]	1980	300	95%
Niazi[10]	1981	50	82.6%
Ackerman et al[11]	1985	420	98%
Silverman et al[12]	1986	60	95%
Hawkins et al[13]	1987	415	95%
Cusick et al[14]	1990	307	58%
Larosa et al[15]	1991	827	97.7%
Alam Saeed[16]	1991	56	94.4%
Present study	1992-93	62	95%

FNABC' positive for malignancy' cases have priority for admission and treatment with better treatment plan and single operation.

Its limitations are that it cannot differentiate between follicular adenoma and follicular carcinoma because this distinction is made on histological criteria, including capsular and vascular invasion[22].

Cells obtained by FNABC can be subjected to culture and the effects of radiotherapy and chemotherapy can be detected. It is also useful for follow up in patients with carcinoma treated with radiotherapy and chemotherapy[23].

Sometimes it is both diagnostic and therapeutic in cystic swellings. Aspiration of the fluid is the only treatment in many cysts, owing to the fact that some thyroid cysts are deprived of epithelial lining and emptying of their content often results in regression of the cyst[24], risk of missing intracystic carcinoma is negligible (0.5%) (Gasper and Barness)[24,25]. Usefulness of FNA can be increased if used in combination with electron microscopy.

Other Uses of FNABC

DNA flow cytometry on the material obtained by FNABC gives rise to aneuploid peaks in malignancy but not in benign thyroid pathology[27].

Enzyme Study[28]

It can be carried out like for acid phosphates in matastatic carcinoma of prostate. Medullay carcinoma of thyroid can be diagnosed by immunoperoxidase staining for calcitonin on the aspiration material.

CONCLUSION

FNABC has an excellent patient compliance, is very simple, ecnomical and quick to perform in an Out Patient department and leaves no scar.

It is the only pre-operative diagnostic test that can differentiate between benign and malignant conditions and it has very high accuracy in experienced hands.

REFERENCES

1 Scanlan P, Dowling M, Dervan P, Carrigan T, Hetternan S, Firth R In. J. Med. sci., 1993, 5:177-9.

2. Martin HE and Ellis EB, Biopsy by needle puncture and aspiration. Ann. Surg., 1930, 92:578-89.

3. Schwartz, shires Spencer, Principles of Surgery

4. Bailey and Loveís Short Practice of Surgery 22nd Edition.

5. Lowhagent T, William JS, Lundell g et al. , Aspiration Biopsy and Cytology in the diagnosis of thyroid cancer world. J. surg., 1981, 5:61.

6. Boey et al., FNAC versus drill needle biopsy of thyroid nodules. A controlled clinical trail surgery, 91.

7. Frable WJ Thin needle aspiration biopsy. A personel experience with 409 cases. AM. J. Clin. Pathol., 1979, 65:168-182.

8. Handbook of Histopathological and histochemical Techniques, Culling third edition, pp, 216-17.

9. Collachio and Oertel, 1988, Cancer.

10. Niazi M, FNABC of a cold nodule of thyroid gland. M. Phil thesis, University of Karachi, 1988.

11. Ackerman: Surgical Pathology.

12. Silverberg E: Cancer Statistics, 1977, 27:26.

13. Hawkins H, Bellidod Bernal C et al., Fine needle aspiration biopsy cytology in the diagnosis of thyroid cancer and thyroid disease. Cancer, 1987, 59:1206-9.

14. Cusick, Crawny N.P., Bernal C. et al. Fine needle aspiration biopsy cytology in the diagnosis of thyroid cancer and thyroid disease. Cancer; 1990, 26:807-811.

15. LaRosa, Belfiore A, Padove G et al., The frequency of cold thyroid nodules and thyroid malignancies in

patients from iodine deficient area cancer, 1991, pp, 3096-2.

16. Alam Saeed, FNAC in cold nodule of thyroid. M. Phil thesis University of Karachi, 1991.

17. Caraway NP, Sneige N, Samman NA: Diagn cytopathology, 1993, 3:345-50.

18. Cuschieri, Giles, Moosa: Essential surgical practice.

19. Davis Cristopher: Text book of surgery.

20. Burkitt, Quick, Gatt, Deakin Essential surgery.

21. Beg MHA and Mirza RA, Endemic goitre and the realities (editorial) Specialist, 1991, 7:2.

22. Current Surgical diagnosis and treatment.

23. Einhorn J Sand Franzenís: Thin needle biopsy in the diagnosis of thyroid disease. Acta Radiological, 58, pp 321-26.

24. Khan S H and Khalig T: Evaluation of solitary cold nodule of thyroid. FNABC as a major diagnostic tool. J Pak Inst Med Sci, 1990, 1:11-3.

25. Gasper MR: Aspiration of the breast cyst. Clif Med, 1961, 95:227-31.

26. Barness MC: Aspiration biopsy of rumors in obscure or difficult localities under roentgenoscopic guidance. AM J

27. RM Rodriguez P Parilla J Sola et al: Comparison between preoperative cytology and intraoperative frozen section biopsy in the diagnosis of thyroid nodules. Br. Med. J., 1994, 81:1151-54.

28. Palmer JO, MC Divitt RW Stone KR et al flow cytometeric analysis of breast needle aspirate. Cancer, 1988, 62:2387-91.

BENIGN ORO-FACIAL TUMOURS

NASREEN AMANAT

ABSTRACT

Among the various developmental and acquired causes of facial disharmony, odontogenic cysts and tumors, dental hamartomas and reactive bone disorders rate the most common. Most of these lesions are discovered fortuitously. These patients either receive no treatment or undergo extensive resections. Detailed are four cases of oral pathology i.e. odontogenic tumor (ameloblastoma), odontgenic cysts (odontogenic keratocysts), dental hamartoma (compound odontome) and a reactive-bone disorder (central giant cell granuloma), as causes of facial disharmony and their surgical management.

Key words: Central giant cell granuloma, ameloblastoma, ameloblastic carcinoma, odontogenic keratocysts, primordial cyst.

INTRODUCTION

A patient suffering from benign oral tumor or cyst is likely to undergo different types of surgical treatments because of lack of consensus as to their management. We present our experience of four such cases.

CASE NO. 1

A seven years old female presented with a painless swelling on the left side of face, which was noticed 8 months earlier (Fig. I). A gradual increase in the size of this swelling led to facial asymmetry. Clinical examination revealed expansion of buccal and lingual plates of the left body of the mandible from the first mandibular molar to the central-incisor region with bowing of the lower border. The swelling was hard, well circumscribed and non-tender. Panoramic view of the mandible revealed a multilocular radiolucency involving the left body of the mandible . Hematological and biochemical indices were within normal limits. Aspiration biopsy of the lesion was inconclusive . Under general anesthesia, the lesion was enucleated through an intra-oral, subperiosteal approach. The lesion was non-encapsulated, dark red in colour, friable and bled easily. The patient required transfusion of one unit of blood per-operatively. Primary dentition and tooth buds of the permanent dentition were preserved. The lesion was identified as an osteoblastoma on histological examination. Since the clinical picture was not consistent with the histopathological diagnosis, a second opinion was sought and the lesion was reported to be a central giant cell granuloma. Radiological examination at one year follow up shows normal bone trabeculae.

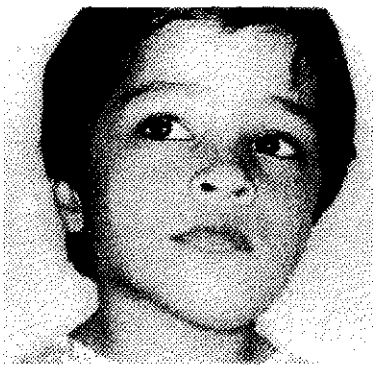


Fig. I Facial assymetry with bowing of left border of mandible

CASE NO. II

A married 22 year old female, presented with a swelling on the left side of the face. There was a discharging sinus on the left angle of the mandible. The swelling had first been noticed 14 months earlier with a gradual increase in size. There was a history of pain and antibiotics were used on more than one occasion . Clinical examination revealed a swelling consistent with expansion of the buccal plate of the left mandible from the angle to the pre-molar area with bowing of the lower border of the mandible . Intra-orally the left lower first, second and third molars were missing. Panoramic view revealed a compound composite odontome occupying the left

ASSISTANT PROFESSOR ORAL SURGERY
A.K.U.H. KARACHI

body of the mandible (Fig. II). There was a separate unilocular radiolucency present distal to the odontome in the mandibular ramus and a green stick pathological fracture at the angle. Under general anaesthesia, both lesions were enucleated, via the subperiosteal approach, after removing the buccal plate of the left mandible. The odontome consisted of numerous discrete denticles . The fracture at the left angle of the mandible was reduced and immobilized with eyelets and intermaxillary fixation. Histopathologically the proximal lesion was identified as a compound odontome with dentigerous changes in the follicle and the distal lesion was a Keratocyst. There was complete resolution of the asymmetry over an eight months' period and there has been no recurrence of the keratocyst over a three years follow up.

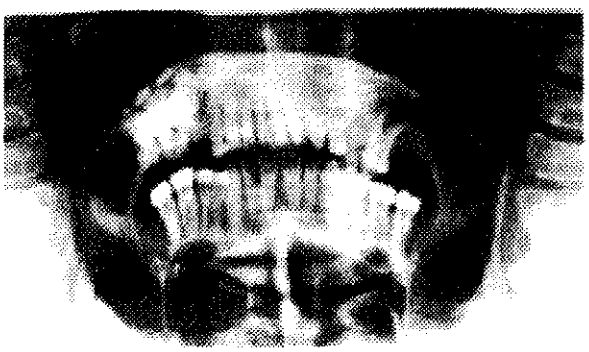


Fig. II Panorax showing missing teeth, compound odontome and a radiolucency in the left ramus

CASE III

A 25 year old male presented with a grossly enlarged mandible of unknown duration. An aspiration biopsy had been attempted with inconclusive result. Clinical examination revealed hard, irregular surface of the entire mandible. Intra-orally the dentition was intact with a sinus in the right premolar area. The panorax revealed a large multilocular radiolucency from angle to angle (Fig. III). Both canines were lying along the lower border of the mandible with both the mandibular third molars in horizontal impaction. The diagnosis of a keratocyst was made. Since the patient did not wish to have a dental clearance, the lesion was enucleated intra-orally via a subperiosteal incision. Mandibular canines were removed and apicectomy and retrograde fillings were done on exposed dental roots. Immediate post-operative recovery was uneventful but for a wound breakdown in the area of the sinus. Pus from the sinus was noticed about twelve weeks after the surgery. Samples were taken for the culture and sensitivity and appropriate antibiotic therapy was instituted . The infection could not be controlled with antibiotics five months after the first procedure a recurrence was observed; this was again enucleated and curettage was performed. At this time the right lower third molar was also removed and the post-operative recovery was uneventful. Orthograde root fillings were carried out on all mandibular teeth after the second operation. There has been a gradual reduction of the radiolucency observed on the panoramic views . Three years after the second operation the patient was lost to follow up.

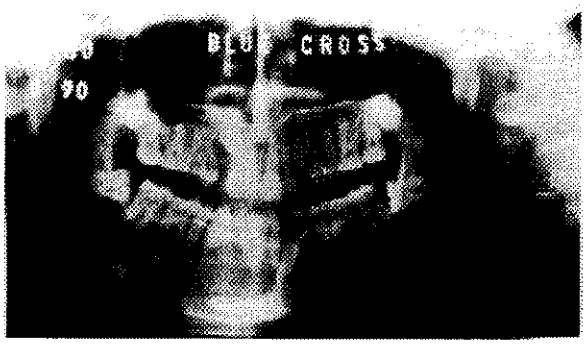


Fig. III Panorax showing radiolucency from (R) coronoid to the left coronoid. Note all 4 impacted 3rd molars and canines

CASE - IV

A fifty-year old female presented with a massive growth in the mandibular symphysis, extending from the right premolar to the left premolar region due to which the patient was unable to close her mouth (Fig. IV). The growth itself was well demarcated, non-lobulated and hard in consistency with no fluctuation, bruit or tenderness to palpation.

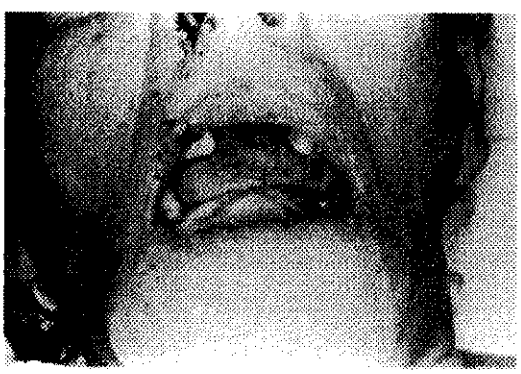


Fig IV Grossly enlarged mandible. Patient is unable to close her mouth

Panorax revealed a well circumscribed radiolucency with a fair amount of interspersed calcification (Fig. V). A diagnosis of a calcifying odontogenic tumour or an ameloblastoma was made. Dental clearance was performed and the lesion was approached intra-orally, which was found to be calcific, hemorrhagic and well circumscribed. Segmental osteotomy was performed leaving an intact lower border of the mandible followed by thorough curettage. Profuse bleeding was encountered

from the soft tissues as well as from the diseased bone requiring transfusion of 8 units of whole blood. At the time of recovery from anesthesia, collapse of the right upper lobe of lung was observed. Bronchoscopy was performed to remove the blood clots from the tracheo-branchial tree. Subsequent recovery was uneventful. Histopathology report confirmed an ameloblastoma. There was a wound breakdown one week post-operatively which was allowed to granulate. Eighteen months after operation there was radiological and histological evidence of recurrence but the patient had declined further surgery.

Fig V Panorax showing radiolucency interspersed with radio-Opacities

DISCUSSION

The oro-facial lesions discussed above are all essentially benign and are usually discovered on routine dental examination or when facial asymmetry becomes prominent. Pain is rare, unless there is secondary infection and under the circumstances a draining sinus may also be present. On radiological examination these lesions appear mostly as radiolucencies with variable amount of interspersed radio opacities. Some pathological entities that commonly present as radiolucency in the jaw are odontogenic keratocyst, ameloblastoma, central giant cell granuloma, cherubisum, odontogenic myxoma, aneurysmal bone cyst, central hemangioma and other vascular lesions[1]. Since there is much variation in the behavior of the multilocular lesions, their management differs accordingly. During evaluation of radiolucent lesions of the jaw the possibility of a vascular lesion must always be kept in mind due to its potentially serious implications. All of the lesions discussed in this report are locally aggressive but do not metastasize, in contrast to the malignant tumours of the head and neck, which are not only locally invasive but spread via lymphatics or the blood stream. For these malignant tumours extensive surgical resection coupled with pre and post operative radiotherapy is recommended. The role of chemotherapy still remains palliative in head and neck malignancies. For benign oral tumors, a more conservative approach is preferable The entire mandible or maxilla can be exposed through an adequately designed intra-oral subperiosteal flap. This approach not only gives good access for enucleation, curettage and segmental resection but also allows retrograde root fillings to be done at the same time.

Central giant cell granulomas may occur at any age, but the majority present between 10-25 years and occur in the tooth bearing areas of the jaw - usually the mandible[2]. The lesion usually presents as swelling of the bone and facial asymmetry[3]. Radiologically it appears as non-corticated radiolucency causing displacement of involved teeth and their root resorption[4]. Growth is usually rapid and when the cortical plate is perforated, the lesion may present as a peripheral giant cell granuloma. Enucleation and curettage is the treatment of choice. Recurrence does not occur even if some tissue is left behind.

The term odontome originally included all odontogenic tumors and cysts. Currently it is used to designate a non-neoplastic, developmental anomaly that contains fully formed enamel and dentine. Odontome can, therefore, be considered a dental hamartoma containing calcified dental tissues[5] . Complex and compound odontomes are closely related malformations. Complex odontomes consist of a mass of enamel, dentine and cementum which is haphazardly arranged, whereas compound odontomes consist of a collection of numerous small discrete tooth-like structures[6]. All gradations between compound and complex odontomes exist, Most odontomes arise in association with a missing tooth. Radiographically, the compound odontomes show as a radiolucency with numerous small radio opaque denticles. These lesions are asymptomatic unless there is a secondary infection when pain becomes a feature of their presentations. Surgical enucleation is the treatment of choice. The condition does not recur and a radical approach is contraindicated .

Ameloblastoma is a benign, locally invasive tumour derived from the odontogenic epithelium. It accounts for about 1 percent of all oral tumours. About 8% of these tumors occur in the mandible, of which about 70% arise in the molar region and the ascending ramus. Ameloblastoma is a slowly growing tumor and is usually discovered in the fourth to fifth decade of life. Extension of the tumor in the soft tissues is a late feature. Radiologically ameloblastoma appears as a unilocular or a multilocular radiolucency. Although most ameloblastomas arise intra-osseously, extra oral or peripheral ameloblastomas have also been reported[7,8]. Metastases have been reported[9,10,11,12,13] but this is still an area of contro-

versy. Recurrence is a common feature in the multilocular type of ameloblastoma.

CONCLUSION

Central giant cell granuloma must be differentiated from brown tumours of hyper-parathyroidism by radio-immune assay of parathormone. It must not be confused with the giant cell tumour of the limbs which is an extensively destructive lesion. Central giant cell granuloma and compound, complex odontomes are benign, non recurrent entities, which are adequately treated by enucleation and curettage.. Ameloblastoma and dental keratocysts are both locally aggressive and exhibit a variable potential for recurrence. Segmental resection with preservation of the lower border of the mandible, wherever possible, minimizes the risk of recurrence . However, since loss of teeth may not be acceptable in younger patient is enucleation and curettage may be performed, despite the greater chances of recurrence. Use of cryosurgery after enucleation is recommended to minimize recurrence. Patients who have undergone removal of ameloblastoma and keratocyst must be monitored post operatively for several years. Where dental extractions, segmental or partial resection of the mandible have been carried out, occlusal harmony should be restored with bone grafting and osseus integrated implants. Minor facial asymmetry will resolve under muscle stress. Where occlusion is intact, orthognathic surgery may be considered as a second step to resolve remaining facial asymmetry. The advantages of an intra-oral sub mucoperiosteal flap is that it does not leave a facial scar and the morbidity is low. It also allows apecectomy as well as enucleation, curettage and/or segmental resection through the same approach.

REFERENCES

1. Katz JO, Underhill TE.Multilocular radiolucencies.Dent Clin North Am. 1994 38 (1) 63-81

2. Soames JV; Southam JC. Oral Pathology. Oxford Medical Publication, 1985. 237.

3. Potters BJ. Tiner BD. Central giant cell granuloma. Report of a case. Oral Surg Oral Med Oral Pathol. 1993 75 (3): 286 -9.

4. Horner K. Central giant cell granuloma. Clin Radio. 1989 40 (6): 622-6.

5. Pindborg JJ; Kramer IRH. Histopathological typing of odontogenic tumors, jaw cysts and allied lesions. 1971 Int Histological Classification of Tumors. No. 5 World Health Organization, Geneva.

6. Budnic SD. Compound and complex odontome. Oral Surg. Oral Med. Oral Pathol. 1976), 42, 501 - 6

7. Stevenson AR, Austin BW. A case of ameloblastoma presenting as an exophytic gingival lesion. J Peridontol. 1990 61 (6): 378-81.

8. Bucci E., Lo-Muzio L, Mignogna MD, de-Rossa-G. Peripheral ameloblastoma: Case report. Acta-stomatal-Belgi. 1992 89 (4); 267 - 9.

9. Laughlin EH. Matastasizing ameloblastoma. Cancer. 1989 1; 64 (3): 776-80.

10. Ramdas K, Jose-CC, Subhashini J. Chandi-SM. Viswanathan FR. Pulmonary matastasis of the mandible treated with cisplatinum, adriamycin, 9 cyclophosphamide. Cancer. 1990 61:66 (7): 1 475-9.

11. Philips SD, Corio RL, Brem H, Mattox D. Ameloblastoma of the mandible with the intracranial matastasis. A case study. Arch Otolaryngol Head Neck Surg. 1992 118 (8); 861-3.

12. Ueda M: Kosaki-K, Kaneda T; Imaizumi M, Abe-T. Doubling time of ameloblastoma metastasizing to the lungs. Report of two cases. J Craniomaxillofac-Surg. 1992 20 (7): 320-2.

13. Nagai N, Takeshita N, Nagasuka H, Innoue M, Nishijima K; etal . Ameloblastic carcinoma: Case report and review. J Oral Pathol Med. 1991 20 (9): 460-3.

14. Albuquerque K, Mehta S, A, Sarkar S Mehta-AR. Recurrent Amelo blastoma of the mandible and maxilla. Indian J Cancer. 1993 30 (2) 77-81.

15. Muller S, DeRose PB, Cohen C. DNA ploidy of ameloblastoma and ameloblastic carcinoma of the jaws. Analysis by Image and flow Cytometry. Arch Pathol Lab. Med. 1993 117 (11): 1126-31.

16. Thompson IO, Ferreire R, Van-Wyk CW. Recurrent unicystic ameloblastoma 07 the maxilla. Br J Oral Maxillofac Surg. 1993 31 (3) 3: 180-2.

A STUDY OF CASES OF LARGE INTRA-ABDOMINAL LYMPHANGIOMAS- A REVIEW OF LITERATURE

IMTIAZ LEWIS QURESHI, JAFFER-UZ-ZAMAN, AHMED SULTAN KHAN,
ABID QAZI, IFTIKHAR JAN, NAEEM-UZ-ZAFAR KHAN.

ABSTRACT:
Intra-abdominal lymphangiomas are rare lesions. In literature different diagnostic modalities are used in detection of early lesions. But our experience at the P.I.M.S. with intra-abdominal lymphangiomas has been totally different. All the 6 lesions in the last 8 years presented with enormous abdominal distention and a provisional diagnosis of ascites. Some of these were even tapped for ascitic fluid analysis. In most cases the final diagnosis was made only at laparotomy. Interesting features of age, sex, site of origin, type of lesion, and management are discussed in this article.

KEY WORDS:Abdominal neoplasms, lymphangioma, Diagnostic errors, ascites.

INTRODUCTION

Lymphangioma is a benign growth of lymphatic origin mostly noted at birth[1,3]. Whether these are congenital malformations, hamartomas or true neoplasms is still controversial[7,17]. 95%are seen in the neck, 2-3% in chest and the rest occur in other parts of the body[8]. Intra-abdominal lymphangiomas are rare. The incidence of these has been estimated to range from I in 27,400 to 1 in 100,000 of all hospital admissions[12]. This article discusses our experience with the management of this rare entity and the diagnostic confusion which we encountered during their management.

MATERIALS AND METHODS

The medical records of all the patients with proven diagnoses of intra-abdominal lymphangiomas, presenting to the Children's Hospital, P.I.M.S, Islamabad from January 1987 to December 1994 were reviewed retrospectively. All the cases were analyzed from the standpoint of age, sex, presenting symptoms, physical examination, investigations, site of lesion, management and outcome.

RESULTS

During 8 years (Jan. 87 - Dec. 94) we saw 6 cases of intra-abdominal lymphangiomas. The average age at the time of presentation was 5.3 years ranging from 3 to 9 years. 66.6% (n=4) were females. Two patients had omental lymphangiomas, 2 had mesenteric lymphangiomas (Figure 1), One had a retroperitoneal origin (Figure 2) while another arose from the greater curvature of the stomach (Figure 3) but was not attached to the omentum. All the patients presented with chronic abdominal distention, which in one case increased suddenly in size (case#l). In one patient it was also associated with colicky abdominal pain and occasional vomiting (Case#4). Every patient had good general health at the time of presentation. Each patient had a positive fluid thrill. The provisional diagnosis in 66.6% (n=4) patients was ascites based on the characteristic history and physical findings were inadvertently tapped in 2 cases and showed straw-colored fluid.

Fig. 1: Partly cystic partly solid multilobulated lymphangioma arising from the root of the mesentery extending from the fourth part of the duodenum to proximal jejunum (Case #4)

Prof. M. Naeem-uz-Zafar Khan
The Children's Hospital
PIMS, Islamabad

Fig 2: Multicystic lymphangioma densely incorporating the left kidney. The specimen also shows the kidney which had to be sacrificed.

Routine hematologic tests and LFTs in all 6 cases were within normal range. Ultrasonography revealed large intra-abdominal cystic masses in 4 cases which displaced the viscera peripherally but origin of these cysts could not be ascertained. In one patient the provisional diagnosis of multicystic kidney was entertained due to the presence of multicystic mass in the renal region on ultrasonography (Case #5). Definitive diagnosis in these cases could be made intraoperatively which also revealed their site of origin. The patient who presented with a sudden increase of abdominal girth had a lymphangioma filled with dark black colored fluid - an indication of hemorrhage into the lesion (Figure #I).

Fig 3: Intra-abdominal lymphangioma from case #1. This large, multilobulated, unilocular thin walled lesion arose from the greater curvature of stomach. It contained darkblack fluid- an indication of hemorrhage.

In one patient the abdominal cavity was full of turbid fluid with lymphangiomatous lesion on the omentum and peripheral surface of peritoneum in a large umbilical hernia (Case#6). These were the remnants of lymphangioma which actually were ruptured by needle punctures. These lymphangioma were excised after careful dissection from the surrounding tissues except the one which was retroperitoneal in origin (Case #5). In this case kidney had to be sacrificed because of the involvement of the vascular pedicle(Figures 2 & 4). The results are summarized in Table #1.

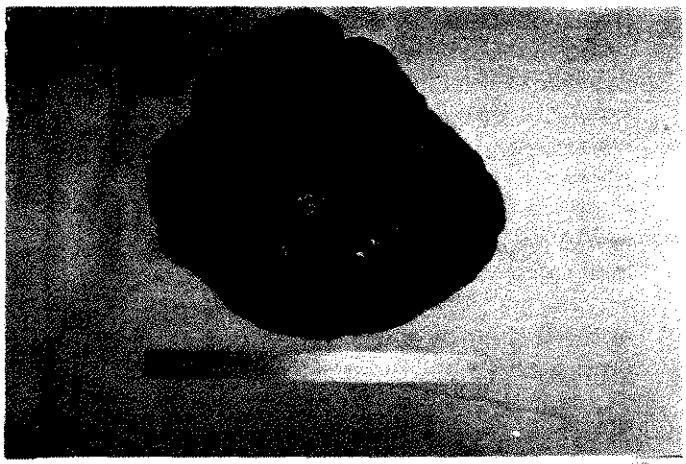


Fig. 4: Multicystic lymphangioma densely incorporating the left kidney. The specimen aslo shows the kidney which had to be sacrificed.

Table 1

No	Age	Sex	Symptoms	Provisional Diagnosis	site	procedure
1	5 Yrs.	M	Chronic Abdominal distention, with sudden increase	Ascites	Greater Omentum	Excision
2	3 Yrs.	F	Chronic Abdominal distention	Ascites (tapped)	Greater curvature stomach	Excision
3	7 Yrs.	F	Chronic Abdominal distention	Ascites	Mesentery	Excision
4	4 Yrs.	M	Chronic Abdominal distention	Mesenteric cyst	Mesentery	Excision
5	4 Mons	F	Loin Mass Kidney	Multicystic	Retroperitoneal	Excision & Nephrectomy
6	9 Yrs.	F	Chronic Abdominal distention	Ascites (tapped)	Omentum	Excision

Table 1: Results of management of intra-abdomianl lymangiomas

DISCUSSION

Among the intra-abdominal lymphangiomas it has been estimated that 60% are mesenteric, 25% are omental and 10% are mesocolic in origin, other abdominal sites are rare [11-14]. In children, it is reported to be the most frequent tumor of omentum[2]. Majority appear in the first year of life. 90% are evident by the second year of life[3]. Reports are also available in which it was detected at the age of 78 years[8]. In American population there is a 2:1 female to male ratio while in Japanese population the ratio is reported to be opposite[8]. Apart from these reviews no

sex predilection has been observed[11].

Numerous theories have been proposed concerning the origin of lymphangiomas. The important postulates are: disturbance of endothelial secretory function of the vessels, defects in embryonal organization of the lymphatic system, obstruction of existing lymphatic channels by an inflammatory process or a failure of lymphatic spaces in embryo to join the venous system[3,4,5,8,11]. Other less plausible etiologies include failure of the leaves of the mesentery to fuse, localized degeneration of lymph nodes or intra-abdominal trauma[11].

Lymphangiomas are classified primarily on a histological basis. The original classification established by Wegner in 1877 is still considered to be the most reliable. It divides lymphangiomas into simple, cavernous and cystic types[1,3].

These overgrowths are endothelium lined, round, smooth, thin walled and have clear serous or milky fluid within them. They may vary from small growth to a very large sized lesion. Sudden enlargement can be due to hemorrhage, vascular obstruction or a superimposed infection. These lymphangiomas may be uni or multilocular with communicating cysts[2-6]. Lymphangiomas are histologically benign but two patients with changes of lymphangio-endotheliomas have been reported[11].

Lymphangioma may vary in size from a few centimeters to a very large lesion containing several liters of fluid occupying the entire abdominal cavity and producing marked distention. In older children the lymphangiomas are generally of large size. This may be owing to the fact that they may have been growing for a number of years. They may be intra-mucosal, intramural or intraluminal and sessile and broad-based or pedunculated lesions. Generally there may be an abrupt transition between the lesion and adjacent normally displaced tissues. Generally lymphaniomas are solitary, but they may be multiple. When multiple lymphangiomas are present the term lymphangiomatosis is applied. Histological examination is necessary for diagnosis because they must be differentiated from angiomas[3]. In contrast to hemangiomas they usually do not have blood within the spaces and do not demonstrate immunoreactivity to factor VIII related antigen[4].

It is reported that the diagnosis is mostly incidental with the help of lymphangiography, during elective abdominal surgery, in an acute surgical abdomen or at post-mortem examination, but some are symptomatic[11]. The symptoms are nonspecific and depend on site and size of the lesion. The most frequent symptoms being pain, digestive problems, palpable soft cystic mass or even externally visible tumors such as pseudo-ascites. Most remain occult until pressure symptoms occur. It can lead to obstruction of tubular structures like intestine, ureter and bile-duct. Acute abdomen may occur due to inflammation of the cyst wall, bleeding, torsion or rupture of the cyst[15]. Galifer has reported that acute abdomen caused by lymphangiomatous cyst mimicked acute appendicitis[11]. Occasionally they may lead to intussusception. In rectal lymphangiomas the most common complaint is pain or bleeding[8].

Although it is rare for lymphangiomas to be huge in size but all our cases were noted in this category. At this stage imaging techniques help differentiate lymphangioma from ascites[6,18]. On a plain radiograph it may appear as rounded sharply outlined soft tissue mass displacing the adjacent bowel. This sign may be better appreciated in a gastrointestinal barium series. Calcification of the cyst wall may be occasionally noted. On ultrasound it appears as well outlined, sonolucent transonic abdominal mass. CT scan demonstrates a non-enhancing near water density mass[5,16]. They clinically and radiologically are difficult to distinguish from other intramural masses such as lipomas, leiomyomas or early epithelial polyp. Lymphangiomas are generally pliable and change shape in response to compression. This can be appreciable both endoscopically and radiologically. CT scan is reported to provide the exact topography[17]. MRI is reported to be superior to CT or U/S in diagnosing a large hemorrhagic abdominal lymphangioma which is reported to clinically mimic a recurrent hemoperitoneum[19]. Protein losing enteropathy has been reported to be associated with the lesion which can be cured by resection of the lesion[5,9,10].

Effective treatment is surgical removal. Regression is unlikely[3]. They invade tissue planes and appear to infiltrate normal tissues but are benign[8]. Aspiration of large cysts lessens the difficulty with exposure during operation. If the cyst has invaded an abdominal organ like small bowel, spleen or tail of pancreas, total excision of the involved organ is indicated[11]. Prognosis is good and it seldom recurs[15,16]. Internal and external marsupialization have become obsolete[11].

REFERENCES

1- James Lister, Irvene M. Irving: Neonatal surgery 3rd Ed, 1990, Buterworth, p.59.

2- Juan Rosai: Ackerman's Surgical Pathology, Vol.2 7th Ed, 1990 Mosby p.549.

3- Sabiston: Textbook of Surgery, 11th Ed. 1989 Saunders.

4- John H Davis: Clinical surgery, 1987 C.V. Mosby. Company p. 3077-78,1485.

5- Ronald. L. Eisenberg: Diagnostic Imaging in surgery, 1987, Mc-Graw Hill. p. 466,507.

6- Louis. P. Dehner: Pediatric surgical pathology, 2nd Ed, 1987 Williams & Wilkins. p. 77, 883-885.

7. Anderson's Pathology, Vol.1 8th Ed, 1985, Mosby. p. 724-726.

8. Frank. A. Oski: Principles and practice of paediatrics 1990, J.B. Lippincott. p. 388.

9. Fenoglio, Preiser, Lartz, Listrom, Davis,. Rilke: Gastrointestinal pathology - An atlas and text, 1989 Raven press p. 578-579.

10. Colin L Berry: Paedriatic pathology, 2nd Ed. 1989 Springer Verlag. p. 237.

11. I. Roisman, J. Manny, S. Fields, E. Shiloni: Intraabdominal lymphangioma. Br. J. Surg. 1989, Vol. 76. May, 485-9.

12. G. Silecchia, A. Materia, D. Surgo, A. Capua: Accidental successful treatment of a rare case of diffuse peritoneal lymphangiomatosis: Fr. J. Surg. 1994, 81, 1286-1289.

13. Walker AR, Putnam TC.: Omental, mesenteric, and retroperitoneal cysts: a clinical study of 33 new cases. Ann. Surg. 1973: 178: 13-19.

14. Henzel JH., Pories WJ, Burget DE, Smith JL.: Intraabdominal lymphangioma. Arch. Surg. 1966; 93: 304-8.

15. Morger R., Bollinger B., Muller M.: Abdominal lymphangioma in childhood. Paediatr. Pathol. 1991; 26(1): 31-4.

16. Schimidt M.: Intra-abdominal lymphangioma. Kans. Med. 1992 May; 93(5): 149-IS0.

17. Liessi G., Sandini F., Spaliviero B.: Cystic lymhangiomas of the abdomen. CT and US findings. Radiol. Med. Torino. 1989 Sep; 78 (3): 204-9.

18. Lugo Olivieri CH., Taylor GA.: CT differentiation of large abdominal lymphangioma from ascites. Pediatr. Radiol. 1993; 23(2): 129-130.

19. Stoupis C., Ros PR., Williams JL.: Hemorrhagic lymphangioma mimicking hemoperitoneum: MR imaging diagnosis. J. Magn. Reson. Imaging. 1993 May-June; 3(3): 541-2.

Sacrococcygeal Teratoma Presenting as a Benign Lesion in Older Age Group

JAFFER-UZ-ZAMAN, IMTIAZ LEWIS QURESHI, AHMAD SULTAN KHAN, NAEEM-UZ-ZAFAR KHAN

ABSTRACT

Although it is a known fact that high percentage of sacrococcygeal teratoma left untreated will undergo an early malignant transformation. But recently we have had an interesting experience with 3 cases of sacrococcygeal teratoma 2 of which were entirely untreated upto the ages of 4 and 6 years and third one was inappropriately managed, but did not undergo a malignant transformation and presented with obstructive bowel and urinary symptoms. Details of presenting symptoms, investigative findings and management is described. A review of literature is also presented.

KEYWORDS : sacrococcygeal teratoma, persistent benign nature, teratoma

INTRODUCTION

Sacrococcygeal teratoma is the most common congenital neoplasm of neonates.It is most commonly observed at birth, only a few cases have been observed in older children and adults[1, 3, 7]. The incidence varies from 1 in 10,000 to 1 in 40,000 live births[1, 2, 3, 4, 21]. There is a female preponderance with a ratio of 4:1 [1, 3]. It is reported to be autosomally dominant with a strong familial tendency [1, 4].

CASES

Case 1

A girl who had a previous surgery at the age of 8 days for acute urinary retention caused by intra pelvic cystic lesion, in another hospital, was admitted in October, 1994 at the age of three and a half years, once again with urinary retention. On examination she had a large cystic lower abdominal swelling which was presumed to be distended urinary bladder. She also had bilateral diffuse deep cystic swelling causing the buttock to bulge out. Ultrasound showed remarkably distended bladder occupying lower half of the abdomen. There was another cystic swelling of 10 cm. circumference deep in the pelvis which presumably had caused bladder neck obstruction resulting in urinary retention. She also had bilateral gross hydronephrosis. Her BUN was 55. IVU failed to secrete the dye. X-Ray chest and CT scan did not show any distant metastasis. Alpha feto protein was normal.

On exploration through lower transverse abdominal incision, a large cystic mass in the pelvis was noted (Figure 1). It had displaced the bladder up into the abdomen. This was excised and it contained whitish smell-less fluid and culture revealed no growth. The cyst had endothelial lining and was reported as mature cystic teratoma. Subsequently, this relieved her symptoms and when she was readmitted on 22 november 1994 her kidney and bladder had returned to normal size. Her BUN was 24. She was once again explored from behind after placing her in jack-knife position and rest of the multi-cystic teratoma along with the coccyx was removed. In addition to cyst there were solid areas containing cheesy material and loose hair. Once again histology revealed no evidence of malignancy.

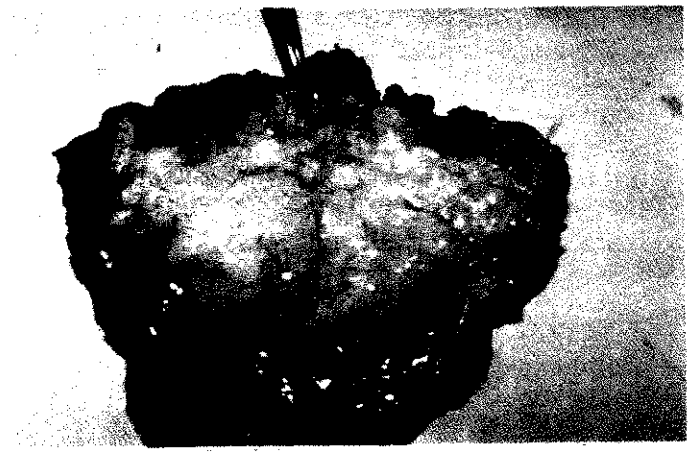


Fig.1: Specimen of Case #1 who had recurrence of inadequately removed sacrococcygeal teratoma and on each occasion presented with acute urinary retention. Artery forceps is pointing to removed coccyx which is an essential step in preventing recurrence

Prof. M. Naeem Khan, Chairman and Prof. of Paediatric Surgery
The Children Hospital,
PIMS, Islamabad

Case 2

4 years old female consulted us for the first time with massive swelling of the size of a football arising from the sacrococcygeal area (Figure 2 & 3). The swelling underwent repeated ulcerations and resulted in physical disability as she was unable to sit properly.On examination there was a broad mass of 25 cm. circumference with loss of gluteal folds and anterior displacement of anal canal. The overlying skin was very thick with multiple areas of ulceration and necrosis secondary to repeated trauma. The tumor was rubbery in consistency with areas suggestive of cystic lesions. Plain X-ray revealed patchy calcification. On barium enema there was evidence of intra pelvic presacral extension of tumor, but this extension was not palpable perabdominally. IVU was normal. X-ray chest and CT scan revealed no evidence of metastasis. Alpha feto protein was within normal limits indicating a benign lesion.At operation a firm large multicystic tumor with intense fibrosis fat and gluteal muscle was noted. This resulted in loss of tissue plains. There were scattered patchy calcifications of the cyst walls. With some difficulty it was excised in its entirety. Histopathology revealed tissues of ectodermal, endodermal and mesodermal origin. The tissue had cystic cavitations which were lined by chondroid endothelium and neural tissue was also present. No evidence of malignancy was found in multiple sections. In view of histopathology, prognosis may be considered good. Post operative progress was satisfactory and she was sent home. (Figure 4)

Fig 4: Patient managed conservatively following an esophageal perforation from attempted bouginage for dilatation. Note intercostal chest tube. mediastinal widening is significant due to edema fluid.

Fig. 3: Cut surface of the removed specimen of sacrococcygeal teratoma in Fig. 2. The increase in size was due to hemorrhage in the cystic part of mature teratoma

Fig 2: A splinter which had perforated through the first part of duodenum was lying half outside and half within the lumen. This was the content of ghutti given to the neonate by grandmother. It had caused peritonitis.

Case 3

A six years old boy consulted us for the first time with complaints of constipation and urinary retention for three months. He carried a barium enema with him which showed massive dilatation of colon and impacted fecal matter. His intravenous pyelogram showed normally functioning kidney with no distal urinary tract obstruction or hydronephrosis. With the provisional diagnosis of Hirschsprung disease he was taken to the operation room for rectal biopsy. Examination under anesthesia revealed a solid mass displacing the rectum anteriorly. The upper margin of the mass could not be reached with the finger tip. Examination also revealed bilaterally palpable masses in the glutei with gluteal asymmetry. Lateral view of the pelvis in barium enema showed a pre-sacral mass displacing the rectum anteriorly (Figure 5) while ultrasound showed a 10cms X 8cms solid mass in the pelvis. At operation a large solid mass arising from the coccyx and the last segment of sacrum was found plastered to the rectum. It was, with some difficulty, removed and coccygectomy was done. Histopathology revealed a benign sacrococcygeal teratoma containing neural tissue.

Fig.5: Lateral view of pelvis of barium enema shows marked anterior displacement of the rectum which is occupied by a presacral intrapelvic solid mature sacracoccygeal teratoma with predominantly neural tissue in a 6 years old child (Case 3)

DISCUSSION

Usually sacrococcygeal teratoma presents as a bulging mass over the sacrococcygeal region with variable cystic to solid consistency [1,6,12]. Histologically elements of all three germinal layers may be found[6, 12]. Most sacrococcygeal teratoma have malignant potential and with increasing age tendency of malignant transformation increases proportionally. In neonates frequency of malignancy is approximately 7% whereas above the age of two years it rises to 50% or more [1,11]. In an interesting case a 67 years old male presented with sacrococcygeal teratoma complicated by ulceration and necrosis, histopathology showed benign nature of tumor [12, 13]. Metastasis of malignant tumor may occur in regional lymph nodes, lungs, liver and vertebrae [14]. These tumors are being diagnosed more frequently in utero due to the widespread use of obstetric sonography. In fetal life it may cause hypertrophy of placenta, fetal hydrops, high output cardiac failure, pre-eclempsia and respiratory insufficiency [5, 9, 10, 15, 24]. A baby may be born with bilateral hydronephrosis, abdominal distention or failure to pass meconium [4]. It may also cause abortion or still-birth [18]. Associated abnormalities occur in 20% of the cases, the most common being musculoskeletal, renal, cardiac, gastrointestinal, central nervous system, spina bifida, undescended testis and congenital hip dislocation [1, 2, 3, 17, 24]. Tumor with large presacral component may present with weakness or paralysis of legs, constipation, anal stenosis [25] and urinary symptoms secondary to bladder outlet obstruction [4]. Sacrococcygeal teratoma in females is often associated with agenesis of corpus collasum and arachnoid cyst. Males born with sacrococcygeal teratoma may have leydig cell dysfunction, abnormal spermatogenesis or both [19,22, 16, 30].

Routine ante-natal ultrasound examination helps not only in diagnosing the sacrococcygeal teratoma but also determines secondary uropathy and helps in planning the mode of delivery [2]. Calcifications, which may be diffuse or in the form of a recognizable structure may be a helpful radiological sign but do not help differentiate benign from malignant lesions. N- acetyl cholinesterase [23] has also been measured for diagnosing but the most useful marker is alpha-feto protein, which differentiates between benign and malignant tumors and can be used as a prognostic factor as well as a sign of recurrence [1, 20, 25].

In all cases of this tumor, radical attempt should be made to remove the tumor regardless of the size of the tumor and histological stage, as soon as possible after birth. Unusual delay in surgical treatment may be associated with malignant degeneration, pressure necrosis, infection or hemorrhage in the tumor. Coccygectomy is mandatory to ensure against recurrence [25]. Incision in this operation is very crucial. An inverted V-shaped incision has been found to be very important in maintaining the shape of the gluteal fold (Figure 4) and reconstitution of the anorectal sling. Prognosis depends on type, extent, histology and age at the time of surgery, but interestingly not the size of the tumor.

Malignant transformation is most likely to occur between the age of 4 months and 5 years. However, a few cases have been reported in later ages without malignant change.

REFERENCES

1. James Lister, Irvene M. Irving: Neonatal surgery.3rd Edition.1990. Butterworth. p 142- 52.

2. M. Feldman, P. Byrne, M. A. Johnson, J. Fischer, and . G. Lees: Neonatal Sacrococygeal Teratoma: Multimoding Assessment. Journal of Pediatric Surgery, Vol.25, No.6 (June), 1990: pp 675-678.

3. P.S. Malone, L. Spitz, E. M. Kiely, R. J. Brereton, P.G. Duffy, and P.G. Ransley: The Functional Sequelae of Sacrococcygeal Teratoma. Journal of Pediatric Surgery, Vo.25, No.6 (June), 1990: pp 679-680.

4. Roberta E. Sonnino, Shirley Chou, and Frank M. Guttman: Hereditary Sacrococcygeal Teratomas. Journal of Pediatric Surgery, Vol.24, No.10 (October), 1989: pp 1074-75.

5. Sheldon J. Bond, Michael R. Harrison, Klaus G. Schmidt, Norman H. Silverman, Alan W. Flake, R. Nathan Slotnick, Robert L. Anderson, Steven L. Warsof, and Donald C. Dyson: Death Due to High-Output Cardiac Failure in Fetal Sacrococcygeal Teratoma. Journal of Pediatric Surgery, Vol.25, No.12 (December), 1990:pp 1287-1291.

6. Havranek-P, Hedlund-H, Rubenson-A, Guth D., Husberg M., Frykberg T., and Larsson L.T.: Sacrococcygeal Teratoma in Sweden between 1978 and 1989: long-term functinal results. J. Pediatr-Surg. 1992 Jul; 27(7): 916-8.

7. Bloechle M., Bollmann R., Zienert A., Kalache K., Korner H., Bartho S. Specht-U: Fetal teratoma diagnosis and management. Zentralbl-Gynakol. 1992; 114(4): 175-80.

8. Drut R. M.; Drut R., Fontana A., Grosso J.J.: Mature Presacral Sacrococcygeal Teratoma associated with a sacral epignathus. Pediatr. Pathol. 1992 Jan-Feb: 12(1): 99-103.

9. Nakayama D.K., Killian A., Hill L.M., Miller J.P., Hannakan C, Lloyd D.A., Rowe M.I. The new born with hydrops and sacroccocygeal teratoma. J. Pediatr. Surg. 1991 (Dec; 26(12): 1435-8.

10. Alter D.N., Reed K.L., Marx G.R., Anderson C.F., Shenker L.: Prenatal diagnosis of congestive heart failure in a fetus with a sacrococcygeal teratoma. Obstet. Gynecol. 1988 Jun; 71(6 Pt 2) 978-81.

11. West D.K., Touloukian R.J.: Meconium pseudocyst presenting as a buttock mass. J.Pediatr. Surg. Ig88 Sep; 23(9): 864-5.

12. Harbon S. and Pheline Y.: Giant sacrococcygeal teratoma in adults. Ann-Chir-Plast-Esthet. 1989; 34(2): 153-9.

13. Langer J.C., Harrison M.R., Schmidt K.G., Silverman N.H., Anderson R.L., Goldberg J.D., Filly R.A., Crombleholme T.M., Longaker M.T., and Golbus M.S.: Fetal hydrops and death from sacrooccygeal teratoma: rationale for fetal surgery [see comments] Am. J. Obstet. Gynecol. 1989 May; 160(5 Pt 1): 1145-50.

14. Ouimet A., and Russo P.: Fetus in fetu or not? [see comments]. J. Pediatr. Surg. 1989 Sep; 24(9): 926-7.

15. Roig J.; Aguilar X. Maestre J. Domingo C. Morera J.: Pulmonary mass and air meniscus sign in a 19-year-old man. Chest. 1991 Aug; 100(2): 562-3.

16. Lahdenne P; Dunkel L, Heikinheimo M, Koskimies Al, Siimes M.A.: Hypergonadotropic hypogonadism and sperm abnormalities in men born with benign sacrooccygeal teratoma. J. Androl. 1991 Jul-Aug; 12(4): 226-30.

17. Lahdenne P, Heikinnheimo M, Jaaskelainen J, Merikanto J, Heikkila J, Siimes M.A.: Vertebral ahnormalities associated with congenital sacrococygeal teratomas.: J. Pediatr. Orthop. 1991 Sep-Oct; 115): 603-7.

18. Ikeda H., and Sasano N.: Demonstration of pituitary tissue with 6 cells immunoreactive to pituitary hormones in a sacrococcygeal teratoma. Acta. Pathol. Jph. 1987 Jan; 37(1): 117-22.

19. Borges A.C., Festugato R.R., de-Cerqueira J.C., de-Souza M.R.: Sacral agenesis associated with a teratoma Arq. Neuropsiquiatr. 1987 Mar; 45(1): 81-7.

20. Peterson C.U., Strickler J.G., Gonzalez R. Dehner L.P.: Uterus like mass of the small intestine. Heterotopia or monodermal teratoma? Am. J. Surg. Pathol. 1990 Apr; 14(4): 390-4.

21. El-Qarmalawi, M.A., Sadik M. el-Abdel. Hadi F., Muwaffi. R; Nageeb K.: Diagnosis and management of fetal sacrococcygeal teratoma. Int. J. Gynaecol. S\Obstet. 1990 Mar; 31(3): 275-81.

22. Shinoura N., Kondo T., Muraoka I., Tsukamoto Y, Yoshioka M, Miyazawa H. :Sacrococcygeal teratoma of new born infants; a report of two cases. No.Shinkei. Geka. 1990 Jun; 18(6): 557 61.

23. Kainer F. Winter R.; Hofmann. H.M. Karpf. E.F.: Sacrococcygeal teratoma. Prenatal diagnosis and prognosis. Zentralbl. Gynakol. 1990; 112(10): 609-16.

24. Book B, Ries R, Wunsch. P.H., Feige A. : Prenatal diagnosis of a sacrococcygeal teratoma with hydrops fetalis and placental hypertrophy—consequences for the further course of pregnancy. Gaburtshilfe. Frauenheilkd. 1990 Aug. \50(8): 647 9.

25. Dominici C. Bosman C. De-Sanctis-S; Clerico. A. Thorel M.F. Ceccamea A. : Endodermal sinus tumor. Histological changes induced by chemotherapy. Minerva. Pediatr. 1990 Jun. 42(6): 249-52.

REVIEW ARTICLE

MINIMALLY INVASIVE SURGERY

IRSHAD WAHEED, KULSOOM JUNEJO

KEYWORDS : Minimally invasive surgery, laparoscopy

INTRODUCTION

Endoscopic surgery is rapidly becoming a popular alternative to open procedures. Its introduction in the field of general surgery has changed the speciality forever. The Laparoscope, once used by gynaecologists for evaluation of pelvic disorders has emerged as a leading therapeutic tool for various surgical procedures. The most widely practiced amongst them is the laparoscopic cholecystectomy, considered as a "gold standard" treatment for uncomplicated symptomatic cholelithiasis. Others include laparoscopic appendicectomy, hernia repair, thoracoscopic treatment of pneumothorax, persistent pleural effusion, oesophageal disorders, sympathectomy, arthroscopic examination and joint repairs. Various urological and gynaecological procedures have all become accepted endoscopic procedures.

HISTORY

Lapara is a Greek word meaning fluent. Laparoscopy is endoscopic visualization of peritoneal cavity. The earliest description of endoscopic examination was from KOS school led by Hippocrates (460-375 BC), the father of modern Laparoscopy . The one who introduced this was Jacobeus. He used a cystoscope for examination of both peritoneal and pleural cavities in 1910 and used both air and water for distention of cavities. Later on the procedure underwent several modifications as Zollikofer in 1925 introduced the concept of CO2 for insufflation.

ADVANTAGES

Laparoscopy has revolutionized general surgery. The increasing demand for minimally invasive surgery is being driven by a combination of developments in surgical instrumentation and patients' expectations of less pain, shorter hospital stay, improved cosmesis and early return to normal activity.

Prof. of Surgery
Jinnah Postgraduate Medical Centre
Karachi.

The advantages of Laparscopy are:

- Small incisions,minimal trauma, reduced post-operative pain, early moblization and virtual absence of wound infection.
- Minimum handling of gut and rapid return of G.I.T. functions, therefore less chances of developing future adhesions.
- Smaller wound size causes less depression of cell mediated immunity.
- Surgeons contact with patients' blood is decreased therfore,the chances of HBV & HIV transmission is correspondingly decreased.

DISADVANTAGES

- Image quality is inferior to normal binocular vision.
- Loss of depth perception.
- Reduced field of vision.
- Improper location of surgeon's eyes & hands relative to visual display,causing misperception of angular relationship.
- Minimal access causes organ extraction difficult.

The operating time cannot be ignored because of less experience and setting up of instruments etc.; operating time may be prolonged. Therefore inspite of shorter hospital stay, the total hospital cost is not reduced because theatre is in use for a longer time and the instruments, especially the disposables, are quite expensive.

INSTRUMENTS AND PROCEDURE

It requires full video endoscopic facilities including provision of camera, television monitor and insufflation equipment. Instruments required are needle, trocar & cannula, sheath, specialized clamps, clip applicators, retractors, staplers, scissors & specialized instruments for specific procedures.

Laparoscopy is carried out in operation theatre under general anaesthesia, observing full aseptic precautions. A pre-operative consent for conversion to open procedure should always be taken. Initally pneuoperi-

toneum is created. Then laparoscope is inserted through a sub-umbilcal port and initial laparascopy performed. The site of placement of ports is different in various procedures. It is ideal to have enough ports to allow the surgeon to use two handed dissection to avoid clashing of instruments, ports should not be too close to each other.

COMPLICATIONS

Experience in laparoscopic surgery is increasing. Success depends upon skill and experience.A sharp learning curve exists for first 25-50 cases and is characterized by longer operative time and risk of complications . But there is good evidence to suggest that with adequate training and experience, the incidence of operative complications will be reduced to a level comparable to open surgery.

Complications due to introduction of instruments and pneumoperitoneum include:

- Emphysema.
- Gas embolism.
- Thromo-embolic complications.
- Haemorrhage.
- Haemotoma.
- Intestinal perforation.
- Hyper-capnia leading to respiratory or circulatory embrassment.
- Hernia at puncture site,recurrence of malignancy at port site.
- Aggravation of existing hernia.

Due to the operative procedure

- Diathermy injury of skin and abdominal wall.
- Diathermy injury of intestine .
- Bile duct injuries.
- Haemorrhage.
- Pelvic sepsis
- Peritonitis.
- Trauma to retroperitoneal structures

CONTRAINDICATION

- Cardiac or respiratory insufficiency.
- Extreme obesity
- Advanced pregnancy or large abdominal tumors.
- Previous surgery (conversion rate is increased).
- Intestinal obstruction.
- Generalized pertonitis.

Most of the contraindications have become only relative ones as the experience and the confidence of the surgeons increases. Thus, acute cholecystitis is no longer an absolute contraindication.

FUTURE

There is no doubt that laparoscopic surgery is here to stay, it has been through three evolutionary steps:

1. Diagnosis
2. Minor surgical techniques as biopsy.
3. Removal of large tissue volumes as nephrectomy or splenectomy. The currently developing fourth step is the laparoscopic reconstruction.

The future of laparoscopic surgery should be directed towards identifying limitations, eliminating source of operating errors, improvement in skill and experience and improving existing technology and also to make it economically justifiable. The emphasis should be on the evolution of proper, accredited training programs in teaching hospitals.

REFERENCES

1. Farquharson's Text book of operative surgery - 8th edition R.F. Rintoul Minimal access surgery : 353 - 360.
2. General Surgical Operations 3rd edition R.M.Kirk Minimal access surgery : 167 -174.
3. Laparoscopy in Urology 1st edition 1994. Malcolm.J.Coptcoat and Adrain D.Joyce. History of Laparoscopy :5 -9.
4. Shaw's text book of operative gynaecology 5th edition Endoscopy in gynaecology : 37 - 45.
5. Maingots Abdominal operation 8th edition Seymour Schawartz Laparoscopy : 241 -245.
6. W.A .Kmoit and S.D. Wexner 1995 Laparoscopy in Colorectal surgery: a call for carefull appraisal Br.J.Surg 82: 25 - 26.
7. J.R.T.Monson, A.D.K. Hill and A.Darzi 1995 Laparoscopic Colonic surgery . Br.J.Surg.82 : 150 - 157.
8. M.S.Wilson, G.T. Deans and W.A. Brough Prospective trail Lichtenstein with laparoscopic tension free mesh repair of inguinal hernia Br.J.Surg.82 : 274 - 277.
9. S.O.Wexner and S.M. Cohen Port site metastases after laproscopic colorectal surgery Br.J.Surg 82 : 295 - 298.
10. A.J.Mc Mohan, G.Fullarton, J.N.Baxter and P.J.O. Dwyer Bile duct injury and bile leakage in laparoscopic Br.J.Surg . 82: 307 - 313.
11. J.P.Griffith, N.J.Everitt, F.Lancaster, A. Boylston, S.J. Richards, C.S.Scotl, E.A. Benson, H.M. Sue-ling Conventional cholecystectomy upon cell mediated immunity Br.J.Surg. 82: 677 - 680.
12. G. Crosthwaite, T. Chung, P.Dunkley, S.Shimi and A. Cuschiexi Comparison of direct vision and electronic two and three dimensional display system on surgical task efficiency in endoscopic surgery Br.J. Surg. 82 : 849 - 851.
13. A.S. Cole, M.N.Vipond and H.J.Espiver. Technique for closure of port site under laparscopic visual control Br.J.Surg. 82: 1134 -1135.

LIPOSARCOMA OF CLAVICLE

A Case Report

JAMSHED AKHTAR, FARHAT MIRZA, ABDUL AZIZ

ABSTRACT

Liposarcoma rarely arises in bone but when it does it almost invariably occurs in long bones of extremities.A case of Liposarcoma arising in clavicle is reported. To our knowledge liposarcoma of clavicle is never reported in literature.

KEY WORDS: Liposarcoma, Bone tumors.

INTRODUCTION

Liposarcoma is one of the most common malignant soft tissue tumors in adults but liposarcoma arising in bone is extremely uncommon[1]. Although some pathologists have doubted the very existance of primary intraosseous liposarcoma, a number of acceptable cases have been reported[2].

CASE REPORT

A nine year old female child presented with a swelling over left clavicular region which was noticed three months earlier. History of dull ache at the same site was also given. On physical examination a hard bony swelling in the line of clavicle was found. Skin over clavicle was normal. Radiologically, a well defined expanding osteolytic lesion with thinned out cortex in the lateral half of the left clavicle was found (Fig. I) Soft tissue invasion was absent. No swelling was present in other parts of the body. Lesion was resected. There was no local infiltration. Grossly, the tumor was greyish yellow in colour and soft in consistency. Microscopically, most of the cells had vacuoles in their cytoplasm. Areas of myxoid degeneration were also present. No futher treatment was given. Fifteen months postoperatively patient is well with no recurrence.

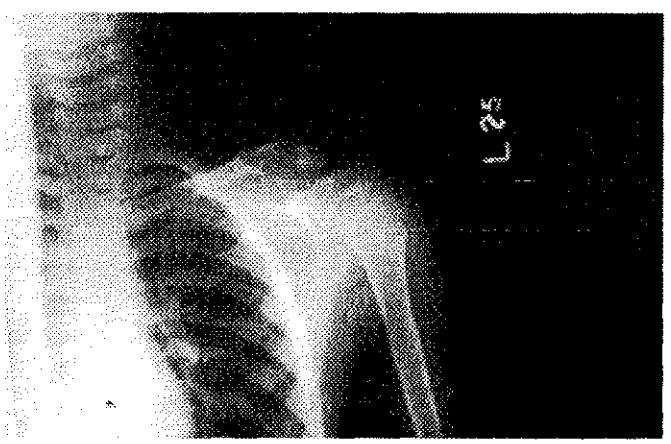


Fig. I Liposarcoma of Clavicle

Dr. Jamshad Akthar
Paediatric surgeon
Dept. of Paediatric Surgery, NICH

DISCUSSION

Stewart, one of the first to report liposarcoma of bone, discribed three cases in 1931[3]. The tumor is slightly more common in males and may be seen in patients of all ages. Most common site of involvement is long tubular bones of the extremities. Femur and tibia are commonly involved[4]. Schwartz collected fourteen cases of proven liposarcoma from literature[3]. To establish a firm disgnosis of an intraosseous liposarcoma two criteria must be met, the tumor must have the histologic features of liposarcoma and a soft tissue origin or metastatic neoplasm must be excluded[4]. Our case fulfills both the criteria. It is also unique as to its site, the clavicle, which is never reported previously.

REFERENCES

1 Carnesale PGH Malignant tumors of bone in Crenshaw AH. Campbell's operative orthopedic. 7th ed. Vol. II, chap. 33,C.V. Mosby company. 1987; 777-805

2 Ross CF, Chertsey GH. Primary osteo-liposarcoma of bone (Malignant mesenchymoma). Report of a case. J.Bone joint Surg. 1968; 50 B, 639-643

3 Schwartz A., Shuster M., Becker SM. et al. Liposarcoma of bone. Report of a case and review of the literature. J. Bone joint Surg. 1970; 52-A, 171-177

4 Resnick D. tumors and tumor like diseases in Resnick D. Niwayama G. (ed) Diagnosis aof bone and joint disorders. 2nd. ed. vol. 6, chap. 91. W.B. Saunder Co. 1988: 3602-4011.

TUBERCULOSIS OF PENIS

A Case Report

ABDUL SATTAR MEMON, JAN MOHAMMAD MEMON, ABDUL GHANI SOOMRO

ABSTRACT

A rare case of tuberculosis of penis in a man of 50 years is presented, followed by review of literature.

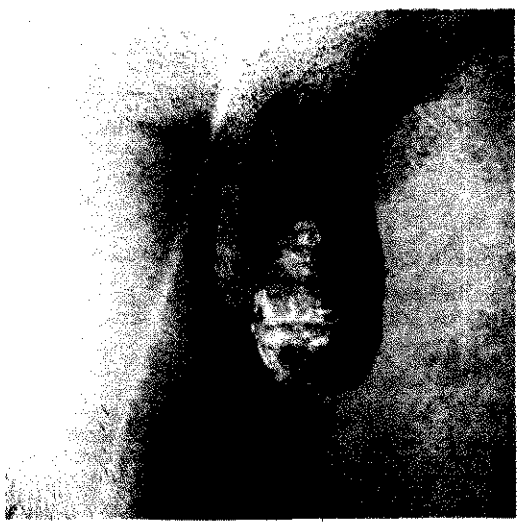
KEY WORDS: Penis, tuberculosis, genito-urinary tuberculosis, tuberculosis of the penis.

INTRODUCTION

Tuberculosis of penis is a very rare manifestation of the disease; upto 1971 only 139 cases had been reported in the literature[4]. Quite a long ago it was seen as a complication of ritual circumcision, when the operator who had open pulmonary tuberculosis used to suck the circumcised penis[5]. Presently tuberculosis of the penis is essentially a disease of adults, primary or secondary[3].

CASE REPORT

A 50 years old male from rural area of Sindh presented with ulceration of glans penis, burning micturation and pain with low grade fever for 8 months. Examination revealed pale underweight male of average build; no other positive finding was present on general examination. Systemic examination did not reveal any positive finding. Local examination of external genitalia revealed ulceration of glans (circumcised) penis, resembling carcinoma penis. (photograph I).



Photograph I

Photograph II

Urine DR was clear, Hb 9 g% ESR 105/hr, X-ray chest revealed bilateral foci of pulmonary tuberculosis in both lungs and sputum was positive for AFB. Incisional biopsy under general anaesthesia revealed it as a typical tuberculous ulcer and the patient was put on antituberculous drugs. He responded well to treatment and the ulcer healed (photograph II). At present the patient is completely cured.

DISCUSSION

Tuberculosis is a common disease of the developing countries, however tuberculosis of penis is an extremely rare manifestation of the disease. Presently tuberculosis of penis is a disease of adults either primary from coital contact from disease already present in the genital tract or due to contamination from infected clothing[1-6]. Rarely penile lesion may be caused by re-inoculation from male partner through an infected ejaculate[2]. Secondary penile tuberculosis occurs as a secondary manifestation from active pulmonary tuberculosis.

Dr. Abdul Sattar Memon
Assoc. Professor Surgery
Liaquat Medical College, Jamshoro

The lesion always presents as a superficial ulcer of glans, which clinically is indistinguishable from malignancy. The lesion can also progress to cavernositis involving the urethra[8]. Rarely lesion occurs as a solitary nodule or as a cavernositis with ulceration[2-7]. The diagnosis is confirmed by biopsy and responds well to antituberculous chemotherapy.

REFERENCES

1. Agarwalla, B, Mohanty, G.P., Satu, L.K. and Rath, R.C., Tuberculosis of the penis-report of two cases J. Urol., 124: 927, 1980.
2. Baskin NS and MEC. Tuberculosis of penis presenting as subcutaneous nodule. J. Urol, 141; 1430, 1989.
3. Gow, J.G., Genito-urinary tuberculosis. Cambles Urology 1992, 6th ed, Vol:I chap:22 Philadelphia W.B Saunders Company Scientific publications, 1976.
4. Lal, M.M., Sekhon, G.S., and Dhall, J.C., Tuberculosis of the penis, J. Ind, Med. Assoc., 65: 316, 1971.
5. Lewis, E.L. Tuberculosis of the penis. A report of 5 new cases and a complete review of the literature. J. Urol., 56: 737, 1946.
6. Narayana, A.S., Kelly, D.G., and Duff, F.A., Tuberculosis of the penis. Br. J. Urol., 48(4); 274, 1976.
7. Ramesh V; and Vasanthi R. Tuberculous Cavarnositis of the penis, genito urin. MED, 55: 56, 1939.
8. Veukataramaiah, N.R. Van Raalte, J.A., and Dutla, S.N.; Tuberculous ulcer of the penis Postgrad, Med J., 58: 59, 1982.

PHOCOMELIA

A Case report

IMTIAZ LEWIS QURESHI, UZMA FARHEEN HASHIM, ZIA-UL-HAQUE, NAEEM-UZ-ZAFAR KHAN

ABSTRACT:
An interesting case of Tetraphocomelia is presented, which highlights the importance of regular antenatal monitoring of expectant mothers. In addition, the judicious use of drugs during pregnancy, with an appropriate knowledge of teratogenic effects that they may produce, is advocated.

KEYWORDS: Tetraphocomelia, antenatal monitoring, teratogenic drugs, phocomelia.

INTRODUCTION

Phocomelia is derived from two Greek words "probe" meaning seal and "melos" meaning limb[1]. This deformity is a severe form of transverse limb deficiency in which there is a proximodistal developmental failure. This rare congenital anomaly can involve the upper or lower extremities or both[2]. Three types of phocomelia are identified: complete, proximal, and distal. In complete phocomelia, the upper extremity proximal to the hand is absent, and the hand articulates with the glenoid. Patients with proximal phocomelia have no upper arm, and the hand and forearm articulate with the glenoid. In distal phocomelia, the forearm is absent, and the hand articulates with the arm[2, 3].Similar pattern of deficiencies may be present in the lower limbs.

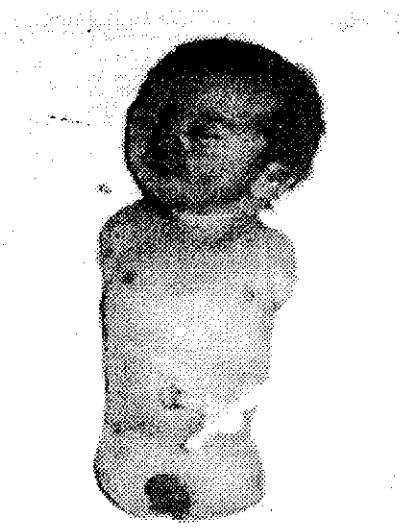


Fig. 1

A Child with Tetraphocomelia

Prof. M. Naeem-uz-Zafar Khan
Head Paediatric Surgery
Children's Hos[otal, PIMS, Islamabad

CASE

The baby boy was a product of non-consanguineous marriage, born to a twenty-eight years old G5 4+0 healthy mother after a full term pregnancy by spontaneous vaginal delivery with no complications.The mother did not undergo any ante-natal monitoring and there was no history of drug ingestion during the ante-natal period. The baby had absence of all the four limbs (Tetraphocomelia) except for a vestigial proximal hand bilaterally [Figure I].The lower limbs were entirely absent, including the digits. There was no other detectable abnormality including the cardiovascular system. With no facilities for special limb fittings, the parents were *"reassured"* and the patient was sent home and was lost to follow-up.

DISCUSSION

Birch-Jensen estimated that it occurred once in about 75000 births[1]. During the late 1950s and early 1960s, phocomelia became prevalent in Europe as a result of maternal thalidomide ingestion, when it affected 60% of the infants born to mothers taking thalidomide in first trimester[1]. However, cases can also occur sporadically[1].

In Great Britain, "thalidomide babies" often had besides phocomelia, preaxial defects that included radial club hand or hypoplasia of the radius, thumb, and index finger. Patients in this group had an ulna, and the humerus was normal, hypoplastic, or absent. Children with proximal agenesis of the humerus also had absence of the shoulder joint. Thalidomide also affected other systems and has been reported to cause depression of the nasal bridge, coloboma, microtia, deafness, hyperhidrosis, and cryptorchidism[2].
Phocomelia may be associated with other congenital

defects [13] as well as syndromes. It is a recognized feature of Robert-SC phocomelia, [6, 7, 8, 10, 19], Holt Oram[4, 5], Schinzel Phocomelia[11] and Acrofacial dysostoses syndromes [9]. Cardiac abnormalities are frequent and may be due to the fact that cardiac structures are formed at the same time that the limb structures are formed in embryo [1].

Etiologically, role of teratogen like thalidomide[1, 2 3], Valproic Acid[15] and Dicyclomine[16] has been well-established while experimental work on mice has postulated retinoic acid[17], and Cadmium Chloride[14] to be of teratogenic nature.

Antenatal routine ultrasonography can reliably detect phocomelia prenatally[6, 7]. It is also very important to identify associated abnormalities or syndromes because in cases of Robert-SC phocomelia Syndrome early neonatal death can be expected. Other syndromes featuring phocomelia may have much longer survival[6]. Tetraphocomelia has been detected as early as 17 weeks of intrauterine life[7].

Surgical treatment of phocomelia is limited[1, 2]. Most surgeries performed in these patients are designed to aid prosthetic fittings [1]. Decisions must be adapted to the total disability of the child. Thus, if one arm is normal, the dysplastic limb may be more valuable as it acts as a holder, and steadier than any contemporary prosthesis. Unfortunately, however, phocomelia is usually bilateral; but even then we should remember the adaptive capacity of the legs if they are normal[18]. Digital function may be improved by web space deepening or by correction of contracture that may facilitate the use of prosthetic devices[1]. Developments in prosthetics are now so rapid that these children should be referred to special centers within the first 3 months. A recent development is the use of myoelectric prosthetics in the upper limb[18].

REFERENCES

1. Lovell & Winter: Paediatric Orthopedics. 2nd Ed., Vol.2, 1986. J.B. Lippincott Company. p. 651-2.
2. F. William Bora, Jr.: The Paediatric Upper Extremity: Diagnosis and Management. 1986. W.B. Saunders Company. p.28.
3. Tachidjian: Pediatric Orthopedics. 2nd Ed., Vol 1, 1990, W.B. Saunders Company, p.III.
4. Dupre. C.T., Fincher. R.H.: Holt-Oram Syndrome associated with hypoplastic per pheral vasculature and midsystolic click. South. Med. J. 1993, Apr; 86(4): 453-6.
5. Pfeiffer-R.A.; Bowing-B; Deeg-K.H.: [Variants of radial hemimelia with and without vitium cordis (Holt-Oram Syndrome) in 2 families]. Monotsschr-Kinderheilkd. 1989 May, 135(5): 275-9.
6. Grundy-HO, Burlbaw-J, Walton-S, Dannai-C: Roberts Syndrome: Antenatal Ultrasound - A Case Report. J-Perinat-Med. 1988; 16(1): 71-5.
7. Robins-DB, LaddarRL, Thieme-GA, Boal-DK, Emanuel-BS, Zackai EH: Prenatal detection of Roberts-SC phocomelia syndrome: Report of 2 sibs with characteristic manifestations. Am-J-Med Genet. 1989 Mar;j32(3): 390-4.
8. Verloes-A, Herens-C, Van-Haldergem-L, Retz-MC, Dodinval-P: Roberts-SC Phocomelia Syndrome with exencephaly. Ann-Genet. 1989; 32(3): 169-70.
9. Le-Merrer-M, Cikuli-M, Ribier-J, Briard-ML: Acrofacial dysostoses. Am-J-Med-Genet. 1989 Jul; 33(3): 318-22.
10. Midro-AT,Iwaszkiewicz-Pawbwska-A, Puchnarewicz-A, Debeck-K: [TAR Syndrome (Congenital Thrombocytopenia and aplasia of the radial bones) in a 5-months-old boy]. Wiad-Lek. 1992 Jan; 45(1-2): 66-9.
11. Chitayat-D, Stalker-HJ, Vekemans-M, Delneste-D, Azouz-EM: Phocomelia, Oligodactyly, and Acrania: the Schinzel-Phocomelia Syndrome Am-J-Med-Genet. 1993 Feb 1; 45(3): 297-9.
12. Schianda-Stumpel-C, De-Die-Smulders-C, Fryns-JP, da-Costa-J, Bouckaert-P: Limb reduction defects and renal dysplasia: Confirmation of a new, apparently lethal, autosomal recessive MCA syndrome. Am-J-Med-Genet. 1990 Sept: 37(1): 133-5.
13. Schinzel-A: Phocomelia and additional anomalies in two sisters. Hum-Genet. 1990 May; 84(6): 539-41.
14. Padmanabhan-R; Hameed-MS: Characteristics of the limb malformations induced by maternal exposure to cadmium in the mouse. Reprod-Toxical. 1990; 4(4): 291-309.
15. Verloes-A, Friklche-A, Gremillet-C, Paquay-T, Decortis-T, Pigo-J, Senterre J: Proximal phocomelia and radial ray aplasia in fetal valproic syndrome. Eiu-J-Pediatr. 1990 Jan; 159(4): 266-7.
16. Correy-JF, Newman-NM, Collens-JA, Burroross-EA, Burrows-RF, Curron-JI. Use of Prescription drugs in the first trimester and congenital malformations.Aust-N-Z-J-Obstet-Gynaecol. 1991 Nov; 31(4): 340-4.

17. Dolfe-P, Ruberte-E, Kastner-P, Petkovick-M, Stoner-CM, Gudas LJ, Chambon-P: Differential expression of genes denoting alpha, beta and gamma retinoic acid receptors and RABP in the developing limbs of the mouse. Nature. 1989 Dec. 7; 342 (6256): 702-5.

18. G.C. Llyod Roberts and J. Fixsen: Orthopedics in Infancy and Childhood. 2nd Ed., 1990. Butterworth-Heinemann: p 100-101.

19. Jones: Smith's Recognizable pattern of human malformation, 4th Ed., 1988. W.B. Saunders Company. p 256-7.