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EDITOR'S NOTE

ORIGINAL ARTICLES

CASE REPORTS

EDITORIAL : SURGICAL PRACTICE AND ITS FUTURE IN PAKISTAN

With the explosion of technology in the last decades, newer innovations and modalities are being brought out each year. Every profession, in fact every person, is being affected by this and surgeons are no exception. Sometimes one feels that we surgeons are lagging behind, especially in Pakistan with our meague resources, inefficient manpower and at times total lack of planning and training, which has always led us to look towards the West and now towards the East, as well, for help.

If we look behind during the last 50 years since independence considering the conditions, the medical professionals and especially surgeons have not done badly. Almost every kind of Surgery, with a few exceptions, is being carried out in our country routinely now. I will confess that affluent people of our country are still not satisfied and they still go to the West for their treatment. Not that they consider our Surgery is of poor quality, but the cause of this dis-satisfaction is poor arrangements, even in the major care centres, and lack of properly trained paramedical staff and defective ancillary services.

At the time of Independence only major government and military hospitals provided Surgical services. But the population boom, especially during the last decades and the flight of population from rural to urban areas, has choked the Surgical services by sheer number of cases. During the last decades private sector has joined hands with the government and started several institutions and universities in the major cities of the country with teaching and training institutes for doctors, both at graduate and postgraduate levels.

With the expansion of surgical knowledge, skills and know-how, it has become impossible for one person to do all the jobs; hence specialities and sub specialities and even super specialities have been introduced. One can imagine the extent of this by just looking at The College of Physicians & Surgeons Pakistan, which awards minor and major diplomas in about 38 specialities. The organisation of surgical services, their practice and training has now taken newer, wider and higher dimensions, which are backed by higher technologies and inventions in the fields of investigation, treatment and management. Sometimes it becomes difficult to keep pace with these developments. Though the surgeons in Pakistan have taken these challenges well and still perform well inspite of several drawbacks and weaknesses in our health delivery system, these highly skilled professionals have to often work not only with less qualified and poorly trained colleagues, but have to compete with quacks of all kinds, from barbers to bone-setters to phony surgeons and have to deal with complications created by them. This is happening even in the major cosmopolitan centres, what to talk of rural areas. Poverty, Illetracy and lack of communication drives the uneducated, ill-informed and frightened citizens to go to the quacks for help.

This inherent danger, with its ramifications, can only be taken care of by mass education and awareness of the population and easy and timely access to properly staffed and well-equipped medical centres managed by properly trained professionals. For this our Government should revise its age old policies and bring them in conformity with the national requirements and needs of our citizens, the "suffering masses", both in the rural and urban areas of the country with especially more stress on training of paramedical staff.

The training of surgeons must be according to the modern day techniques and their evaluation must be standardized according to international standards. These objectives are being followed and updated in the country only by the College of Physicians & Surgeons Pakistan. But wider, urgent and tougher stands has to be taken by the Government and trainers to see that our national objectives are achieved and medicare is made available and affordable to everyone in the country.

ASADULLAH KHAN

LAPAROSCOPIC COST EFFECTIVE HERNIOPLASTY: A BANGLADESH EXPERIENCE

HUMAYUN KABIR CHOWDHURY

ABSTRACT:
Laparoscopic hernioplasty is probably the second most common laparoscopic procedure performed. This effective technique was used on 19 patients to set the ball rolling for laparoscopic hernioplasty in Bangladesh. Cases were done from January 1995 to June 1997. Hernia stapler was not used so as to reduce the cost. Intracorporeal suturing was carried out in all cases, using 3 to 4 stitches with 2/0 polyglycolic acid suture. average operation time was 80 minutes. There was only one early post operative complication of scrotal swelling. 3 to 28 months follow up in 12 cases did not show any recurrence Other patients were lost to follow up. This procedure is definitively less expensive than the technique where staples are used, but more expensive than open surgery, that can be compensated to some extent by decreased hospital stay.

KEY WORDS: Laparoscopic, Hernioplasty, Cost Effective.

INTRODUCTION

There are as many operations for inguinal hernia as there are surgeons[1]. The unquestioned success of laparoscopic cholecystectomy encouraged surgeons to explore other areas and inguinal hernia thus became one of the commonly performed procedure. Lichthenteins[2] tension free repair has definitely influenced laparoscopic repair. The feasibility of hernioplasty has been shown by many surgeons[3]. Cost of staples as a recurrent expenditure remained a major obstacle for developing countries. Intracorporeal suturing to fix the mesh has definitely reduced the cost and will help to popularize the technique in developing countries. Initial results of this pioneering small series are very encouraging.

MATERIAL AND METHODS

The study included 15 male and 4 female patients. Age range was 36 to 78 years (average 48 years). There were 9 direct and 10 indirect hernias and no recurrent hernia.

Transabdominal preperitoneal approach was made in all cases. Under general anaesthesia the patient was put in supine and head down position. Pneumoperitonium was done with veress needle and umbilical 11 mm port was introduced. General survey was done and then the other two ports were introduced, one 11mm on the ispilateral side and one 5mm on the contralateral side, lateral to the rectii a little below the level of the umbilicus. Peritoneum was incised after injecting adrenaline mixed with normal saline to elevate the peritoneum and to reduce blood oozing. Two peritoneal flaps were made to accommodate a prolene mesh of 11x6cm size considered adequate for our patients. Dissection of the sac was then done and if the sac was too big, then part of the it was left behind. Care was taken not to injure the epigastric vessel. Traction to the sac by a grasper, after holding the sac as far as possible from inside and dissection with the scissors were helpful. Pressure from outside on the hernia site was also helpful. After adequately dissecting the area, prolene mesh was introduced with a lateral sleeve cut to be passed behind the inferior epigastric vessels. The mesh is then placed and fixed with three to four stitches of 2/0 polyglycolic acid suture, one to the cooper's ligament, another laterally to fix the lateral sleeve and the other medially and above. Peritoneum was then closed with continuous 3/0 polyglycolic acid suture stitch. In all cases a gauze roll was placed externally on the hernia site and in male a scrotal bandage to keep moderate pressure for about 24 hours was also applied to obliterate the loose space.

RESULTS

In early post operative period there was only one complication of scrotal swelling that improved within one week. Analgesic requirement was minimum. Only 3 patients required single dose of pethedine. On first post operative day all the patients were ambulatory and were discharged from the hospital except one who was discharged on 3rd post operative day. Patients returned to normal activity within 5 to 7 days, which is usually 15 to 20 days in case of open surgery. Three to 24 months follow up in 8 cases

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was satisfactory without any complication or recurrence. Four patients did not return for follow up. Use of intracorporeal stitches definitively eliminated the cost of staples, but in comparison to the open operation it is still expensive, which may be counter balanced by less hospital stay and early return to work.

DISCUSSION

Laparoscopic inguinal hernia repair offers significantly decreased post operative pain, shorter hospital stay, faster return to work, fewer complications and comparable operating time. During the same time frame, surgery itself has evolved into a discipline more concerned with cost-effectiveness, outcomes and consumer acceptance[4]. Use of intracorporeal suturing reduces the cost which will definitely inspire many surgeons in the developing countries to practice this procedure. In the developing countries hospital beds are less expensive, so once the surgeons feel comfortable with this procedure and time of surgery comes down, surgeons will charge less and that will reduce the cost of surgery; only then it will be comparable to open surgery. In this pioneering small series of 19 cases results are satisfactory, cost was less because staples were not used and operating time was almost comparable, which will come down to less than open procedure with experience.

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ELECTRONIC VIDEOENDOSCOPY

MIR ZAFAR ALI, ASADULLAH KHAN, IRSHAD WAHEED

ABSTRACT:
Endoscopy is the visual inspection of any cavity of the body by means of an endoscope. Early methods of endoscopy used rigid instruments with relatively simple optics, as in the cystoscope or without lenses in rigid sigmoidoscope. Fiber optic technology, which became available in the 1960s, led to the development of flexible instruments which have allowed simpler, safer and more extensive examination of the gastrointestinal tract. Electronic Videoscopy is a convenient form of fiberoscopy with a video monitor attachment.

KEY WORDS: Videoendoscopy, Electronic Imaging In Endoscopy.

INTRODUCTION

The principle used in fiberoptic endoscopy is the internal reflection within suitable glass fiber. These are arranged in a 'coherent' fashion i.e. the fiber arrangements at both ends of the bundle is identical and image can be obtained. Most flexible endoscopes are similar in design with a fiberoptic light-carrying bundle which conveys illumination from the light source to the tip of the instrument and an optical bundle which transmits images from the tip to the eye piece. With developments in camera technology, it is now possible to produce endoscopes with solid state imaging device at the tip rather than on the fiber bundle. With digital imaging' the acquisition of accurate endoscopic images is one of the functions that can be processed in a variety of different ways to obtain different information from the same target site. A variety of instruments are presently available for image processing and recording formats.

ELECTRONIC VIDEOENDOSCOPY

Videoendoscopy has been made possible by impressive innovations in the field of micro-electronics. Most significant is the development of the *charge-coupled device*, referred to as the CCD-chip. A CCD is an array of 33-1,00,000 individual photo cells (known as picture elements or pixels) receiving photons reflected back from the mucosal surface and producing electrons in proportion to the light received.

The basic components for a videoendoscope are an electronic scope with a CCD, a video-processor and a TV monitor[1-3]. Electronic Videoendoscopy has been so named because the entire examination can be viewed on a TV screen, in contrast to standard endoscopy, in which visual observation is normally limited to the view from the eyepiece. Videoendoscopy procedure is performed entirely by looking at the TV monitor[1,4].

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Light reflected from the target area is focused by a lens onto the CCD which transmits the image to a video processor for relay to a TV monitor. The image obtained from the CCD is in the form of electrical signals, which are relayed to the video processor through an electrical wire.

Basically Videoendoscope are of two types:
1. Direct Videoendoscope
2. Indirect Videoendoscope

The direct type differs from the indirect type by the use of a CCD chip at the tip of the endoscope and acquires images directly from its lumen.Indirect Videoendoscopes are standard fiberoptic endoscopes with camera attachments. Both types of scopes provide facilities for TV viewing, photography , hard-copy printing and video recording.

CCD chips were first utilized in endoscopy when Welch-Allyn Incorporated included them in the manufacture of gastrointestinal endoscopes. These endoscopes were first tested clinically in the United States in 1982. The result of these tests were reported by Sivak[1] in 1984.

In common with all other television systems, the individual receptors of the CCD respond only to degrees of light and dark and not to color. The CCD was invented by W.S. Boyle and G.E. Smith in 1969. By the seventies multiple scientific applications had been discovered and commercially applied[5]. The main function of the CCD is only image detection. The video processor is necessary for image analysis, reconstruction, and relay of the reconstructed image to a monitoring or documentation system. Interconversion of analog and digital signals takes place within the video processors[5-7]. The videoprocessor contains electronic components dedicated to storing and manipulating the vast volumes of data presented by incoming video signals[7,8]. Finally, after an image is detected by the CCD chip and processed by the video processor, it can be viewed on a TV monitor. The quality of the image obtained, is primarily dependent on the character-

istics of the CCD. The important characteristics are that of resolution and the *signal to nose ratio* (SNR). Resolution is defined by the number of pixels on the CCD and determines how well an image can be reproduced in fine detail. The SNR also determines the ability to perceive the image three dimensionally[9,14]. Images with a low SNR are coarse and flat, and those with a high SNR are more vivid and amenable to three dimensional analysis. Therefore images with a higher SNR are desirable. The CCD has made possible the construction of a miniaturized camera easily mounted on endoscope tips[10]. Thus no fiber optic bundle is used for image transmission. The only fiberoptic bundle found in videoendoscopes is a single bundle used for light transmission from the processor to the tip of the scope[11,12].

There are two types of CCDs color and monochrome (or black and white). The color CCD contains color pixels and can simultaneously detect different color wavelengths of light. Color CCD's have extra pixels to allow for an overlay of multiple primary color filter stripes, transforming the pixels to a particular color. The electrical color signals derived from this CCD are reconstructed with in the video processor to permit a composite image to be viewed on the TV screen. Color CCDs simplify color acquisition but are considerably larger in size to the monochrome CCDs, increasing the diameter of the distal rigid portion of the endoscope. This is one of the reasons why monochrome CCDs are preferred today.

(Black And White) Monochrome CCDs can only provide black and white signals. To generate color image from a Monochrome CCD, light from a Xenon lamp within the video processor, (Fig. 1) is passed through a rotating wheel which has a glass which provides the three primary colors, red, green and blue. Most color images can be simulated by an appropriate blend of these three colors[13-15].

Red, blue and green light components reflected from the target area and detected by the monochrome CCD, are then transmitted as an analogue signal to the processor. Here they are converted into digital data which are analyzed and reconstructed into a composite of the original image. The reconstructed digital image is converted back into an analog signal for transmission to the TV monitor where the original live image can be viewed[2,13,16]. The Monochrome CCD primary color images (in mostly red, some green and a little blue are stored transiently in bands of memory chips in the processor and fed out to the red/blue/green electron gun of the TV monitor every 30th of a second. Thousands of chips and sophisticated computer technology are used to optimize the underlying single CCD output and account for the excellence of the image produced by Monochrome CCDs system. Therefore, the high price involved, as well as the relatively large processor.

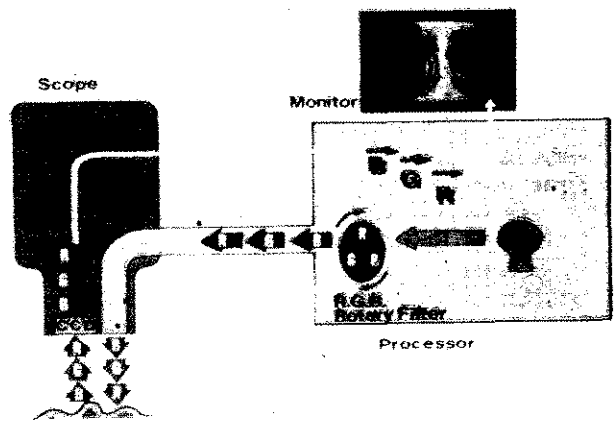

Figure 1 Standard monochrome CCDs can only provide black and white signals. Color generation takes place within the video processor by passing white light through a high speed rotation filter to produce three primary colors; red, green and blue (RGB).

By contrast, a color CCD uses white light from a light source, and inputs directly into the monitor, requiring a cheaper and smaller processing unit.

Technical Restriction of Videoendoscopes

All CCD's, especially the sequential variety, performs rather poorly in red light, such as a blood filled intestinal lumen, in which the view becomes fuzzy and fluorescent in quality. Leaving aside the problem in bleeding patients, the screen image quality of present video endoscope in both color and resolution, otherwise approximately equals that of present fiberoscope. Videoscopes are therefore not well suited to bleeding emergency endoscopy where tried and true fiberoptic large channel instruments, coupled with the remarkable range of light sensitivity and image processing capability provided by the human retina and optical cortex, give result so far unequaled by electronic system.

The objection that video-endoscopes introduce artificial color values in untenable, as :-

i. They can be shown in technical studies to give a remarkable faithful rendering of test charts;
ii. The visual assessment of pathology depends little on absolute color values and is any way backed by histopathology.
iii. There is the inescapable fact that individual perception of color varies significantly, the extreme example being color blindness.

Advantages of Electronic Videoendoscopy

Video Endoscope provide many advantages :-

- Allow number of viewers to observe.
- Monitoring
- Documentation and input of additional data.
- Electrical manipulation of images.
- Magnification.
- Binocular view analysis which is facilitating teaching, training and consultation and assistance in diagnostic

and therapeutic procedure.

- Freeze facility of the scope permits an endoscopist to select the best view and eliminate motion artifacts and better quality of photographs can be obtained.
- Photography with videoscope is simplified by the use of one touch remote control switches which are conveniently located on the control unit of the scope.
- Hard copy in computer, print and video recording is easier.
- Video recording permits extended analysis of the procedure without bothering the patient.
- Images recorded in a floppy disk become more portable. Edited tapes can be used for lectures, conferences and consultations.
- Creation of an endoscopy data base file is made possible by videoendoscopy.

Structural Advantages

- Videoendoscope provides structural advantages. In case of fiber optic technology the scope length is limited by the technical requirements of image transmission, However by placing the CCD at the tip of the scope, there is no practical limit to its length, which is important in gastrointestinal endoscopy
- Electrical cables are thinner allowing thinner insertion tubes which are acceptable to both, endoscopist and patient.
- Viewing the procedure can increase interest of the patient and tolerance.
- CCD is much cheaper as compared to fiber optic bundle, which are prone to breakage while handling, resulting in deterioration of images. In contrast videoendoscopes can be expected to offer excellent images for much longer period.
- Hard copy imaging in employing only the ocular lens system at the instrument tip without the degrading effect of transmission on optical bundle and through a secondary lens system.

Notes for use of Videoscopes

It must be remembered that the scope is an instrument designed to be inserted into the body of the patient Special attention should be paid to the coating of the scope to prevent electrical leakage, the endoscopist should wear rubber gloves and patient bed must be electrically insulated from the floor. Even if the possibility of an electric shock is almost nil, it is better to err on the safe side and meticulously apply the prevention commonly used with electrical equipment.

Future of Electronic Videoendoscopy

Progress in videoendoscopy can be expected to parallel development in CCD chip technology and computer technology.Videoendoscopy equipment CCD chip television cameras have the advantage of providing a far greater amount of information than the eye can see. This can allow image magnification without sacrificing details to focus on a particular aspect of morphology[17].

The development of smaller color CCD chip coupled with high speed electronic shutter would minimize the problem of color black-up during fast movements and more portable video-endoscopes can be developed which can be used at the bed-side, creation of computer program and soft-ware which can permit the creation of automatic diagnostic system linking to the existing diagnostic program in the computer system.

High definition TV (HDTV) system provides higher resolutions. The present TV system formats use approximately 500 lines. this effectively limits the number of usable pixels on the CCD to around 400,000 pixels for each of the three primary colors. With the development of HDTV it is now possible to utilize around 1,500,000 pixels for each of the three primary colors. However, innovation using the so called integrated management system. would permit future coupling of CCD and HDTV technology. Significant improvements in both the quality and nature of the video images obtained can therefore be anticipated.

Over all, it seems likely that Videoendosopes will rapidly take over a majority position in G.I. endoscopic units, although fiber optic instruments will, by virtue of their simplicity and small diameter capability, retain a valuable role.

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INTERNATIONAL SURGICAL EVENTS

KARACHI (PAKISTAN): INTERNATIONAL COMBINED SURGICAL MEETING 97 - KARACHI

An International Combined Surgical Meeting 97 will be held at College of Physicians & Surgeons Pakistan from November 7th to 9th, 1997. It is a joint meeting sponsored by International College of Surgeons (Pakistan Chapter), SAARC Association of Surgeons (Pakistan Chapter) and Society of Surgeons Pakistan. The theme of the Congress is **Healthcare Future in Under Developed World.** The Scientific programme is going to offer updates, plenary sessions, symposia and panel discussions on virtually every field of surgery. The workshops (hands on) are being offered on the subject like internet in the Medicine, Medical Statistics and other related subjects.

SINGAPORE: ORTHOPAEDIC ASSOCIATION OF SINGAPORE, 20TH ANNUAL GENERAL MEETING

Singapore Orthopaedic Association is holding its 20th Annual Scientific Meeting from 20 to 23 October 1997. The theme of the meeting will be **Lower Limb Reconstruction.** It will highlight the state of art in the management of difficult hip and knee revision orthoplasty for osteolysis, loosening and infected joint replacement.

ACAPLCO (MEXICO): WORLD CONGRESS OF SURGERY

The 37th Int. Surgical Week of International Society of Surgery and International Federation of Surgical Colleges was held at Acapulco (Mexico) from August 07, 1997. The theme of the conference was **"Endoscopic Surgery in Developing Countries".**

KARACHI - PAKISTAN - 3RD CONGRESS OF SOUTH ASIAN CONFEDERATION OF ANAESTHESIOLOGISTS.

The 3rd Congress of South Asian Confederation of Anaesthesiologists will be held at Hotel Sheraton, Karachi from 26th to 29th November 1997. Satellite Meetings will be held at Lahore on Sunday, 30th November 1997 and Monday, 1st December 1997 at Islamabad.

CONFERENCES HELD BY COLLEGE OF PHYSICIANS & SURGEONS PAKISTAN, KARACHI

Since 1993, the College of Physicians & Surgeons Pakistan has held three international joint conferences in which the following surgical aspects were covered by experts in their respective fields:-

CONGRESS 93

- "Transplant Surgery in the 21st Century with special reference to laryngeal transplant - a dream or reality" by Dr. Marshall Strome, (U.S.A.)
- "Recent advances in assisted reproduction technology" by Prof. S. Shan Ratnam; (Singapore)
- "New Modalities in the evaluation of abdominal trauma" by Dr. Aurelio Rodreguez: (U.S.A.)
- "Will Islet Transplantation be a Treatment of Diabetes in the 21st Century" by Dr. Gordon C. Weir (U.S.A.).

Hands on Workshops for practicing surgeons and postgraduates on :-

- Arthroscopy & Total Knee Replacement
- E.R.C.P.
- Laparoscopy
- Bronchoscopy and Bronchial Lavage
- Hysteroscopy
- Upper G.I. Endoscopy

CONGRESS 95

- "Organ Transplantation in the year 2000" by Prof. Adibul Hasan Rizvi.

CONGRESS 96

- "Modern Trends in the Orthopaedic Management of Bone and Cartilage Tumours" by Prof. D.L. Hamblen, (RCPS Glasgow).
- Ano-Rectal Surgery: From Pouches to Sphincters by Dr. I. G. Finaly, (RCPSG).
- Newer Perspectives on Thrombogenesis by Prof. C.D. Forbes (RCPSG).
- Cleft Lip and Palate by Prof. Ijaz Ahsan (President CPSP).
- The Golden hour of Trauma care - seven years on by Dr. I. W. R. Anderson (RCPSG).

EXTRA PULMONARY TUBERCULOSIS IN SURGICAL PRACTICE

JAMSHED AKHTAR, ABDUL AZIZ

ABSTRACT:
Tuberculosis can affect any organ of the body. The principal clinical problem of extra pulmonary tuberculosis is difficulty in diagnosis. In two years period from January 1994 to December 1995, 47 cases of Extra Pulmonary Tuberculosis were managed at National Institute of Child Health, Karachi. There were 29 female and 18 male patients. Eleven patients were below 5 years and 36 between 5 and 12 years of age. Cervical lymph nodes were involved in 21 cases, followed by musculoskeletal system that was effected in 12 cases. Rare sites included liver, testis and parotid gland. Early diagnosis can help in decreasing morbidity and mortality associated with this disease.

KEY WORDS: Tuberculosis, Mortality, Children.

INTRODUCTION

Tuberculosis, an ancient disease, is still flourishing; in fact it is on the increase even in developed countries[1] Pulmonary tuberculosis is still one of the commonest conditions treated medically. Tuberculosis in surgical practice (extra pulmonary tuberculosis) is still difficult to diagnose, specially abdominal tuberculosis and tuberculosis of unusual sites[2]. We conducted a study at NICH to collect data on this problem.

MATERIAL AND METHOD

This is a retrospective study conducted over a period of two years from January 1994 to December 1995. in the Surgical Department of National Institute of Child Health (NICH), where patients below 12 years of age are treated. All histologically proven cases of tuberculosis were included in this study. The cases of pulmonary tuberculosis and tuberculosis meningitis were excluded, because they are dealt with by physicians. The clinical data was collected on a proforma that included age, sex, site, operation performed etc and follow up.

RESULTS

In the two years period, January 94 to December 95, 47 cases (18 males and 29 females) were managed. Eleven patients were less than 5 years of age and 36 were between 5 and 12 years. Details of various sites affected are given in Table 1.

25 percent patients had associated pulmonary tuberculosis. All patients underwent some kind of surgery depending upon the site and were later prescribed antitubercular therapy, which included Rifampicin, Isoniazid and Pyrazinamide

Three patients with spinal tuberculosis presented with psoas abscess, which were drained only and bone grafts were not done. They were advised bed rests; splints were not applied.

Peritonitis was the most common presentation in patients with intestinal tuberculosis due to perforation. All patients underwent laparotomy with drainage of feculent material,

TABLE I ANATOMICAL REGION ANVOLVED

Region		No.
Lymph nodes		**21**
Cervical	16	
Axillary	2	
Inguinal	3	
Musculoskeletal		**12**
Pott's disease	3	
Bone (long)	2	
Ribs	3	
Innominate bone	1	
Joint	3	
Abdominal		**8**
Intestinal	5	
Tabes mesenterica	2	
Liver	1	
Genitourinary		**4**
Renal	3	
Testicular	1	
Miscellaneous		**2**
Parotid gland	2	

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perforation could not be identified in any of them. Biopsy was taken and drain kept. One patient later died and one developed faecal fistula.

One patient was a suspected case of hepatic tumor and on laparotomy, was found to have large tuberculoma. Three patients with renal tuberculosis presented with mass in lumbar region and fever and rigors. Diagnosis of pyonephrosis was made. X'ray revealed soft tissue density with calcification. Urine was negative for acid fast bacilli. Nephrectomy was performed. In one patient faecal fistula occurred for which colostomy was made which was closed at a later date.

A patient with testicular tuberculosis presented with painless enlargement of right hemiscrotum. With suspicion of tumor, exploration was undertaken on next elective list. Cord was found to be thickened and testis enlarged. Orchidectomy was performed. On bisecting the testis, it was found to contain caseous material with no grossly identifiable testicular tissue. On further investigation no evidence of tuberculosis was found in the urinary tract. The patient was lost to follow-up and appeared after six month with mass in abdomen. He was not taking antikoch's treatment. With suspicion of abdominal tuberculosis, no surgical intervention was done and anti-tubercular drugs were started, patient responded to treatment.

DISCUSSION

BCG vaccination is given to all children as a part of expanded programme of immunization but its protective role is still undetermined[3]. Tuberculosis can effect any part of the body, the most common site being lungs. A sizable number of patients present with extra pulmonary tuberculosis, which demands high index of suspicion and active intervention and to limit the spread of the disease.

Presentation of extra pulmonary tuberculosis, is usually slow. Cervical lymph node enlargement was the most common site in our study. Diagnosis is suspected on clinical grounds and early biopsy helped in planning management, that included antitubercular drugs and excision of all grossly involved lymph nodes. Radical surgery was not attempted.

Bones and joints are frequently involved in tuberculosis of which spinal tuberculosis is the most common type[4] Tuberculosis of bone and joints is a crippling disease and early intervention is needed to prevent damage to these sites[5], for which early synovial fluid culture and biopsy are of great help.

Abdominal tuberculosis presents with bizarre symptoms and signs[6] and is usually caused by human type rather than bovine strain. The population in which it occurs belongs to poor socio-economic group which is malnourished and lives in unhygienic conditions. Laparoscopy helps in diagnosing these cases as it allows visualization of the lesion and at the same time, biopsy can be taken without subjecting the patient to major surgery[7].

In conclusion, patients with extra pulmonary tuberculosis present with unusual manifestations[8]. Pulmonary tuberculosis may or may not be present[9]. Tuberculin test may not be reliable but induration of more than 10 mm, may be of significance as it may indicate active disease[10]. New diagnostic modalities like Bactec TB system, DNA probe and Polymerase chain reaction may help in rapid identification of specific mycobacteria with high sensitivity and specificity[11]. Culture of the organism and histopathology of accessible sites are diagnostic tools. In addition to surgical treatment antitubercular drugs in appropriate doses at regular times and for adequate period go a long way in controlling the disease[12]. This aspect is often forgotten and is the most neglected part of the treatment. This results not only in spread of the disease but also leads to emergence of resistant strains of the organism[13]. WHO has issued guidelines in this regard[14] and established a standardized S.C.C. (Short Course Chemotherapy).

We must understand local conditions when we are treating patients with tuberculosis and should not forget that there is still a social stigma attached with this disease. Drugs must be prescribed individually or in combination only after the clinician has thoroughly familiarized himself with available preparations. In order to avoid wrong dosing, it is suggested that the patient be instructed to bring all the prescribed medicines at each visit.

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ZINC THERAPY IN WILSON'S DISEASE

SHAUKAT ALI, A. ZAHIR KHAN, HASAN AZIZ

ABSTRACT:
To evaluate the results of Zinc therapy in Wilson's Disease (WD), a study of ten cases of WD treated with Zinc was carried out at the Department of Neurology, Jinnah Postgraduate Medical Centre, Karachi. Patients were diagnosed and assessed according to Yahr & Hochn scale of disability, Kayser - Fletcher rings, estimation of serum ceruloplasmin and urinary copper. All but one patient showed steady improvement in six months of treatment. Zinc therapy is effective in treatment and prevention of Wilson's disease with minimal and reversible side effects. It is cheapest of all the remedies of Wilson's disease available in Pakistan.

KEY WORDS: Zinc Therapy, Wilson's disease.

INTRODUCTION

The Pseudosclerosis of Westphal and Strumpell, progressive lenticular degeneration of Wilson and hepatolenticular degeneration of Hall are the terms adopted at various stages of evolutionary discoveries of Wilson's disease (WD)[1-3]. Wilson knew that the disease runs in families but it was Hall who labeled it autosomal recessive disorder[2,3].

Deposition of abnormal pigment round the limbus of cornea was noted by Kayser and Felischer in patients with lenticular degeneration in 1902. Excessive copper deposition in WD was discovered in the brain in 1930 and in the liver in 1945. A chance observation of Mandelbrote showed excessive copper excretion in urine of WD patients[4]. Estimation of low serum ceruloplasmin, a copper binding and disposing protein has been in use as a diagnostic tool for WD since 1952[5].

Wilson's disease usually presents in childhood or young adult life. Average age at onset is about 11 years for patients presenting with hepatic dysfunction and 19 years for those with initial neurological manifestations; but the disease presenting as late as the sixth decade has been described[6].

Hepatic and neurological presentations are equally common and most patients, if left untreated, eventually develop both types of involvement. Most of the neurological patients initially present with behavioral disturbance[7].

Because treatment and prophylaxis of WD is life saving, diagnosis must be made or ruled out in every individual in whom WD is suspected. Whether an individual is a symptomatic or ill, a diagnosis or WD can be established or ruled out unequivocally in almost all instances by demonstrating a serum ceruloplasmin concentration <200 mg/L and either a hepatic copper concentration>3.9 umol/g dry liver (>250 ug/g dry liver) or corneal deposits of copper (Kayser - Fleischer rings)[8].

Average diet of adult humans in western countries contains from 0.6 to 1.6 mg Cu/day. Salivary, gastric and other secretions contain 2.0 mg. Normal biliary content of copper in 24 hours is 2,5 mg while it is not available in the bile of WD patient. Only 2 to 3% of the dietary copper is excreted in urine of a person while the rest is excreted in faces. In WD the failure of biliary excretion causes positive balance of copper which accumulates to a toxic level, manifestating clinically as behavioral disorder and neurologic and Hepatic dysfunctions. Chellating agents increase the urinary excretion of copper, while Zinc decreases absorption from the gut. The mechanism behind the zinc-copper interaction in the small intestine was studied by Hall et al. They found that high dietary zinc induced intestinal metallothionein and proposed that this newly synthesized metallothionein may act as a "trap" for absorbed copper[11,12]. Zinc induces metallothionin in the gut, liver and other body tissues as well[10].

Copper tends to displace other metal ions from metallothionins and high concentration of the latter induced by diety factors such as zinc can retard the passage of copper from mucosa into the blood, thus enhancing the antagonism to absorption provided by high zinc intake. This tightly bound copper is then released into the gut when the intestinal cells[11,13] slough off. 2.3-Dimercaptopropanol (BAL), a chellating agent, was initially used but was replaced by penicillamine for the treatment of Wilson's disease in 1954[9]. It is a costly drug with many side effects, whereas Zinc in WD provides better remedy and is less expensive.

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Trientine, another chellating agent has been used in case of adverse reaction to penicillamine. These chellating agents have been reported with sensitivity reactions like rashes, fever, leukopenia, thrombocytopenia, or proteinuria. In case more serious toxic effects (including disseminated lupus, cytopenia, pemphigus, and proteinuria) occur, the drug is stopped[14]. Penicillamine has been reported with induction of permanent neurological deficit when used as initial therapy in ptresymptomatic cases[15].

Zinc was introduced for the treatment of WD by Schouwink in 1961[16] Zinc 75mg daily in three divided doses or 150mg daily in six divided doses has been used effectively[17,18].

PURPOSE OF STUDY

To evaluate the effects of Zinc therapy in Wilson's Disease (WD).

MATERIAL AND METHOD

It is a hospital based open study, conducted at the Neurology Ward of Jinnah Post Graduate Medical Centre, Karachi. Patients of all ages and both sexes who presented with progressive movement disorder/and or dementia were subjected to K-F rings examination with slit lamp and serum ceruloplasmin estimation. K-F ring positive cases with low ceruloplasmin were diagnosed as WD and put on Zinc therapy. Family members of WD patients were screened by 24 hours urinary copper estimation and the positive cases were subjected to serum ceruloplasmin estimation and K-F rings examination. Yahr and Hoehn scale of disability were used for clinical evaluation of disability.

All the patients were put on 100 mg zinc in four divided doses daily. High copper diet meat, liver, brain, potato and nuts were restricted. 24 hours urinary copper estimation was carried out at the start of Zinc therapy.

RESULTS AND DISCUSSION

Nine cases of progressive movement disorder with low ceruloplasmin and positive K-F rings were diagnosed as suffering from WD and were put on Zinc therapy. Their mean age at presentation was 16.3 +3 years with age ranging from 7 to 21 years. Five were males and five females (Table 1). One case was presymptomatic found during family screening for WD (Table 2).

TABLE I ZINC RESPONDERS

Responders	No.
Male	5
Female	5
Total	10
Age: Range	(7-21) years
Average age:	16.30 + 3.28
Male to Female ratio:	1:1

TABLE II CLINICAL FEATURE IN ORDER OF FREQUENCY

Clinical feature	No.
Dysarthria	8
Dystonia	8
Athetosis	5
Inappropriate smile	5
Akinesia	5
Tremor	3
Dysphagia	3
Drooling of saliva	2
Aggressive behavior	1
Myoclonus	1
Chorea	1
Writer's cramps	1

At the start of Zinc therapy, 4 patients were in grade 5, 2 patients in grade 4 and one patient in grade 3 and one patient in grade 2. Symptomatic improvement started in the first month of zinc therapy in all cases. In the second month of therapy patient started having improvement in their disability.

In the course of 3 to 6 months of zinc therapy, 8 cases improved from worst grades of disability to the better grades of disability evaluated according to the disability scale of Yahr and Youhen (Table 3). After 6 months of zinc therapy one patient was in grade 4 and one in grade 5 while 2 patients were in grade 1, grade 2 and grade 3 each. One presymptomatic case remained asymptomatic during the course of therapy. He complained of nausea on taking zinc in the begining of the therapy. Zinc failed to halt the progression of disability in one case .Four totally disabled, dependent, bedridden and mute cases are now independent and can speak meaningful and understandable sentences easily.

All cases of dyarthria are improving. One case of writer's cramps, who could not write due to spontaneous spasm of hand musculature, is now capable of writing three

TABLE III IMPROVEMENT IN DISABILITY OF INDIVIDUAL CASES AFTER ZINC THERAPY

Cases	Grade before Theray	Grade After Therapy
1	5	2
2	5	3
3	5	4
4	5	4
5	4	2
6	4	3
7	3	2
8	2	1
9	1	2 (Could not halt the progress)
10.	Presymptomatic	Presymptomatic

pages per day. Six cases whose urinary copper estimation were carried out after six months of zinc therapy, has shown decreased urinary copper excretion. The case which did not improve clinically with zinc therapy has also showed decreased copper excretion with zinc therapy (Table 4).

TABLE IV BIOCHEMICAL RESPONSE TO ZINC THERAPY. 24 HOURS URINARY COPPER

Cases	Before therapy	After therapy
1	576 ugm/24 hours	186 ugm/24 hours
2	260 ugm/24 hours	224 ugm/24 hours
3	133 ugm/24 hours	60 ugm/24 hours
4	150 ugm/24 hours	103 ugm/24 hours
5	153 ugm/24 hours	46 ugm/24 hours
6	45 ugm/24 hours	34 ugm/24 hours

Two cases which improved clinically in the course of two months of zinc therapy, abandoned the drug. Both of them started to deteriorate ten days after stopping the drug.

One patient lost 5 kg in 6 months while another lost 2 kg in three months. Four patients developed anemia. Two patients developed sore throat and fever and one patient who did not improve with zinc therapy, developed pain in the dystonic limb (Table 5).

TABLE V SIDE EFFECTS

Side effect	No.
Transient nausea	2
Weight loss	2
Pain in the dystonic limb	1
Anemia	4
Fever with sore throat	2

Eight patients started symptomatic improvement in the third and fourth week of Zinc therapy. Improvement in the disability grades started in the fifth and sixth week of therapy. Four totally dependent, bed ridden and mute cases, are now capable of walking, eating, washing and cleaning themselves independently; as well as speaking meaningful and understandable sentences easily. Zinc also helped improve behavioural disturbances, as one patient with agressive behaviour and five patients with inappropriate smile totally recovered after three to six months of therapy.

One presymptomatic patient on zinc for six months is still symptom free but had nausea and sometimes vomitting after taking zinc during the first month of therapy. His twenty four hours urinary copper dropped to lower level with zinc therapy for six months. This chemical manifestation of his metabolic improvement provides an indirect evidence of disease prevention.

Five of the cases improved clinically as well as biochemically as evidenced by clinical evaluation and 24 hours urinary copper estimation with zinc therapy.

Zinc therapy could not halt the progress of disease in one case (Table 4) on the above mentioned regimen of treatemnt, though his urinary copper in 24 hours decreased. He probably needs larger and more frequent dosages of Zinc.

In none of the patients any acute deterioration occured. Complications like anemia and sore throat etc could be managed with iron and antibiotics. Two patients, who were obese, lazy, and socially inactive lost weight. After zinc therapy they became socially active and probably this was the reason for their weight loss.

Two of the cases abandoned therapy after two months of therapy in spite of improvement. They started deteriorating after the tenth day of cessation of therapy. Symptomatic improvement in the third week of therapy in eight cases and deterioration at the tenth day of cessation of therapy in two cases may be reflection of the fact that probably maximum induction of metallothionein takes place in the third week of zinc therapy and half life of metallothionein may be less than two weeks.

CONCLUSION

We conclude that zinc therapy is cheap and effective in treatment and prevention of Wilson's disease. It has reversible minimal side effects and does not deteriorate the disease. It costs WD patients seven dollar per year. Twenty four hours urinary copper estimation is a good index to use in monitoring the response of zinc therapy biochemically.

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VASCULAR TRAUMA : EXPERIENCE IN JPMC, KARACHI

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ABSTRACT:
All the forty seven patients, with 49 arterial injuries, who reported at the Department of Orthopaedic Surgery, Jinnah Postgraduate Medical Centre, Karachi from December, 1994 to June, 1997, were included in this retrospective study. Time lapse between the injuries and arrival at the hospital varied from seconds to three months. Average follow-up was eighteen months. Male/ female ratio was 9:1. Firearm injuries (42.55 %) were the most frequent cause of vascular injury, followed by road traffic accidents and iatrogenic injuries. Femoral, popliteal and brachial vessels were the most common injured arteries. Majority of the injuries were complete tears. Most patients had associated injuries (51.08 % had fractures). Twenty nine arteries were repaired primarly, 8 with vein graft. Nine (31.03 %) repairs failed. Most of the failures could be attributed to iatrogenic factors. We had worst results with conservative treatment. Overall amputation rate was 27.67 %. The mortality rate in our series was 10.68 % with only one case dying due to bleeding from the artery.

KEY WORDS: Vascular Trauma

MATERIAL AND METHODS

This is a retrospective study conducted at Jinnah Postgraduate Medical Centre from December, 1994 to June, 1997. All of the forty seven patients with arterial injuries were included in this study. The diagnostic criteria for vascular injuries were mainly clinical, that is, the type and location of wound, amount of bleeding, the quality of distal pulse and circulation. However for late cases angiography was performed. Various treatment options suitable for individual patients were tried and the results were evaluated.

The treatment options used were: Primary repair with or without vein graft, ligation conservative with anticoagulant therapy and amputation. They were used according to the clinical needs of the patient. Heparin was not used post operatively in any case. The criteria for failure of repair were: development of ischaemia or gangrene, severe bleeding, and absent or weak distal pulses. Follow- up was done six months to three years.

The data was recorded for sex, age, mechanism of injury, type and location of injury, mode of presentation, associated injuries and results of the treatment. It has been analysed and compaired with other studies.

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RESULTS

Sex

Male	44 (93.62%)
Female	03 (6.38%)*
Age	28-6 years**

*All females had popliated injury due to gunshot wounds during family feuds.

**Range: six to fifty years

TABLE I MECHANISM OF INJURY

Mechanism	No.	%
Firearm	20	42.55
Raod traffic accident	12	25.53
Latrogenic trauma	05	10.63
Sharp penetration injury	04	8.5
Others	06	12.76

DISCUSSION

Vascular injuries are not uncommon and occur more often in violent societies such as ours or during military conflicts[13,15]. The usual victims are young males. Hardins[5] reported 86.69% of the victims being male, while Menzoian[8] reported 94.64 % being male. 93.62 % victims in our series were male female 9:1. The mean age reported by Hardins[5] and Menzoian[8] are 28.8 years and 32 years respectively. The mean age in our series was 28 ±6 years.

Fire arm injuries are the commonest cause of vascular injuries reported by many authors[3,5,7,8,11,13,15,19]; the highest

TABLE II ARTERIES INJURED	
Arteries injured	No.
Common carotid	01
Subclavian	02
Axillary	01
Brachial	09
Radial	05
Ulnar	01
Seperior gluteal	01
Inferior gluteal	01
External iliac	02
Femoral	12
Popliteal	10
Anterior tibial	01
Posterior tibial	02
Dorsalis paedis	01

TABLE III TYPE OF INJURY		
Type	No.	%
Complete tears	37	75.57
Partial	11	22.45
Thrombosis	01	2.04
Pure arterial	30	63.83
Combined arterio venous	17	36.17

TABLE IV FINDING AT THE TIME OF PRESENTATION		
Type	No.	%
Weak/Absent pules	46	97.87
Shock/Bleeding	18	38.29
Expanding haematoma	08	17.02
Gangrene	07	14.89
Visible arterial injury	10	21.27

TABLE V ASSOCIATE INJURIES		
Type	No.	%
Fractures	24	51.08
Muscle/Tendon injury	17	36.17
Nerve injury	12	25.53
Visceral injury	08	17.02
Skin loss	13	27.66

TABLE VI TIME INTERVAL BETWEEN INJURY AND ARRIVAL AT HOSPITAL		
Type	No.	%
< 6 hours	28	59.57
6 to 12 hours	08	17.02
12 to 24 hours	02	04.25
> 24 hours	09	19.15

TABLE VI TREATMENT CARRIED OUT		
Type	No.	%
Primary repair	21	42.85
Interpositional vein graft	08	16.32
Ligation	09	18.36
Conservative	06	12.24
Primary amputation	05	10.20
Amputation	14	27.67
Mortality	05	10.68

(60%) been reported by Hardin[5]. 42.55 % of the victims in our series had firearm injuries. Blunt injuries are second most common as reported by Franklin[3], Menzoian[8] and Snyder15, however treiman had sharp penetrating injury (38 %) as the second most common cause. 25.53% of our vascular injuries were caused by road traffic accidents.

The third most common cause reported by many authors is sharp penetrating injury; however in our series iatrogenic injuries (10.63 %) were third on the list. Iatrogenic vascular injuries occur after cardiac catheterization[9] or arterial punctures[11] but rarely can occur after total hip replacement[2,12], disc surgery[9], arteriovenous[9] shunt formation and others[7]. In our series, of the five iatrogenic injuries, three occurred during tumor surgery, one during surgery tibial nonunion and one during revision surgery for fractured femur with femoral arterial injury, repaired initially with interpositional vein graft. Femoral, popliteal and brachial arteries are commonly affected[2,3,9,19], as in our series.

Pure arterial injuries are more frequent than combined arterial / venous injuries or only venous injury as reported by Hardin[5] (Arterial 61.98%, Arterial / venous 28.51%, venous 9.5%) and Menzoian[8] (Arterial 64.28 %, Arterial / venous 23.21% venous 12.5%). Our series had similar findings. Most of our arterial injuries (75.57%) were complete. The others were being partial tears except one radial artery which showed thrombosis. Pearce[11] has pointed that most of the carotid artery injuries are partial, as was our one. Mode of presentation[2,3,10,11] is usually bleeding, shock, absent or weak pulse, expanding haematoma, distal limb ischaemia or obvious open vascular injury. Vascular injuries are frequently accompanied by other injuries, Hardin[5] reported 48.2% had associated injuries including nerve injuries (29 cases), fractures (16 cases) and soft tissue damage (13 cases). Menzoian[8] reported 37.33 % coincidence of fractures with vascular injuries. Subclavian, axillary and brachial artery injuries are usually associated with nerve[7,11,17] injury. The highest reported coincidence of axillary artery and brachial plexus injury 92%. McCready[7] also draws attention to the supracondylar fracture and brachial artery injury. Mucha[10] observed that the vascular injury in pelvis is usually accompanied by fractures. Snyder[15] noted that 32% of the knee dislocations had associated vascular injury. In our series 51.06 %

had fractures, 36.17% had muscle and tendon injury, 27.66% had skin loss, 25.53% had nerve injury and 17.02% had visceral injuries.

The significance of time interval between injury and the repair of arteries cannot be over emphasized because of the tissue anoxia and subsequent ischaemia. Snyder[15] reported that if the injured popliteal artery with knee dislocation was repaired within eight hours, the amputation rate was 13%, which rose to 86 % with progressive delay. Treiman[19] noticed that most of the limbs survived if artery was repaired with in nine hours of injury and none survived after the delay of fifteen hours. 59.57% of our patients arrived in the hospital with in six hours after trauma, In our cases 17.02% reported within six to twelve hours and 23.40% after twelve hours (Table - V). There was on average, delay of three hours before circulation to the distal part could be re-established.

Indications for surgery included uncontrolled bleeding[10] obvious open vascular injury, absent or weak pulse[5,14] expanding haematoma[3] and sensory or motor deficit[7,11]; the last requiring urgent surgery[7,11]. Pearce[11] has warned against the Surgery of occluded carotid artery due to trauma in the absence of neurological deficit because of the danger of emboli formation.

Rutherford[14] reported frequent presence of distal pulses in the presence of vascular injury. He also reported 42% false positive and 20% false negative exploration rates if done on the basis of clinical grounds alone. Sirinek[14] and associates reported 64% unnecessary explorations when done routinely for the proximity wounds. Therefore Menzoian[8] and Rutherford[14] advocate angiography in all cases. Blaisdell[1] however finds it more useful in cases of multiple level injuries and does not find it 100% accurate. Angiography is also advised under specific circumstances[1,5,9,10,15]. Thirty six patients with vascular injury underwent exploration in our series with 97.87% having weak or absent pulse, 38.29% with bleeding and shock, 17.02% with expanding haematoma and 21.27% with open visible vascular injury. We did not perform angiography routinely because of its non availability in our hospital in emergency and the risk of delaying reperfusion of the tissues. However in cases arriving late, we did get angiography to evaluate the status of collaterals, the level and type of vascular damage and future planning.

We did not get false positive or negative explorations, however; one patient, had a partial tear of the popliteal vein while the artery was being pressed by a bony fragment, thus causing absent distal pulse. In another cases radial artery was thrombosed with no evidence of the tear following firearm injury.

Average requirment of blood transfusion was four units (the range being from zero to thirty six units). Mucha[10] reported average requirement of 15.7, units blood in haemodynamically unstable patients with pelvic fractures and 3.4 units of blood for haemodynamically stable patients. We had two patients with vascular injury and pelvic fractures. One with common iliac artery and vein injury died on the table due to shock and uncontrolled bleeding inspite of transfusing thirty six units. The other required nine units of blood and ligation of the gluteal vessels; he survived.

Primary repair[5,7,8,9,11,19] of the injured vessel gives best results, however interpositional grafts[8,18,19] are also recommended in the specific circumstances.

Various types of interpositional grafts have been tried such as arteries[16], veins[8,18,19], intestinal submucosa[6], dacron[18] and poly tetra flouroethyline. Best results are achieved with arterial grafts but they are not available frequently for use. The second choice is saphenous vein graft. Its long term patency is better than the synthetic grafts (Feliciano[18]) and is reported to be 56% and 58% after five years in femorofemoral and femoropopliteal bypass surgery[17]. The basic problems with vein graft are due to its weakness in the presence of infection, which causes transmural necrosis and lethal bleeding. Shah[16] and associates lost three out of the four experimental animals, when interpositional vein grafting was done in the infected bed. The reported incidence[18] of interpositional vein graft thrombosis is 19.3 % and of bleeding is 4.5%. It has also the tendency to aneurysm formation in long term.

It is here that thomas[18] et al and Feliciano[18] propose the role of synthetic graft in the presence of infections as it leaks less often. However; Rich[18] found 100% failure of dacron graft in axillary, femoral and popliteal vessels. The over all rate anastomosis failure, as reported by Treiman[19] is 9.48 % and by Menzoian[8] 12.5 %.

We had failure rate of 28.57% in primary repair of the arteries and 35.7% in repairs with the interpositional vein graft. The overall failure rate was 31.03% which is significantly higher. However out of our nine failures, six had iatrogenic factors for failure. Two had malignancy and in one, vein graft was done in the tumor bed and in other femoral, saphenous and external iliac veins were completely blocked by tumor cell (malignant fibrous histiocytoma). The third one had crush injury of the artery with massive soft tissue loss and required bone shortening of 5 inches to anastomose the radial artery with ulnar end of deep palmar arch. These three were the cases of poor patient selection. In one patient, the artery was repaired under tension and bone was not fixed, thus thrombosed. In another patient a packing gauze introduced to treat gunshot wound was lying on the interpositional vein graft, thus causing infection and bleeding. The sixth had femoral

artery injury with fracture femur and was repaired with interpositional vein graft. He did well for three weeks; later during revision surgery, the graft was injured. It was repaired, however this trauma and tight bandage to control the bleeding caused its thrombosis and failure. Thus only three (10.34%) were genuine failures, the rest being either due to poor patient selection or due to poor management.

Whether drugs can improve the success rate of repair is controversial Gloviczki[4] reported that heparin can prevent thrombus formation but is not useful to dissolve the formed thrombus. We do not use heparin postoperatively routinely on the basis that if the anastomosis is good it does not require heparin and if it is poor, it requires revision, not heparin.

Ligation of the small[7,10] arteries is indicated to control bleeding particularly in the presence of good collaterals. Large[7,8] arteries can also be ligated under life threatening conditions, but the risks of gangrene[7,15,16] are to be considered. We ligated five arteries as a primary treatment Anterior tibial, (I) Posterior tibial (2), Ulnar (I) radial (I), Superior gluteal / inferior gluteal (I) Brachial (I) and Subclavian (I) without significant after effects except one.

Consesrvative treatment is associated with higher complication rate. Treiman[19] reported fourteen of the fifteen consevvatively treated patients has complications including eight death. Pearce[11] however suggests .conservative treatment for the completely occluded carotid artery in the absence of neurological deficit. We treated six (10.63 %) patients conservatively, all ending with complications (Gangrene (3), Arteriovenous fistula (1), Ischaemic contracture (I)). The last two had intermittent claudication. Primary amputation can be performed if the patient has severe associated soft tissue and bone injury with neurological[15] deficit or when gangrene is already established. We did five (10.63 %) primary amputations, four below the knee and one through the mid thigh. The overall amputation rate has reduced significantly from the World War-II. Franklin[3] reported 49% over all amputation in World War-II. Tremein[19] reported 13.5% amputation rate in 1966. The rate of the amputation following axillary artery was 43.2% (De Bakey and Simeone[7]) which came down to 1.5% during Vietnam war[7]. Popliteal artery presents the greatest challenge[3]. The amputation rate following the popliteal artery injury was 72.5% (Sullivan) in World War-II, 32.5% in Korean War (hudges and Jaenke[16])

The ligation or repair of the venous injury is controversial[13], though some advocate repair[6]. Hardin[5] has reported better results with the primary repair or ligation than with the interpositional graft. However the type of injury or locationt[15,16] is more deficit[7,11]. The last required urgent surgery[7,11] Pearce[11] however warned against the surgery of occluded carotid artery due to trauma in the absence of neurological deficit because of the danger of emboli formation.

Menzoian[8] reported 37.33% coincidence of fractures with vascular injuries. Subclavian, axillary and brachial artery injuries are usually associated with nerve[7,11,17] injury. The highest reported coincidence of axillary artery and brachial plexus injury 92%. McCready[7] also draws attention to the supracondylar fracture and brachial artery injury. Mucha[10] observed that the vascular injury in pelvis is usually accompanied by the fractures. Snyder[15] noted that 32% of the knee dislocations had associated vascular injury. In our series 51.06% had fractures, 36.17% had muscle and tendon injury, 27.66% had skin loss, 25.53% had nerve injury and 17.02% had visceral injuries.

We had over all 40% failure rate of our venous repairs (all primary). Whether they were due to primary arterial anastomosis failure or local factors is difficult to say as both patients required amputation.

Pearce[11] has warned against the Surgery of occluded carotid artery due to trauma in the absence of neurological deficit because of the danger of emboil formation.

The over all rate anastomosis failure, as reported by Treiman[19] is 9.48 % and by Menzoian[8] is 12.5%.

We had over all mortality rate of 10.61% with only one death caused by bleeding due to vascular injury. Three victims had severe associated injuries. One patient died of acute myocardial infarction post-operatively.

CONCLUSION

It is surprising that our results and those reported by many authors are comparable in the terms of sex, age, mechanism of injury, type of vessels injured, pattern of injury, associated injuries, results after ligation and conservative treatment and over all mortality rate. However there are significant differences also. The firearms used in our society are usually high velocity type, thus causing severe damage. The iatrogenic injuries were more frequent and out numbered the sharp penetrating injuries in our series. Significant number of cases arrived in our hospital very late, thus contributing to our failures. We did not have false positive or negative explorations. We had high failure rate and the fault lies with us and the hospital. Poor patient selection, lack of the facilities and poor coordination between various surgical teams were the main reasons for most of our failures. The higher amputation rate reflects our failures. We believe that by learning from our mistakes, we as well as other young surgeons can improve results which will be comparable to best ones.

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POST-FRACTURE AVASCULAR NECROSIS OF FEMORAL HEAD IN CHILDREN AND ADOLESCENTS

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ABSTRACT:
Avascular Necrosis of femoral head is a frequent complication following femoral neck fracture and traumatic hip dislocation in children with poor results. In a 10 years study of 100 femoral neck fractures in children at Jinnah Postgraduate Medical Centre, Karachi, 51% developed complications; avascular necrosis of femoral head, coxa vara, non union and premature capital epiphyseal fusions. Age range of children were 1-17 years. Followup period ranged 18 months to 8 years. Avascular necrosis was the most debilitating complication which was seen in 35% cases. Avascular necrosis developed in 21% children managed non-operatively and in 43% managed with internal fixation. The aim of study was to evaluate the role of different factors, like; Age of patient, nature of traume, type and displacement of fracture and method of management in causation of avascular necrosis and to draw conclusion for better method of management with minimum incidences of avascular necrosis.

KEY WORDS: Children, Femur Neck, Avascular Necrosis

INTRODUCTION

Avascular Necrosis of femoral head following fracture neck femur or hip dislocation is the most debilitating com avascular necrosis of femoral head is remarkably higher in children compared to adults. It has also been observed following basal femoral neck fractures and inter-trochanteric fractures in children contrary to similar fractures in adults[1]. Various factors has been attributed to the causation of this complication including changes in pattern of vascular supply of femoral head with increasing age. Severity of trauma, initial displacement of fracture and type of fractures has been observed to be directly related to incidences of avascular necrosis[2,3]. These factors are not in the control of the surgeon, whereas the mode of treatment which is in control of surgeon do not significantly affect the rate of complication[2,4,5], unless it is not carried out properly, and with due care to prevent further vascular damage by careful handling. The results of hip fracture in children are good only if they escape complications however, the problem of how to prevent the complications remains largely unsolved. This study is conducted to evaluate different factors leading to avascular necrosis and to suggest better method of management to minimize complications.

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MATERIAL AND METHODS

A prospective study on femoral neck fractures in children and adolescents was carried out in department of Orthopaedic Surgery, Jinnah Postgraduate Medical Centre, Karachi. Study was based on 100 cases treated during ten years from April 1987 to March 1997. Patients included in study were aged between 1-17 years, with femoral neck fracture of less than one week duration of trauma. Patients who come late, (after one week[6]), were excluded from the study. Patients with slipped capital femoral epiphysis, Pathological fractures, Sub trochanteric and Intertrochanteric fractures and adolescents with fused capital femoral epiphysis were also excluded.

All the patients were admitted in the hospital and put on skin traction with counter traction and derotation P.O.P. boot, irrespective of type and displacement of the fracture. 38 patients, were managed non-operatively, that were below 5 years age, had undisplaced or mildly displaced fractures, they were continued on skin traction for 1-2 weeks. They were subsequently put on double leg hip spica cast for 6 weeks, then in single leg spica cast for another 4-6 weeks until fracture showed signs of complete union. 62 patients were operated and internal fixation done within 3-5 days after trauma. 43 fractures were reduced by close method of lead better technique[7], whereas 19 fractures needed open reduction (anterior capsulotomy), due to failure of attempted closed reduction. None of these patients were operated in emergency (Table I & II). Moor's pins were used in all type I fractures,

TABLE I CORRELATION OF AVN* WITH TYPE AND DISPLACEMENT OF FRACTURE

Type	Degree of displacement			AVN	
	Undisplace (n=33)	Mild (n=44)	Gross (n=33)		
I (n=6)	--	4	1	5 (83.3)	
II (n=42)	--	6	11	17 (40.4)	P<0.01
III (n=52)	--	6	7	13 (25.0)	
	--	16 (36.3)	19 (57.5)		

*AVN = Avascular necrosis of femoral head
Figurs in parentheses are percentages.

TABLE II CORRELATION OF AVN* WITH METHOD OF TREATMENT AND IMPLANTS

Treatment	No.	AVN*	
Non-operative	38	8 (21%)	
Operative	62		
Close RIF**	43	12 (27.9%)	
Open RIF	19	15 (78.9%)	P<0.01
Implant used for fixation			
Compression screw	34	20 (58.3%)	
Moor's/knowle's pin	28	7 (25%)	P<0.01

*AVN = vascular necrosis of femoral head
**RIF = Reduction and internal fixation

TABLE III CORRELATION WITH AVN* WITH AGE GROUP OF PATIENTS

Age group (yrs)	No.	%
0 - 3 (n=5)	--	
4 - 8 (n=16)	7	34.75
9 - 13 (n=47)	19	40.4
14 - 17 (n=32)	9	28.12

*AVN = Avascular necrosis of femoral head

and displaced type II & III fractures in patients aged less than 10 years. Whereas Screws were used in all type III fractures and in displaced type II fractures in patients above 10 years (Table II). Fractures fixed with Moor's pins were further stabilized in single leg hip spica cast for 3-4 weeks.

Patients were allowed walking with crutches and non weight bearing on affected limb after removal of spica cast. They were allowed gradual weight bearing after 10-12 weeks post operation. Weight bearing was delayed in patients with signs of avascular necrosis on conventional periodic radiography confirmed on TC 99 Bone Scan. Clinical and Radiological evaluation was carried out fortnightly for 8 weeks, then monthly for 12 months and then every 3rd month.

RESULTS

The total number of patients in the study were 100, male to female ratio was 1:1.5. 75% children sustained femoral neck fracture due to severe trauma (fall from height greater than 5 feet and road traffic accident), 25% children had fracture with mild trauma (fall while playing on level ground and fall from cot). 6 children had Delbet's[2] Type I fracture, 42 had type II and 52 had type III femoral neck fracture. 23 fractures were undisplaced. 44 fractures were mildly displaced and 33 were grossly displaced (Table I). On Ratliff assessment Scale[8], 60% patients behaved good, 19% behaved fair and 21% behaved poor. The fair and poor results were due to development of avascular necrosis of femoral head, coxavara and non-union. Avascular necrosis (AVN) of femoral head developed in 35% patients, 4 of these were associated with coxavara, whereas 4 other had associated coxavara and non union. Premature capital epiphyseal fusion associated with avascular necrosis was seen in 6 patients.

Avascular necrosis was seen in 83% of type I fractures, 40% of type II and 25% of type III fractures. AVN developed in 57% of grossly displaced fractures and 36% of mildly displaced fractures, whereas none in undisplaced fractures (Table I). Avascular necrosis of femoral head developed in 21% of patients who were managed conservatively, whereas it was seen in 43.5% fractures managed operatively. Among operated patients, avascular necrosis was seen in 79% of patients requireing open reduction compared to 28% reduced with closed method. AVN developed in 59% fractures fixed with cancellous screw, majority of them were type II fractures, whereas AVN developed in 25% fractures fixed with Moor's pins, majority of these were type I fractures (Table II). The age of patients did affect the incidence of avascular necrosis. The age group between 9-13 years were affected more (40.4%) than age group 14-17 years (28%).

Minimum followup period was 18 months and maximum 9 years. Hip arthodesis as a secondary procedure was carried out in 5 patients at ages between 22-25 years. Who had developed disabling Osteoarthritis due to avascular necrosis of femoral head following femoral neck fracture in adolescence or childhood.

DISCUSSION

Avascular Necrosis (AVN) of femoral head is the most frequent and debilitating complication of femoral neck fracture in children and adolescents. To compile an accurate statistical data on AVN is difficult due to number of variable factors involved in its causation. Incidences of post fracture avascular necrosis of femoral head are remarkably high in children compared to adults. The problem is worse in younger than older children due to: (a) intra articular position of nutrient vessels of femoral head, (b) presence of insufficient femoral arterial supply between 3-8 years of age. (c) presence of impermeable barrier of epi-

physeal plate and (d) high risk of lateral epiphyseal vessels occlusion with femoral neck fracture. Superior retinacular vessels are usually torn if fracture is displaced by one half the diameter of femoral head.

The degree of displacement has been found to be the main factor leading to high risk of AVN in grossly displaced fractures[5,8-10]. AVN has exceptionally been seen in undisplaced fractures[8]. Similarly in this study none of the undisplaced fractures developed AVN. Whereas, AVN developed in 57% of grossly and 36% of mildly displaced femoral neck fractures. AVN has been frequently seen in basal neck femoral fractures in children in contrast with adults. However, the incidence of AVN occurs with decreasing rate in type I, II and III femoral neck fractures[2,8,13,15]. Similarly, in our study AVN developed in 83% of type I, 40% of type II and 25% type III fracture (P<0.01). The lower incidences of AVN in type III fracture has been attributed to better anastomotic vascular channel at distal portion of hip joint capsule[1,2,10].

The age between 11-13 years is considered a transition period, when anastomosis between foveolar and retinacular vessels takes place across the epiphyseal plate. This has been the explanation for lower incidence of AVN after 13 years and higher before 11 years of age in majority of published series[6,8-11]. In our study findings were little different; 40% AVN occured in age group 9-13 years, 34% in age group 4-8 years and 28% in age group above 13 years. That the AVN was more in ages of transition period (11-13 years) than beforre or after this period. This may be due to the severe insult to developing anastomosis across epiphyseal plate at the time of injury.

AVN following femoral neck fracture in children and adolescent seemingly occur regardless of the type of treatment and implants used for fixation[2,4,5,7,10,12-19]. The insult to vascularity of femoral head mostly occurs at the time of trauma[2]. Whereas, further damage can occur with improper attempt at closed reduction and improper method of fixation, which leads to distraction at fracture site, malrotation and subluxation. Therefore, meticulous handling and preserving the periosteal ratinaculum that connects head and neck and the pilot hole preparation (Drill+taping) has been made mandatory prior to insertion of pins/screws/nails for fixation to prevent considerable torque during fixation with implant[5,18,20]. Various screws, multiple pins fixation devices with side plate compared with nail and multiple pins fixation without side plate have reduced the incidences of non-union in adults[5], but the reports differ as to whether they also significantly reduce the incidences of AVN in children. In our study AVN developed in 58% fractures fixed with compression screw compared to 25% of fracture fixed with 3-4 Moor's pins. Three patients with Moor's pin fixation and one with screw fixation had developed associated non-union. The error in these cases were the persistant distraction at fracture site. 54% (19 cases) of avascular necrosis of femoral head were of type I, 25% (9 cases) had type II, whereas 20% (7 cases) had type III. All type II and type III AVN were observed in fractures fixed with compression screw. Type II necrosis were due to eccentetric fixation, screw tips were lying at lateral or medial subchondral areas causing damage to physis. Our findings were contrary to the statements of Clandruccio[5], that the incidences of AVN with impaction screw fixation of femoral head has been found approximately the same as that following less stable fixation with nail or pins.

The vascular compromise has also been noted in undisplaced fractures managed non-operatively by keeping the leg in internal rotation, extension and adbuction[5,21]. Therefore, mild flexion abduction and external rotation posture of leg has been adviced for patients treated conservatively[5,12,21]. The affects of urgent aspiration of hip in undisplaced fractures managed conservatively, early open reduction (with capsulotomy) operation within 6-8 hours and the evaluation of the additional damage, if any produced by delay between time of fracture and operation has been debated in various publications, that lack statistical data of significance and concludes without concensus[3,5,6,13,16,20]. However, latest experimental studies of Gerber etal and Canale[18,19] has remarked that immediate open reduction and internal fixation may not prevent AVN after displaced type II and type III fractures, and they not favour immediate aspiration or decompression and fixation as the resultes were same whether to be operated on same day or after 2 days.

The initial physical symptoms of AVN are pain and limitation of motion secondary to synovitis. The radiographic signs may be seen as early as 6 weeks of fracture, whereas, the MRI and radioisotope scanning are of more value to detect AVN within 24 hours of injury. However, after two weeks or more reactive changes remarkably maskup the presence of AVN[18,22]

The prognosis of AVN is directly related to severity of involvement of femoral head i.e. type of AVN and to the age of the child[12,13,18]. The type I necrosis, that occur in majority is related to higher percentage of poor results, whereas type II and II necrosis have better results. The younger child have higher percentage of better results, primarily because of ability to repair or remodel the necrotic bone.

No effective treatment is known or proposed for post fracture AVN of caput femoris, the number of approaches has been described but no one method is found completely satisfactory. Treatment recommendation for AVN include bed rest, avoidance of weight bearing for prolonged period

to prevent collapse of head and subsequent osteoarthritis[18,20]. Patients younger than 18 years of age who have post fracture AVN usually do not have sufficient pain to warrant surgery for many years. The early surgical procedure can be soft tissue releases about the hip for flexion abduction contractures and osteotomies. Arthrodesis and arthroplasty are the treatment of choice for end stage of the disease when joint has been completely damaged and symptomatic[20,21] this has been carried out in 5 patients included in this study. For children less than 10 years old with type I AVN, removal of fixation device, followed by the use of an abduction containment orthosis for 12 months may produce acceptable results[13,18].

On the basis of above study following conclusion can be drawn that: The age of child, type of fracture and degree of displacement has been found directly related to incidences of AVN; these are factors which the surgeon is helpless to control. Whereas, the proper handling, prompt and particular treatment are definitly in the hands of surgical team. The categorical statement cannot be made about the best method of treatment for displaced fractures. The selection of proper treatment for every case need awareness of incidences of AVN in all cases and coxavara in non-operated cases. However, published literature today is in favour of operative management for children with all displaced fractures and in children above 5 years age[13,18,22]. The choice of implant shall be decided in reference to type of fracture, age of the child and size of the femoral neck. The prevention of AVN in these fractures are almost synonymous with prevention of displacement[5]. The prevention of trauma seems to be the only major factor to prevent AVN but to prevent trauma is not always so easy to do.

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NON-ULCER DYSPEPSIA AND GASTRITIS-CLINICAL ASPECTS

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ABSTRACT:
A prospective clinical study was conducted in Medicine Unit III, Jinnah Postgraduate Medical Centre, Karachi from January 1996 to September 1996 to determine the endoscopic findings in patients with epigastric pain. After complete clinical examination endoscopy and gastric antral biopies were done in all cases. There was a strong co-relation of clinical, endoscopic and histological findings as in 0.58 (82%) of cases with NUD was there evidence of gastritis.

KEY WORDS: Gastritis, Non-ulcer dyspepsia (NUD)

INTRODUCTION

In a large number of patients with epigastric pain often after appropriate investigations no cause can be found. Epigastric pain can be due to a large number of structural and functional disordes related to stomach, duodenum pancreas and hepatobiliary system. The typical sites of pain caused by different disorders are well known, but variations occur. The introduction of modern techniques has made definitive diagnosis possible for most of the patients[1].

The term gastritis has various connotations. To the endoscopist it may mean visible gastric mucosal abnormalities, to the pathologist gastric mucosa infiltrated with inflammatory cells, to the clinician aspirin induced haemorrhage of the upper gastrointestinal tract and to the patient, epigastric pain. The term gastritis can simply be defined as inflammation of gastric mucosa; inflammation may be predominantly acute (or active) with poly-morphonuclear cell infiltration, chronic with lymphocytes and plasma cells predominating or a combination of both.

In this study non-ulcer dyspepsia (NUD) implies the occurrence of dyspeptic symptoms when no ulcer can be found in oesophagus, stomach, or duodenum, not previously subjected to surgical operation. Patients with anemia, hiatus hernia, oesophageal reflux, carcinoma of stomach, hepatic, biliary, pancreatic.intestinal, renal or cardiac disease were excluded by history, clinical examination, laboratory investigations and by ultra-sonography and radiologically.

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Another factor which is certainly common in dyspeptic patients is an infective agent, Campylobacter pylori. It has been suggested that this bacteria may cause non-ulcer dyspepsia as chronic gastritis is found histologically in upto 50% of patients with these symptoms[2].

According to Colin Jones[3] any patient experiencing upper abdominal symptoms might reasonably be asked to have an endoscopic examination especially for gastric and duodenal symptoms. Gross gastroscopic examination is not sufficient since many lesions need histological identification.

Barium X-rays have been used as a common technique for the diagnosis of gastrointestinal lesion. According to Gower[4] barium meal may not be the final answer in lesions of stomach and persistent dyspepsia. Thus most of the radiologists advise endoscopic examination, in their reports.

MATERIAL AND METHODS

70 Patients with dyspeptic symptoms attending the G.I. Clinic, Medical Unit-II between June - September 1996, Jinnah Postgraduate Medical Centre, Karachi were studies. Following symptoms were asked for including the patients for endoscopic study.

- Pain after eating.
- Flatulence.
- Vague epigastric distress.
- Nausea and / or Vomiting.
- Weightloss.
- Nervousness.
- Pyrosis

Seventy patients who complained of these symptoms

were included for investigation of dyspepsia. They were given an appointment letter for one visit Upper Gastrointestinal Clinic in Medical Unit-III of JPMC. The letter contained the time of appointment, a short explanation of the clinic arrangement, a simple description of endoscopy and a request to attend fasting. Patients seen at morning clinics fasted from previous evening. The clinics were conducted by two doctors and an endoscopy nurse.

Biopsy material was obtained from three to five different sites on the gastric mucosa, preserved in formalin and sent to the pathology department of JPMC.

Patients were told the results of endoscopy immediately after the procedure and sent home as no sedation was given in most cases, with an advice to collect their biopsy report after ten days.

RESULTS

Out of seventy patients with NUD, from whom biopsy specimens of the gastric mucosa were obtained, 46 (65.7%) were male and 24 (34.3%) were female. The mucosa was classified as normal in 12 (17%). 20 (28.5%) were placed in miscellaneous group, 21 (30%) were classed as chronic superficial gastritis and 17 (24.3%) were classed as chronic atrophic gastritis. There were no examples of gastric atrophy in the series. Twenty two cases (about 31%) were positive for Helicobacter pylori organism.

Thus in 58 (82%) cases rather more than three fourths of the patients examined there was evidence of gastritis. Of these 20 (28%) cases had only minor changes (miscellaneous group). Major abnormalities were found in about 38 (54.3%) of the whole NUD group.

TABLE I GASTRIC MUCOSAL FINDINGS IN 70 PATIENTS WITH NUD ACCORDING TO AGE AND SEX ON HISTOLOGICAL BASIS

Age in years	Males					Females				
	N	M	SG	AG	T	N	M	SG	AG	T
10 - 19	--	1	--	--	1	--	1	2	--	3
20 - 29	4	--	3	2	9	2	1	3	1	7
30 - 39	1	6	1	1	9	--	1	--	--	1
40 - 49	--	3	4	3	10	--	3	--	3	6
50 - 59	4	1	1	2	8	1	2	1	2	6
60 - 69	0	1	2	2	5	--	--	--	--	--
70 - 79	--	--	1	1	2	--	--	--	--	--
80 - 89	--	--	2	--	2	--	--	1	--	1
Total	9	12	14	11	46	3	8	7	6	24

NUD = Non-ulcer dyspepsia; N = Normal; M = Miscellaneous minor mucosal changes; SG = Chronic superficial gastritis; AG = Atrophic gastritis; T = Total.

Sex

The distribution between the sexes of patients with normal and different type of abnormal mucosa is shown in Table I. Fewer women than men were examined, chiefly because fewer of those who presented suffering from NUD and more declined the examination or were unsuitable for it. Males predominated in about the same proportion in each decade except in the first. Fewer women than men showed severe changes.

Age

The mucosal findings are grouped according to the age of the patients in the table. Most of the patients who presented were in the second to fifth decade at the time of examination. Atrophic gastritis is commoner in old age; but in our study it was also found in the third, fourth and fifth decade of life. Few patients were examined in sixth and seventh decade and they showed chronic gastritis and atrophic gastritis.

There was another study done by A.W. Williams[5], in which 200 patients were with NUD from whom biopsy material was obtained and examined.

A comparison of the studies is as below in the Table II

TABLE II COMPARISON OF THE STUDIES

Study	Male	Female	N	M	CG	AG
A.W. Williams (200 patients)	134 67%	66 33%	90 45%	72 36%	9 4.5%	20 14.5%
Presnt study (70 patients)	46 65%	24 34%	12 17%	20 28.5%	21 30%	17 24.3%

N = Normal; M = Miscellaneous; CG = Chronic gastritis; AG = Atrophic gastritis.

In our study more patients were diagnosed as chronic gastritis and atrophic gastritis, and they also belong to younger age group. This is probably because of eating habits and environmental factors.

DISCUSSION

The availability and widespread use of flexible endoscope has revolutionized clinical and pathologic approaches to the disease of upper gastrointestinal tract. Biopsy examination provides better correlation between symptomatology and morphology, because histological examination can detect many lesions which are missed on gross endoscopic examination. On the other hand, histological examination provides restraint to the endoscopists in over-diagnosing the lesions. Thus histopathological examination of endoscopic biopsy could be a better source of determining true frequency of different lesions in the upper gastrointestinal tract.

CONCLUSION

In conclusion, it may be submitted that in great majority of the patients studied, the common symptoms related to the upper gastrointestinal tract were associated with some physical lesions. In only about 17% cases no significant changes were seen, that is to say that possibly the symptoms of these patients were functional. Secondly, although endoscopy is a highly useful technique in the diagnosis of the disease of upper G.I. tract, it should be used in combination with histopathological examination of biopsy material.

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SPONTANEOUS RESOLUTION OF A HEMANGIOMA IS A BETTER OUTCOME: EXEMPLIFIED WITH FOLLOW UP OF FOUR CASES

Case Report

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ABSTRACT:
Hemangioma is a common birth defect, management of which is still controversial. Three cases of hemangiomas, two of which were extensive lesions, regressed over a period of time with good cosmetic outcome. One smaller lesion is showing signs of regression and is expected to involute completely. We therefore advocate conservative approach for best cosmetic results in uncomplicated cases.

KEY WORDS: Hemangioma, Expectant Management.

INTRODUCTION

Congenital vascular malformations occur in 20-40 percent of newborn infants and comprise the largest group of neoplasms in children, with hemangiomas being the commonest. Though 10% of children under the age of one year have hemangioma, only 2.6% of these are detectable at birth[1]. Hemangiomas are benign neoplasms which result from failure of establishment of communication of islands of angioblastic cells with the adjacent vascular system. Sometimes there is canalization and re-establishment of communication between the angioblastic tissue and the vascular system which results in rapid increases in the size of lesion. This feature is usually seen in capillary hemangiomas, the commonest of hemangiomas. These hemangiomas are also characterized by a slow period of involution resulting in fibrosis and diminished cellularity. They differ from the vascular malformations which are hamartomas composed of mature endothelial cells that neither proliferate nor involute[2].

Most of the hemangiomas are known to resolve spontaneously within the first few years of life. There is a lot of controversy about the treatment of hemangiomas, though most agree that the best option is to allow the hemangioma to resolve on its own. This, however, applies only to uncomplicated cases. Deferring treatment in complicated case can lead to serious and at times life threatening complications.

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CASE REPORTS

Case #1

A baby boy presented with extensive congenital disfiguring hemangioma involving almost one half of the face including the forehead, eyelids, cheek, nose and lips. (Figure 1). In the course of involution he developed extensive lacerations and bleeding which were treated conservatively with blood transfusions and compression dressings. Over a period of time complete involution of hemangioma occured (Figure 2). The child is now of two years age and further improvement in cosmetic appearance is expected in years to come.

Figure 1 Extensive capillary hemangioma which underwent rapid proliferation and ulceration during the initial few weeks of life (perhaps this is why it is termed filed fire by some).

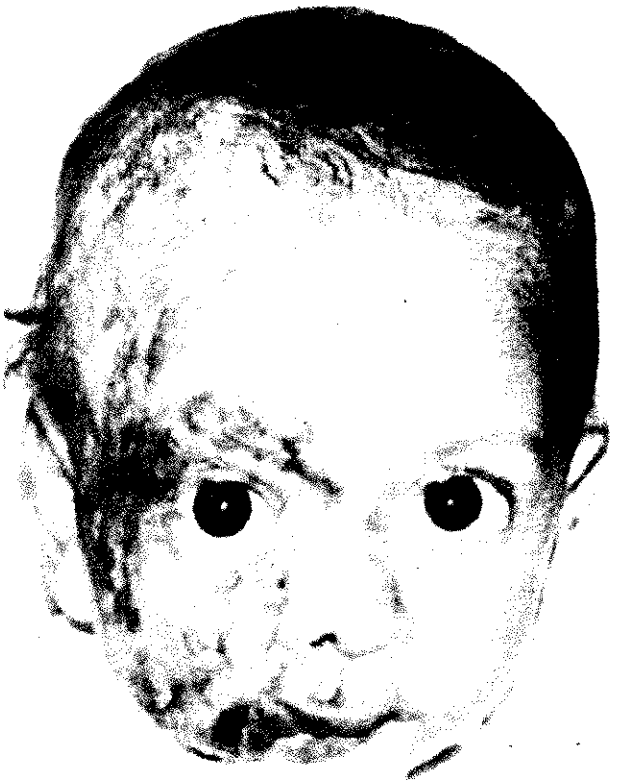

Figure 2 The same patient as in Fig. 1 showing remarkable regression at the age of two years.

CASE # 2

A two months old girl presented with extensive capillary hemangioma involving the neck, chin, cheek and the lower lip with intraoral extension (Figure 3 and 4). We employed periodic injection of 10% hypertonic saline. Gradual regression of the hemangioma took place and only minor surgical procedures were carried out to excise loose and lax skin under the cheek and to provide better cosmetic appearance to the lower lip (Figure 5). Further improvement has taken place with time. Residual scars are negligible.

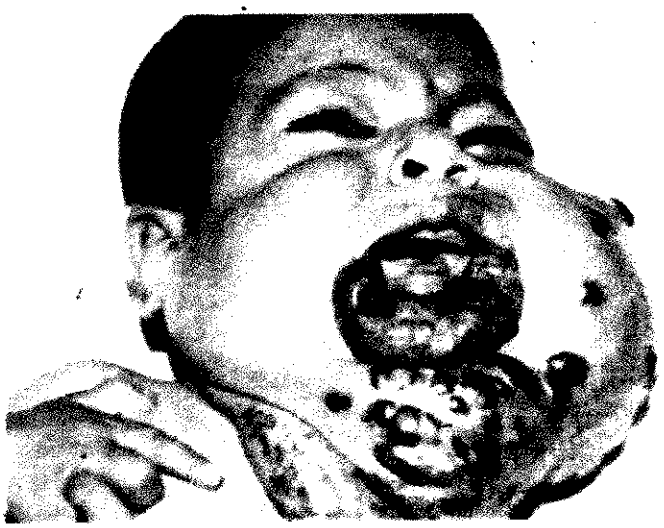

Figure 3

Figure 4

Figure 3 & 4 Two month old girl with capillary hemangioma which later under went spontaneous regression assisted with periodic injection of hypertonic saline solution.

Figure 5 Same patient as in Fig. 3 & 4 at the age of three yea[illegible] fairly pleasent appearance.

CASE # 3

A female child with a small hemangioma on the medial aspect of the thigh near the groin area at birth, presented with rapid increase in the size of the lesion in the first few months of life. The child was followed up with no active interventions and fortunately the hemangioma started showing signs of regression in the seventh month (Figure 6). By the time the child was two and a half years old, the hemangioma had completely regressed leaving behind minimal scar.

CASE # 4

A neonate was born with a tiny hemangioma on one side of the upper lip. It grew in size in the first year of life. The child was regularly followed-up. The lesion is showing signs of involution (Figure 7) and is expected to regress completely.

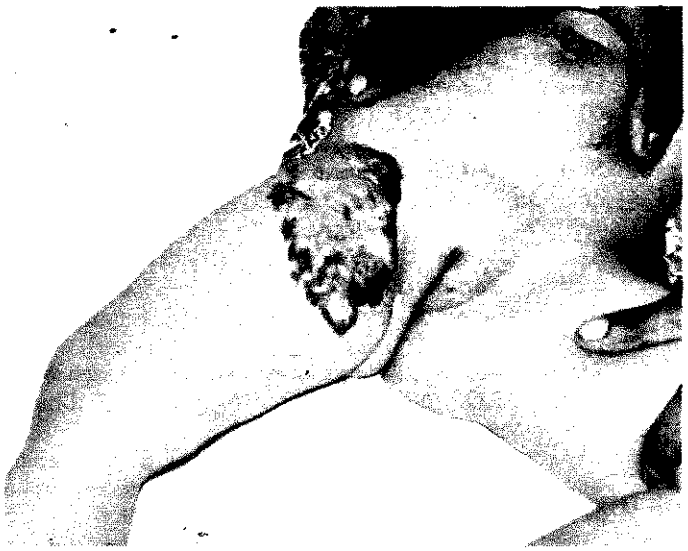

Figure 6 Hemangioma, in this girl grew with in a few months to acquire quite a large ssize. Rapid growth resulted in an equally rapid resolution as seen by central whitish avascular looking areas on the surface. Further progression with ulceration and involution of the lesion resulted in barely discernable scarring.

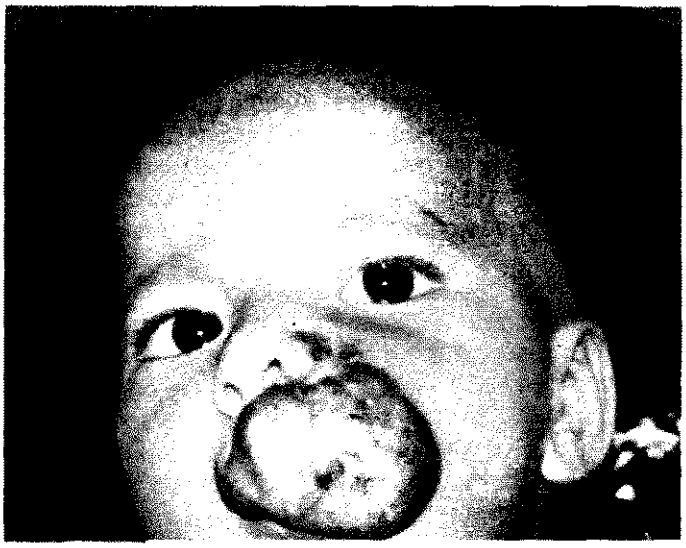

Figure 7 Hemangioma of the upper lip and the cheek after the initial rapid growth has started involuting as evidence by central whitish areas which results due to thrombosis of the vessels in the lesion. Surgical excision would have resulted in the loss of upper lip and considerable disfigurement.

DISCUSSION

Hemangiomas usually present at or soon after birth. They may be single or multiple and vary considerably in size. Hemangiomas result due to continuous development of the cutaneous microvasculature during the first 3-4 months of life. Therefore, aberrant new vessel growth may occur during this time of dynamic vascular reorganization[3]. A fully formed vascular lesion in a new born is more likely to be a vascular malformation, while those developing during the neonatal period are more likely to be hemangiomas. The hemangiomas usually grow with the patient for the first few months of life, sometimes the growth accelerates, specially if the lesion is present on the face or the neck. This occurs due to filling up of the endothelial spaces within the developmental nest with blood. Those that grow rapidly are usually the ones with most potential for involution.

Juvenile hemangiomas are capillary hemangiomas and are further classified on the basis of their feature. Involuting hemangiomas are raised above the surface and comprise of capillary (strawberry nevus) and cavernous hemangiomas[1]. The capillary hemangiomas are superficially placed in the skin and are characterized by capillary proliferation. The cavernous hemangiomas penetrate deeper in the dermis and sub-cutaneous tissue and are made up of larger blood-filled spaces. Involution and regression are more common and less complicated in case of capillary hemangiomas. Cavernous hemangiomas, on the other hand, have a lower potential for regression and are likely to cause complications.

It has been suggested that the basic fibroblast growth factor is the primary angiogenic factor responsible for growth of hemangiomas. The angiogenic factor heparin and histamine released from the abundant mast cells, present in the proliferating hemangiomas, augment this process[3]. The period of growth is followed by a stationary period of variable length. Involution is characterized by clinically documented growth and subsequent regression, histological evidence of normal number of mast cells and of fibrosis and fat deposition. At times, involution is coincident with growth; central thrombosis with necrosis occurs with spontaneous regression during the first two years of life, often leaving minimal residual scar. High levels of estrogen receptors have been measured in proliferating hemangiomas, which may explain the preponderance of girls with these lesions[5].

More than 90% of the hemangiomas resolve without complications or significant cosmetic deformity. Beginning of the involution is observed during the second year of life for the strawberry portion of a lesion and more than 50% resolve by the fifth year of life. Several studies have shown that upto 75% disappear by seven years of age and 90% are gone by the age of ten (approx 10% per year)[4].

Parents overtly concerned about their child's cosmetic appearance must be informed about the risk of scarring with therapeutic interventions. Scarring, on the other hand, is only minimally and uncommonly encountered if the lesion is left for natural resolution. The final result in untreated lesions is generally far superior than that obtained by therapeutic intervention[6].

Aggressive therapy is indicated only in a few complicated cases. These are either the large hemangiomas which

involve the vital organs or produce life threatening complications. In these cases results without treatment are unacceptable with mortality being as high as 20%.

Intervention usually is reserved for life-threatening or vision threatening lesions. Surgical reconstruction may be indicated for the 10-30 percent of hemangiomas that do not regress adequately[7,8]. Future mode of treatment of superficial hemangiomas includes lasers which selectively destroy ectatic dermal vascular tissue through intact epidermis[9]. Another newer investigative modality is subcutaneous injections of Interferon alpha 2a for treatment of complex hemangiomas of infancy and childhood[10]. Biologic suppression by anti-angiogenic factor is also a mode of therapy under investigation[8]. Despite these new modalities, conservative treatment is thought by most to be the option of choice in uncomplicated cases especially cases that are entirely or largely capillary in nature.

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GIANT CONGENITAL NEVI OF BATHING TRUNK DISTRIBUTION

Case Reports

FOUZIA SHAFIQUE, FARAH KHAN, ATHAR QURESHI, SYEDA BATOOL, TASLEEM ANWAR, WAHEED HASHMI, ZAKIR KHAN, M. NAEEM KHAN

ABSTRACT:
An experience with six cases of giant congenital nervus of bathing trunk distribution, seen at The Children's Hospital, Pakistan Institute of Medical Sciences (PIMS), Islamabad between the years 1987-1997 is presented. In four cases surgery was carried out with good results .Two patients were not operated because parents refused to give consent. One lesion was found to be malignant. Each intervention is recommended to achieve good results, which also decreases the chance of malignant transformation.

KEY WORDS: Bathing trunk nevi, Malignant Melanoma, Variable malignant potential, Staged surgery.

INTRODUCTION

Giant nevi are being studied extensively. There is a potential risk for malignant transformation. Though a strong association has been shown between giant nevi and malignant melanomas[1-4], there are reports of origin of spindle cell, undifferentiated tumors[5] and ependymoma[6] in the giant nevi, as well.

Giant congenital nevus, a variant of the nevocellular nevi, is an uncommon lesion seen with the frequency of less than one case in 20,000 live births[7,8]. It has been referred to by different terms on the basis of its size, site, the depth of the lesion and its constitution. If the lesion is more than 20 cms in diameter[9], it is appropriate to call it a giant nevous and it may not be feasible to remove the nevus in a single operative procedure[10].

CASE REPORTS

Case #1

A twenty days old male neonate presented with a large pigmented lesion occupying the back of the trunk extending up to the nape of the neck, both the shoulders and proximal upper arms. The surface of lesion had multiple nodular cystic / semisolid tumorous lesions of varying sizes. There were areas of ulceration on the hump of the largest lesion with secondary infection (Figure 1). In addition, satellite pigmented nevi were present all over the patient's body, through none of these had nodules similar to the primary lesion. No other abnormalities were detected clinically.

A biopsy done from the peripheral area of the nevus elsewhere was reported as an intradermal nevus. However, because of the clinical appearance and the presence of multiple solid lobulated lesion, a repeat biopsy was done which revealed it to be a malignant melanoma. Excision was performed in stages. In the first stage only the smooth areas of the margin of the lesion were not removed. A very wide defect thus created was covered with meshed skin (Figure 2). During the next three weeks the fringes of the nevus were also excised in three stages.

CASE #2

A two and a half months old male child presented with giant hairy nevus of typical bathing trunk distribution with

Figure 1 Deep dermal nevus in a new born involving the entire back and extending to the neck, shoulder and anterior chest on one side. Several solid multi-lobulated lesions of varying sizes are seen on the involved area. These proved to be malignant melanoma.

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Figure 2 Same patient in Fig. 1. Immediate appearance after palliative excision of most of the lesion and sking grafting is planned in near future.

Figure 3

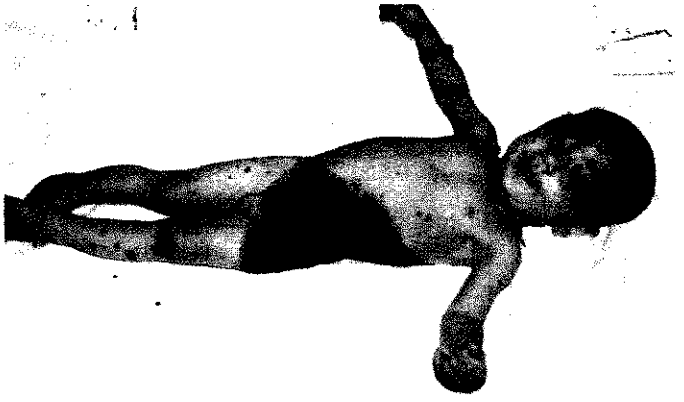

Figure 4

Figure 3 & 4 A male child of two and a half months of age with congenital deep dermal nevus of typical bathingg trunk distribution with multiple satelllite lesions and no detectable meningeal and neurological involvement.

irregular surface. The patient had multiple satellite lesions all over the body (Figure 3 & 4). CT scan was carried out to rule out asymptomatic nervous system involvement. It was reported to be normal. Treatment was commenced in stages by excising the peripheral aspect of the nevus and by pulling and mobilizing the adjacent skin to cover the defect. The treatment was well on its way and most of the lesion was removed. The patient was lost to follow-up.

CASE #3

An eight months old male child presented with seizures. One examination he was found to have a typical bathing trunk nevus covering the buttocks, lower back and upper thighs. There were also a number of satellite lesions all over the body (Figure 5). The main lesion had rough surface and growth of hair, these features were not seen on the satellite lesions. Surgery was not considered because of the symptomatic involvement of the central nervous system.

CASE #4

A three years old boy presented with giant hairy nevus covering the middle of the back and the anterior abdominal wall. There was a history of repeated surface infection of the nevus. After local and systemic treatment of the infection, surgery was performed. Skin expansion technique, by using five large fate's catheters, was employed (Figure 6). This was followed by excision of the nevus and approximation of the skin. Post operatively the patient developed minor dehiscence of the wound but good healing of the wound occurred on application of mercurochrome.

CASE #5

An eleven years old boy was brought to us for the treatment of undescended testes. On examination the child

Figure 5 Eight month old child with bathing trunk nevus and scattered sattelite lesions with scalp involvement. Presented with repeated episodes of seizures due to meningeal involvement. Also had encephalitis like illness.

Figure 6 Child with large congenital nevus on the back. Note the application of multiple folley's catheters with blown up balloons to acquire rapid skin expansion.

was found to have a large pigmented lesion with hairy growth. Clinically the lesion was diagnosed to be a congenital giant bathing trunk nevus, with presence of age related changes in the lesion, including dark pigmentation and hard brittle hair. The parents refused surgery for the lesion.

CASE #6

A case of a four month old baby boy presented with bathing trunk nevus bottom and several satellite lesions all over the body, including the scalp (Figure 7). The child was admitted and the lesion was completely excised in stages. The defect was closed using skin graft. The perianal portion of the lesion could not be removed because of being technically inacessible.

DISCUSSION

There are a few reports of presence of large papules and swellings on the surface of the nevus at birth. With time the pigmentation darkens, the surface becomes irregular and papules and hair appear on the involved area[11,12]. The sites commonly involved are the extremities, trunk, head and neck. Rarely the mucosa of palate, bulbar conjunctiva and tongue may be involved[13]. Through no sex preponderance has been reported in most of the literature, a prospective study currently being carried out shows a female/male ratio of 3:2[13]. In our series all cases were males.

Histologically the giant congenital nevus has all the features of a congenital nevocellular nevus[14]. In a recent study, 89% of all congenital melanocytic nevi involved the lower half of the reticular dermis and 51% infiltrated the subcutaneous tissue. Depth of the nevus cell infiltration correlated positively with size of the lesion[15,16].

The exact etiology of the giant congenital nevus is not known. However, higher numbers of nevi and cafe-au-lait spots seen in the families of affected children suggests a hereditary component[13]. An increase in the incidence of

Figure 7 Typical bathing trunk nevus with satellite lesions all over the body. Note rough, irregular and thickened surface of the lesions.

influenza-like illness and severe nausea and vomiting in the mothers of these children during pregnancy has been noted[13].

A giant nevus located in the head and region may be associated with leptomeningeal melanocytosis (a rare combination referred to as neurocutaneous melanosis)[17] and secondary neurological disorders like epilepsy[18]. Lesion over the vertebral column may be associated with underlying spinal defects such as meningomyelocele, spinal bifid and hydrocephalus[1,17,18]. A higher number of common moles and cafe-au-lait spots are a common finding as well[13].

Giant congenital nevus presents a difficult therapeutic problem as the surgeon must seek to achieve two objectives: first to minimize the risk of malignancy and second to obtain acceptable cosmetic results. The first objective calls for early and radical excision of all pigmented areas; this may be impossible for two reasons. First is the operative risk of surgery in infancy, the age group in which most of

the patients present and the second is the risk of leaving the patient with deformity or disfigurement. Cosmetic considerations may call for less aggressive surgery.
Most authors agree that if feasible, giant congenital nevus should be excised in toto[10,19], though some belive that the dangers of operative procedure outweigh the risk of development of malignant melanoma. If surgery is considered, then excision should be performed early, since malignant melanoma may arise even in the first 5 years of life[10,19]. The psychologically traumatic effect of disfigurement may be significant.

Excision should be carried to the deep fascia since the nevus may extend to this level[16]. However, in some unfortunate cases, the pigment cells extend deeper into bones, leptomeninges and other sites, making total excision impractical[20]. In cases where excision is possible, margins need to be excised carefully as recurrences have been reported even in benign cases[11]. Closure of the defect created by the removal of the nevus may be possible by primary closure. However skin grafting may be indicated in most cases with large defects, amongst these some may require a single skin graft while others may need to have multiple grafts, either in single operative procedure or multiple operative procedures, spaced at various intervals. In cases where the area of the skin invloved is so vast as to leave insufficient normal skin to cover the defect, the recommendation is to do selective excision, removing as much of the abnormal skin as possible and monitoring the untreated area.

Immediate skin expansion is an old concept. We used a modification of this technique in one of our cases. This technique is now being attempted intra-operatively using hypodermic needles to inject air and dental wire. With this technique the wound can be gradually approximated and subcutaneous sutures placed with the device in situ. An intradermal or simple running suture is then placed to approximate the skin edges under little or no tension and the device is removed. Upto date the technique has been tried on 15 patients with minimal morbidity and good results i.e. healing of wounds with primary intention[21]. Another new technique being tried for larger defects is repeated skin expansions with mean delay between expansions being 9 months. It has shows good long-term functional and cosmetic results[22].

Another mode of treatment being currently advocated is Dermabrasion. This involves removal of superficial layers of the nevus taking away the superficial melanocyte mass. This is followed by healing with non-pigmented or much less pigmented skin improving the appearance. However the bulk of melanocytes in the deeper layers remain unchanged, alongwith the risk of malignant melanoma[23,24]. Where a single technique is not likely to give good enough results, combination of the above mentioned techniques, i.e. removal of superficial layers shortly after birth, full thickness resection of the severely affected areas and reconstructive plastic surgery results in an acceptable cosmetic appearance and a significant decrease in the risk of malignancy. Completion of these surgical procedures is recommended before the age of 4-5 years.

Management and long term results of deep dermal nevus with malignamt melanoma is not clearly defined in literature because of its rare occurrence. Our first case, born with malignant melanoma, has been surgically managed mainly to provide palliation and to allow proper handling. We have not cantemplated other treatment modalities like radiotherapy and chemotherapy at this neonatal stage.

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TRICHOBEZOAR IN A CHILD (RAPUNZEL SYNDROME)

Case Report

SATTAR MEMON, K. ALTAF TALPUR, JAN MOHAMMAD MEMON

ABSTRACT:
A rare case of 112cm long Trichobezoar of stomach and small gut in a child of 9 years is presented (Rapunzel syndrome).

KEY WORDS: Trichobezoar, children

CASE REPORTS

A 9 year old girl of Hyderabad was admitted in the Surgical Ward with nausea and colicky upper abdominal pain, associated with a lump for 4 weeks. She had recurrent attacks of mild pain in the upper abdomen on and of for last two years but the parents ignored the symptoms. Patient had severe pain and anorexia for two weeks. There was no history of haematmesis or malena.

On examination, she was an anaemic child of average built with pitting oedema of feet. A huge intra-abdominal mass involving whole of the upper abdomen was palpable. It was quite mobile from side to side and from above downwards. Liver, spleen and kidneys were not palpable.

Her psychiatrie examination was normal. She had hemoglobin of 9.0gm%, ESR, 0.3mm/1st hour, blood urea 17mgm%, albumin 3gm%, globulin 3gm% and A/G ratio 1:1. Her urine D.R., serum electrolytes and LFT were within normal range. X-Ray chest was clear. Gastrograffin Meal showed a large filling defect in the stomach.

Under general anaesthesia, abdomen was opened through mid-line incision. A huge 112cm long Trichgobezoar occupying the stomach and upper intestine was removed through gastrotomy incision (Figure 1 & 2). Post-operative period was uneventful and the patient was discharged on 8th post-operative day.

DISCUSSION

Bezoars has been classified into three categories by Debakey and Oschner[1].

1. Trichobezoar or "Hair-ball" composed of masses of hair and decaying food materials. Usual sufferers are young girls who pick their hair and swallow it.
2. Phytobezoar consists of elongated masses of vegetable fibres, seeds and food.
3. Hard concretion which are organic masses of calculi containing calcium.
4. The fourth type can be added as large masses of Carradia Albicans especially in post-gastrectomy patient.

Figure 1 Trichobezoar being removed from stomach.

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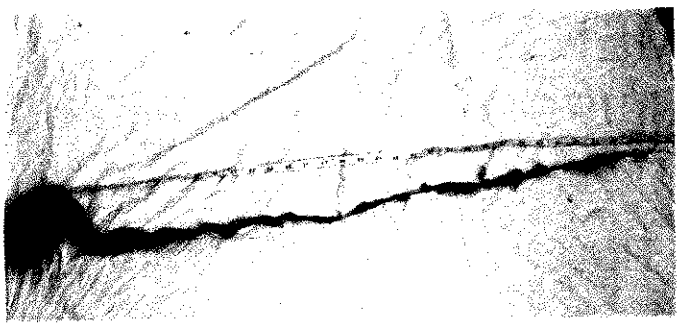

Figure 2 112 cm Trichobezoar after working

Trichobezoar are commonly seen in ladies with psychiatric problems. It is rare in children. This baby used to pick up lice from elder ladies and while picking lice she used to pluck the hair and then sallow the bunch of hair.

Various medications like hygroscopic bulky laxatives, non-absorbable antacid etc have been reported to produce iatrogenic bezoars leading to gut obstruction[2]. First post-vagotomy bezoar possibly due to delayed gastric emptying has been reported by Oliver[3].

Most of the bezoars in the stomach can be fragmented and destroyed with gastroscopy. As in our case huge and big bezoars however need operative intervention. Phytobezoars may be softened with enzymes and then removed with gastroscopy.

In our case the total length of trichobezoar was 112cm extending from stomach to jejunum. Repunzel is a rare form of gastric trichobezoar extended throughout the bowel (Uroz-Tristan J et al)[4]. They have reported a long trichobezoar extending from stomach to jejunum. Wolfson, PJ et al[5] reported fourth youngest patient of a trichobezoar extending from stomach to caecum suggesting that clinician must consider possible presence of a tail in any patient with a bezoar. Seker,B et al[6] also reported a long trichobezoar in a 6 years old girl extending from stomach to caecum (ninth reported case of Repuuzel syndrome). Rao,PL et al[7] reported 8 children with tricho bezoar suggesting that one should suspect trichobezoar in a child especially a girl who presents with a firm mobile lump in the epigastrium.

Bezoars are diagnosed with conventional radiology and gastroscopy permits identification of its nature (Santiago-Sanchez-CA et al)[8]. Currently management of bezoars consists of dissolution, suction, lavage, mechanical endoscopic fragmentation using pulsating jet of water, fragmentation with extracarporeal shock wave lithotripsy and surgical removal. Treatment should also focus on prevention of recurrence, since elimination of the mass will not alter the conditions contributing to their formation.

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XERODERMA PIGMENTOSUM WITH MALIGNANT MANIFESTATIONS

Two Case Reports

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QAISER MAQSOOD, M. NAEEM KHAN

ABSTRACT:
Xeroderma pigmentosum and its complications are not well understood. Most of the cases present with complications of xeroderma pigmentosum due to delay in diagnosis at an early age. We are presenting two cases of squamous cell carcinoma seen in early childhood secondary to xeroderma pigmentosum. The importance of early diagnosis of xeroderma pigmentosa and practice of proper preventive measures so as to delay the onset of complications is stressed.

KEY WORDS: Xeroderma Pigmentosum; Squamous cell carcinoma.

INTRODUCTION

Xeroderma pigmentosum is a rare genodermatoses. It was described for the first time in 1874 by Hebra and Kaposi[1]. It is an autosomal recessive trait found in all the races worldwide[2]. It is a generally heterogenous group of degenerative disorders in which abnormal DNA repair induced by ultraviolet light[3] manifests as sun sensitivity and cutaneous sun damage, oculocutaneous pigmentary abnormalities, neoplasia, mainly skin malignancies, and at times neurological impairment[4,5]. Despite its rarity, xeroderma pigmentosum has been extensively studied, the main reason being lack of any significant treatment modalities which can improve the standard of living and life expectancy of patients and prevent many life threatening complications of the disease.

Squamous cell carcinoma is a very common complication of xeroderma pigmentosum[2] with very limited treatment options available. We are presenting here two cases of squamous cell carcinoma seen in patients with xeroderma pigmentosum. We are aiming to highlight the lack of proper preventive measures for patients with xeroderma pigmentosum, which if properly followed can delay the onset of complications. The treatment of squamous cell carcinoma is also discussed.

CASE REPORTS

Case #1

A six years old boy presented with multiple scattered pigmented freckle like lesions all over the body, specially the exposed parts and face with predominant involvement. He had polypoidal lesion under the left eye which was diagnosed as squamous cell carcinoma and treated elsewhere with radiotherapy. Besides the lesions already described he also had severe conjunctivitis corneal ulceration and opacities and several friable polypoidal neoplastic growths on the under surface of the eye lid inner canthus, nostril and cheek (Fig. 1). He was diagnosed to be a typical case of xeroderma pigmentosum with its known complications of multifocal lesions of the face. Despite the classical presentation, the patient had not had a definite diagnosis and had never been advised any protective measures from ultraviolet light. He was offered chemotherapy but the parents declined.

Case # 2

An eight year old girl presented with generalized pigmentation and ulcerating lesion below the eye with inrolled edges and central necrosis (Fig. 2). This was diagnosed to be a squamous cell carcinoma arising as a complication of xeroderma pigmentosum.

Wide local excision was performed and radiotherapy was given. The patient was advised various protective measures to minimise the damage caused by ultra-violet light. She was seen regularly for a year and was then lost to follow up.

DISCUSSION

Incidence of Xeroderma Pigmentosum is reported to be between 1 in 250,000 and 1 in million[6]. The incidence also varies in different parts of the world, being much higher

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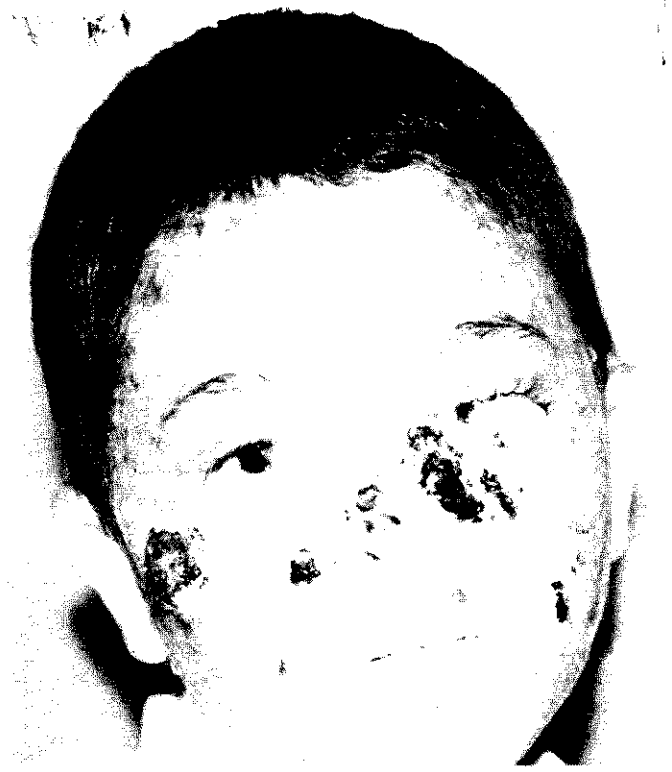

Figure 1 Six year old boy with multiple hyperpigmented frackle-like lesionss all over the body specially the face with several satelite polypoidal lesions on the under surface of the eye, inner canthus, nostril and cheek which on histopathology proved to be multifocal squamous cell carcinoma sceondry to xeroderma pigmentosum.

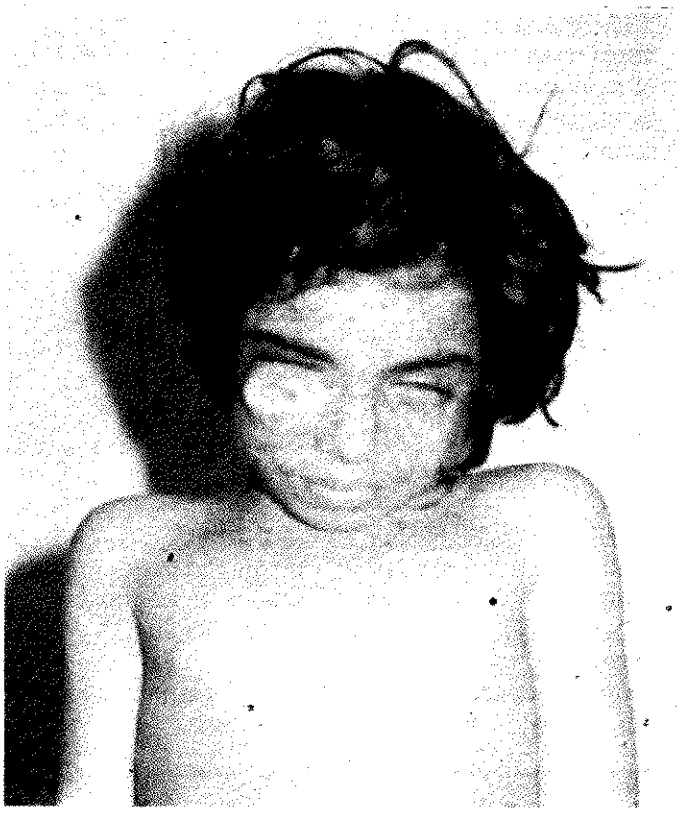

Figure 1 Eight year old girl with generalized pigmentation and an ulcerating lesion under the eye with central necrosis and inrolled margins. Histopathology showed this to be squamous cell cacinoma.

in Japan (1 in 40,000) than in America and Europe (1 in 250,000)[3]. High incidence of first degree consanguinity of the parents of these children is seen and is reported to be anywhere between 12 and 100%[4,7]. Sexes are reported to be effected equally[4,6,7]. The basic pathology is the inability of the cells to repair the damaged DNA caused by exposure to ultraviolet sunlight[3]. Three hundred potential mutation sites have been described as the causal mutations in Xeroderma Pigmentosum[8].

The disease has a bonozgous form, a heterozygous form and a compound heterozygous form. Homozygotes are severely affected and the hetrozygotes are clinically unaffected[2]. The genetic heterogeneity of the syndrome, suspected on clinical grounds, has been confirmed on experimental studies. Approximately 80% of the patients show a defect in the initiation of the DNA excision repair acting on pyrimidine dimers. The other 20% called Xeroderma pigmentosum variants show a defect in S-phase of DNA replication after exposure to ultra-violet radiation.

Presentation is early, usually within the first few months of life with photophobia and chronic conjunctivitis. Soon after, sunburn-like reactions with prolonged erythema and edema and later scaling on the sun-exposed areas occurs. The resolving erythema is then replaced with macules that look like freckles, which are seen alongwith hypopigmentation before two years of age. The number of freckles increases with time as does the variability in their colour. However, the tendency of acute sun sensitivity subsides with age[2]. The skin is dry, scaly and wrinkled and may be so tight that ectropion results. The facial skin becomes atrophic with permanent telangiectasias, blisters, crusts, actinic keratoses, and scar ensue that may cause marked distortion of the nose, eyes, and the mouth. Ophthalmic findings are seen in about 80% of the patients[2]. Pathology is virtually confined to the anterior portion of the globe, although choroidal melanomas have been seen. The lids are also effected. Basal cell and squamous cell cancers commonly occur on the lid margins. Within the first decade of life, basal cell epithelioma, squamous cell carcinoma, and malignant melanomas appear (in decreasing order of frequency) causing considerable mutilation[2]. Less common tumors include keratoacanthomas, sarcomas, angiosarcomas, fibrosarcoma, fibromas, and angiomas[2]. Internal tumors such as medulloblastomas and neuromas occasionally develop[4]. Skin cancer tends to occur earlier in the homozygous and a little later in the heterozygotes. 20-60% of patients have severe nervous system involvement[4]. Various biochemical and metabolic abnormalities may coexist with xeroderma pigmentosa like renal impairment, aminoaciduria, and adrenal hypofunction[2].

The disease is often fatal before the age of ten and two thirds die before the age of twenty, usually due to spread of malignant lesions. But many patients die of infections, to which they are abnormally susceptible. Survival beyond middle age is sometimes possible in mild cases with adequate treatment. Striking inter family variation in expectancy of life has been reported.

A patient diagnosed to have xeroderma pigmentosum needs to be firmly advised concerning measures required to minimize the damage inflicted onto the skin cells by ultraviolet light. Rigorous protection against UV light should be started from infancy; sunscreens should be used regularly before exposure to sunlight, glasses opaque to UV light should be used[9,10].

Regular and frequent follow-ups of these patients are necessary for detection of premalignant and malignant conditions. Premalignant conditions should be treated with either cryosurgery or topical antimitotic agents. Patients presenting with squamous cell carcinoma have limited treatment options. If excision is possible then local excision with wide margin is recommended.

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CARCINOMA OF PARATHYROID A RARE CAUSE OF HYPERPARATHYROID BONE DISEASE

Case Report

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ABSTRACT:
Carcinoma parathyroid is a rare cause of primary hyperparathyroidism. The biological behaviour of this tumour is not well established as only a few cases have been recorded. We are reporting a case of a 45 years old female who presented with musculoskeletal features of primary hyperparathyroidism. The clinical diagnosis of Hyperparathyroidism was supported by results of laboratory and Scintigraphy investigations. Surgery was performed and the biopsy report showed Carcinoma Parathyroid. The patient's symptoms were relieved after Surgery.

KEY WORDS: Primary Hyperparathyroidism, Parathyroid Carcinoma, Osteitis Fibrosa Cystica Generalisa

INTRODUCTION

Primary hyperparathyroidism (H.P.T.) is not as uncommon as thought. Due to availability of facilities for harmonal assays, more and more cases of primary H.P.T. are being diagnosed now. According to recent reports it forms upto 30%-50% of the total metabolic bone disorders[1,2]. We are reporting a case of carcinoma parathyroid which is a rare case of hyperparathyroid bone disease; it constitutes 0.01%-0.4% of primary H.T.P. cases[1,2]. It is the first case report of Carcinoma parathyroid causing metabolic bone disease in Pakistan.

CASE REPORT

In July 1995, a 45 years old female presented with complaints of generalized bone aches, weight loss and progressive muscular weakness for the past six years and was being treated by family physicians. For the last two years, intensity of her symptoms had increased when she had trochanteric fracture of right femur following a trivial trauma (fall), which was treated by bone setter (potter). After six months she was able to walk with the help of a cane. Two months back (April 1995) she had another fall while getting out of bed and developed severe pain in right leg and hip. She also complained of severe low backache, polyuria, polydypsia and increasing muscle weakness. She had no symptoms relating to urinary tract disease or diabetes.

Apart from malunited subtrochanteric fracture and mild tender swelling on right tibial shin, she had no other remarkable findings on clinical examination. She had no swelling in neck.

Her blood picture findings (Hb, TLC, DLC), urine analysis, blood sugar, urea, creatinine and electrolyte were within normal range. ESR (50mm 1st Hr),. Serum Calcium (12.97 mg/dL - N=8.6-10.5), Phosphorous (4.1 mg/dL - N=2.7-4.8), Alkaline Phosphatase (1899 U.I - N=65-306), 24 hour urinary excretion of Calcium (280 mg/ml) and S.Parathramone (1.29 n gm - N=0-0.27) were raised, whereas serum proteins were low (Total 6.8, Alb 3.8) and Urine Bence Jones protein were not found in urine. D.T.P.A. renal scan shows normal functioning kidneys.

The results of these investigations, radiographic skeletal survey and scintigraphy reports (Table-I) supported the diagnosis of metabolic bone disorder, most probably due to primary hyperparathyroidism. The histopathological report of tibial osteolytic lesion suggested Brown tumor. Dual Isotope parathyroid Scintiscaning suggested functioning parathyroid tissue over the left lobe of thyroid (Table-I).

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TABLE I

SKELETAL SCINTIGRAPHY TC 99 m MDP	RADIOGRAPHIC SKELETAL SURVEY
Scan shows involvement of long bones, sparing the axial skeleton which is not characterstic of Metastatic disease. Cold lesion over the neck and trochanteric region of right femur. Possibilities: 1. Primary Bone Tumor 2. Metabolic Bone disorder **Dual Isotope Parathyroid Scintiscaning** (TC 99 m - T1 201 Substraction) Dual Isotope Scintiscan is positive for functioning Parathyroid Tissue over lower part of left lobe of thyroid. Chest does not show any ectopic activity.	**Pelvis and Hips** • Healed subtrochanteric fracture right femur. • Radio lucent cystic changes in both trochanterse, more significant on right side. • Cystic expansion of ischium & Pubii. • Cystic lesion along the iliac crest, supra acetebular region and iliac part of SI joints. **Long Bones** • Generalized punctate osteoporosis and cortical thinning of femur, tibia, fibula, humerus and metacarpals. • Eccenteric cortical expansion with cystic lesions upper and lower right tibia and lower fibula.

Surgical exploration revealed two nodules, one in the left lower lobe of thyroid and another behind lower part of left sternomastoid muscle, whereas the other three parathyroid glands were normal at the usual locations. Regional lymphnodes were normal. Histopathological findings of sternomastoid nodule were consistent with carcinoma parathyroid The size of tumour tissue was 5x4x1 cm. and weight 5 gm. grey white on cutting. and thyroid nodule showed no remarkable changes. The tumour was composed of atypical cells arranged in sheets separated by dilated vascular channels. The cells were round to oval in shape with hyperchromatic to vesicular nuclei and eosinophilic cytoplasm. They are separated by dense fibrous tissue. Increased mitotic activity was also evident. The surrounding fibrofatty tissue revealed foci of these same atypical cells.

Recovery was uneventful except that her calcium dropped to 8.6 mg% within 24 hours after surgery and then remained within normal limits. Serum PTH level was also normal in postoperative period. She was given injection vit.D.3 (3 lac weekly), and calcium supplement for 03 months. She was relieved of her musculo-skeletal symptoms in two weeks and she started walking with the help of a cane in six weeks.

She fell again 2 months after surgery and sustained fracture of the surgical neck left hemerus, left clavicle and undisplaced type I Inter-trochanteric fracture. These fractures healed well in time with non-operative management She was doing well till last followup four months back.

DISCUSSION

Primary hyper parathyroidism and malignancy accounts for 90% of hyper calcemia, whereas the primary hyper parathyroidism constitutes 30%-50% of cases of metabolic bone disorders[1-3]. Primary hyper parathyroidism commonly affects middle aged persons but no age is spared, whereas Carcinoma Parathyroid more commonly affects in 5th and 6th decade of life. Females are more frequently affected than male[2-5]. Carcinoma parathyroid affects bones much more as compared to kidneys. The skeletal changes are more aggressive in carcinoma as compared to adenoma. The common manifestations are osteitis fibrosa cystica and pathological fractures[2,7], as has been observed in this case. Renal stone and nephro- calcinosis are rarely seen despite significant excretion of calcium in the urine. Other common presentations are fatiguibility, hypertension and psychotic symptoms, whereas metatastic calcification, pancreatitis and peptic ulcerations are rarely seen[2,6-8].

The diagnosis of carcinoma parathyroid is extremely difficult unless gross local and distant metastasis have occurred[2]. Clinical and laboratory parameters do help in diagnosis of hyper parathyroidism, but carcinoma parathyroid can only be diagnosed on histopathology. Parathyroid harmonal assay may give a clue if exceptionally raised. Parathyroid scintiscanning is a non-invasive method for localization of normal and ectopic tissues and is of definite help in case of recurrence after surgery. Dual Isotope Technicuim 99m - Thalium 210 or Technicuim 99m MiBi subtraction scannings are superior than previously used Selemethomium and Cesium chloride substraction scanning[9]. High resolution, real time, ultrasonography with 10 MHz probe, transoesophageal endosonography and Cine radiography techniques are truly helpful in localization of parathyroid glands[1,2,9]. C.T. Scan and MRI are of immence value in persistant and recurrent cases especially when the tumour is located in the mediastinum[2,7,8].

In Carcinoma parathyroid en bloc resection of tumour and surrounding involved tissue is recommended to reduce the recurrence rate[2,4,7,8]. Histologically, the tumour cells are arranged in trabeculae and in solid pattern with fibrous band, few cells show mitosis. Invasion of capsule and blood vessels is a frequent feature. Presence of parathyroid harmone in cytoplasm detected by immunohistochemically is diagnostic[6].

Prognosis is usually not good. Patient usually die of hypercalcaemia. Survival depends upon maintenance of serum calcium level below 12 mg/dL[2,3,5]. In addition to parathyroidectomy various medications are used to maintain serum calcium level within normal limits; in inoperable cases that include Mithramycin, Stilbesterol, Calcitonin

and Biphosphonates[2,3,10]. Medical management requires prolonged treatment and continuous monitoring.

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EOSINOPHILIC GRANULOMA OF TIBIA

A Case Report

M. AKHTAR RAJA, ANISUDDIN BHATTI, GHULAM MEHBOOB, SAGHIR AHMED

ABSTRACT:
Eosinophilic Granuloma of long bone diaphysis is a rare, tumour like disorder. It presents as osteolytic lesion with significant periosteal reaction. Often misdiagnosed as Garre's clerosing osteomyelitis or Ewings sarcoma. We report one of such case in a 16 year old boy's tibial diaphysis. The curettage biopsy confirmed the diagnosis and relieved the patient's problem. Recurrence has not reported till year long folllowup.

KEY WORDS: Osteolytic lesion, Periosteal reaction Eosinophilic granuloma.

INTRODUCTION

Skeletal Eosinophilic granuloma is a rare, benign, tumour like disorder of bones. It commonly affects children aged between 01-15 years and rarely affects adults in old age[1]. No Bone is immune to this disease. however, appendicular skeleton is rarely affected. In long bones, it affects shaft, equally distributed in diaphysis and metaphysis; whereas epiphyseal location is exceptional[2]. Every patient is a unique entity and it is very difficult to commit the diagnosis on clinico-radiological grounds. the histopathological feature of biopsy is the only definite way for diagnosis. After thorough curettage of solitary lesion, chances of recurrence are truly nil, multiple lesion often needs curettage or/and corticosteroid and/or low dose radiotherapy. We report this case which is the first to be reported in Pakistan.

CASE REPORT

In September 1996, a 16 year old boy presented with 3 month old swelling of mid leg shin region which was progressively increasing and producing dull ache. There was no history of trauma, fever, sepsis or weight loss. The swelling was diffuse involving diaphysis of tibia. Skin overlying was mobile with normal texture and temperature, swelling was bony hard and tender. Blood investigation (Hb, Tlc, Dlc, Urea, Sugar) urine analysis, Xray chest were within normal limit. Radiograph of leg showed radiolucent lesion involving tibia diaphysis with fusiform expansion, thickening of middle third of tibia and significant periosteal reaction (Fig 1). Tc99 bone scan showed increased tracer uptake in middle of right tibia and remarked as osteolytic lesion/bone tumour.

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With provisional diagnosis of chronic osteomyelitis or Ewings sarcoma, thorough currettage was carried out. The curettage material was greyish white, semi-solid in nature, the surrounding wall of lytic area was sclerotic. The hisotpathology report findings were consistent with Eosinophilic granuloma.

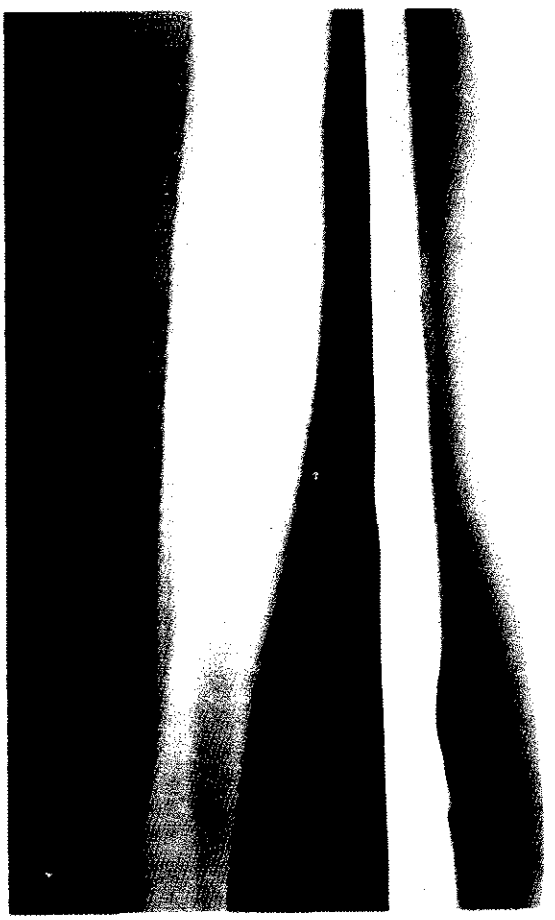

Figure 1 Eosinophilic granuloma of tibia

The patient's wound healed in due time without complications. His symptom of dull ache was relieved. However fusiform swelling of leg persisted with signs of callus formation/healing in previous site of lesion.
On one year followup, he has not developed any signs of recurrence of disease.

DISCUSSION

Solitary of Multifocal Eosiophilic granuloma of bone is a rare benign, tumour like disorder. Histogenetically it is considered as a variant of Histocytosis[1], characterized by Langhan cell histocytosis[3], causing osteolytic lesion of bones. Eosinophilic granuloma is not a new disease, it was first described in 1913 and 1914 by the Russian Pathologist Tartynov, who mistook it for Pseudotuberculous granuloma with a local eosionphilia and Charcot leyden crystals. In 1940 Liechtenstein, Jaffe, otani and Ehrlich recognized the entity as a distinctive, solitary, destructive bone lesion of inflammatory nature and coined it the name "Eosinophilic Granuloma", the term by which it became universally known[2]. Today many relevant literature are available which are either case reports or an intermittent attempt to clarify the disease process and to present new nomenclature or classification. In one of the series, incidence of Eosinophilic granuloma has been reported as 1:200000 per year[4].

Eosinophilic granuloma is a disease of childhood and adolescence (1-15 years) although few cases over age 30-60 has also been reported[1,2,5-7]. A moderately increased male to female ratio, that may reach, 2:1 has been noted among the multifocal cases[2].

Clinically it presents as painful mass affecting skull, spine, rib cage, pelvic bones and proximal skeleton. Bones of hand and feet are affected only rarely[1,2,8-10]. Often it occurs solitary but sometimes as multifocal lesions. In multifocal eosinophilic granuloma extra skeletal manifestations may be featured without obvious skeletal involvement[2].

Typical radiological features of early lesions are those of destructive radiolucent oval area of rarefecation with varying sizes in spongiosa, these are usually well demarcated "punched out", without peripheral sclerosis. On occasions, the lesion may be poorly demarcated and the periosteal gradually merges with adjacent normal bone, and on occasions periosteal bone formation, especially in long bones may occur, giving a typical onion skin lamellated appearance[1,2,7]. This feature closely imitates the periosteal changes seen in osteomyelitis, Ewings sarcoma or non hodgkins lymphoma. Similar was our initial diagnosis.

Hisotpathology of the lesion is the only way of confirmation, characteristic feature of Eosinophilic granuloma is the "presence of mature Eosinophilic leucocytes", although the basic proliferating cell is traditionally considered to be a histocyte[4]. In areas where eosinophilic leucocytes are undergoing fragmentation, proteinaceous crystalline structures, Charcot leyden crystals (Taratynov features) are demonstrable.

It is imperative to confirm the definite diagnosis for the treatment of patient, because the treatment of Eosinophilic granuloma is simple and does not need prolonged medicines, expansive repeated surgical interventions and anticytotic therapies. However, prolonged followup is often required as chances of recurrence and transformation to multifocal eosinophilic granuloma especially in craniofascial bones and ilium are more likely to occur[1,2]. In solitary lesions treatment beyond currettage biopsy is probably unnecessary, as currettage with or without bone grafting, low dose radiotherapy and corticosteroid injections has been found successful[1]. The multifocal (disseminated) form has been treated with radiotherapy, steroid, chemotherapeutic agents (vinblastine) and thymic extracts, all with varying successes[1,2,7]. The case presented above behaved well with curettage only.

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