Humeroradioulnar Synostosis in an African Child: A Case Report.

A. Agaba-Idu Musa
Dept. Of surgery, Usmanu Danfodiyo University, Teaching hospital, Sokoto. Nigeria
E-Mail: agabaidu@ymail.com

This is a report of humeroradioulnar synostosis at left elbow, in a girl from Sokoto, Nigeria in whom there is no history of hereditary/congenital diseases. This case is the thirtieth reported in literature. This defect was not solitary. Plain radiography revealed other deformities: In the right upper limb, the humerus, radius and ulna were well developed. There were three metacarpals and three fingers. In the left upper limb, there was humeroradioulnar synostosis and partial radial hemimelia. There were two metacarpals and three fingers. Treatment was offered but the child was not brought back to the hospital. This is the first case of humeroradioulnar synostosis reported from Africa.

Keywords: Humeroradioulnar synostosis.

Introduction

A girl presented one week after delivery with a stiff left elbow and three digits on both hands. These were the only defects seen. Such a congenital deformity at the elbow had been reported before from this hospital[1]. Plain radiography revealed all the defects. The defects were failures of differentiation and formation on the basis of A.B Swanson's classification[2]. Below are the case report and description of the treatment option.

Case History

SI is a girl who was delivered at term by a 24-year old mother, gravida 4, para 4, (two boys and two girls). Both parents were native Nigerians. There was no history of hereditary/ genetic diseases in both parents' families. The mother attended antenatal care. During pregnancy, apart from iron supplements, she did not take any other drugs or herbal preparation. Pre and post natal periods were uneventful. Delivery was normal, vaginal. The baby cried immediately after delivery and weighed 2.90 kg. The baby was admitted into the special baby care Unit of Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria because of the obvious congenital upper limb anomalies. She was reviewed by an orthopedic surgeon a week after delivery.

The girl looked healthy. She was neither pale nor had jaundiced. Her anterior fontanelle was not bulging. No pathology was seen on the face. Her lungs were clear. Her cardiac contractions were 155 beats / min. No murmurs were heard. The abdomen was not distended and no abdominal wall defect was seen. No enlarged organs were palpated. Both upper limbs had three fingers. The right upper limb was longer than the left upper limb. There was no sensory deficit in upper limbs. The bulk and tone of the muscles in both upper limbs were not the same. The difference was in the biceps branchi muscles. The left biceps branchi muscle was hypoplastic. There were passive and active movements at the right elbow. There were neither movements nor angular deformity at the left elbow. Pronation and suppination were possible only in the right forearm. Both hands had only three digits and no thumbs.

Plain radiography was done and the following deformities were revealed (figure1): In the right upper limb, the humerus, radius and ulna were well developed. There were three metacarpals and three fingers. In the left upper limb, there was humeroradioulnar synostosis[1] and partial radial hemimelia. There were two metacarpals and three fingers. The electrophoretic pattern of the parents' hemoglobin was AA. Both parents were not reactive to VDRL (venereal diseases research laboratory) test.

The child's condition was explained to the parents and they requested for a detailed explanation of soft release and excision arthroplasty[1], which was the treatment offered. Extension and flexion at the elbow are usually accomplished by the biceps and triceps branchi muscles respectively. Force of

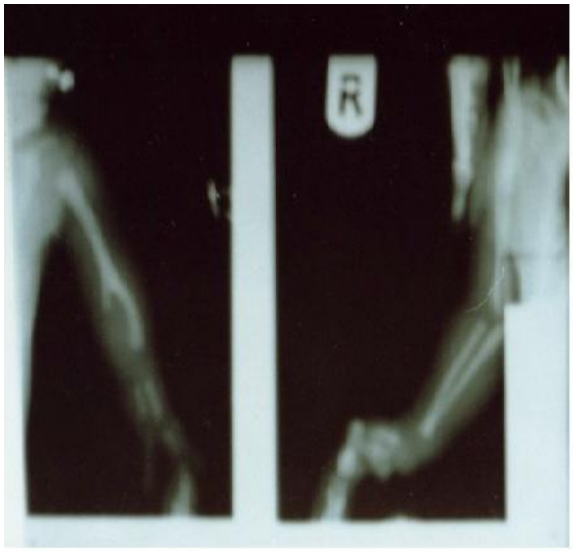

Figure 1

gravity alone can also accomplish extension at the elbow if there is agenesis or any defect of the triceps branchi muscle. They were impressed by the treatment option and requested for a three-month appointment but the child was not returned for surgery.

Discussion

The subject of this report is the thirtieth[3] and the first case of humeroradioulnar synostosis being reported from Africa. The parents of the child were not blood relation. In the right upper limb the congenital limb defects were only failures of formation[2]. In the left upper limb, there were failures of differentiation[2] (humeroradioulnar synostosis[3,4]) and formation (partial radial hemimelia, two metacarpals and three fingers). In the literature reviewed soft tissue release with excision arthroplasty at the elbow is the recommended treatment. Although initial range of movement is always satisfactory, re-ossification occurs[3]. This upper limb congenital defect if associated with lower limb, pelvic and genital malformations is a clinical variant of Al-Awadi/Raas Rothschild syndrome[4].

Conclusion

Although this patient was not operated it is being reported because of the rarity of humeroradioulnar, especially in native African children. This report will sensitise other clinicians to report more cases especially from Africa and shed more light on the mode of inheritance. The result of these new studies will help in prevention or at least reduction in the incidence.

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Patterns of Cardiothoracic and vascular surgical admissions at a tertiary University hospital Addis Ababa, Ethiopia

E. Teffera[1] , S. Kassa[2] , A Ali[3]
[1]Cardiothoracic Surgery Fellow, Addis Ababa University School of Medicine TAH, Department of surgery
[2] Thoracic Surgeon, Addis Ababa University School of Medicine TAH Department of Surgery
[3] Professor of Surgery, Thoracic Surgeon, Addis Ababa University School of Medicine TAH Department of Surgery

Background: ***The subspecialty training in cardiothoracic surgery at the school of medicine in the department of surgery Addis Ababa University is a new field in the country. Virtually all the thoracic surgical patients are referred to our hospital. There are two referral clinic days in a week where varieties of Cardiothoracic and vascular surgical cases are seen as an outpatient and those who deserve and are considered fit for surgery are admitted. The purpose of this study is to observe the disease patterns, evaluate the institutional capacity, implication on training and service delivery in cardiothoracic and vascular surgery***
Method: ***All patients seen at the surgical referral clinic and decided to be admitted for the first time or readmission, from July 2010 to June 2011 were included in the study. Emergency cases were excluded. Data were analyzed using statistical tool SPSS ver.16***
Results: ***A total of 274 cases were decided for admission in this time period. Females were 145 accounting for 52.9 % while men were 129 accounting for 47.1%. Mean age was 38 years while the median age was 39 years, ranging between 13 and 88 years. Neoplasm is the leading pathology for admission accounting 169 (61.7%) cases out of the 274. Malignancies are observed to contribute 126 (45.9%). 98 Esophageal cancer cases were recomeneded for admission making it the single most common diagnosis on admission. Among the esophageal cancers 56 were females (57.1%) while males were 42 (42.9%).***
Conclusion and Recommendation: ***There was an observed change in the male to female ratio with females taking the majority of the admissions and the majority of the esophageal cancer cases admitted. Neoplasm in general and more importantly malignant neoplasm were the leading causes of admission unlike the previous studies indicative of preponderance of non neoplastic and non Malignant causes. The number of cardiothoracic surgery admissions has increased dramatically. We recommend on the commencement of Cancer registry in the country. Further study is needed to have an impact in resource allocation at the institution.***

Key words: Patterns, Cardiothoracic, Vascular, Surgery, Admissions

Introduction

Sub specializations in medicine have evolved for long in developed countries. This has resulted in an improved healthcare delivery, more efficient patients care and better prognosis of surgical pathology that once had fatal outcome. Addis Ababa University School of Medicine Tikur Anbassa Hospital (TAH) is the highest medical training, research and service center in the country. The hospital accepts patients from all over the country as it is the main referral tertiary hospital. However the subspecialty training in cardiothoracic surgery at the school of medicine in the department of surgery (AA), is a new field and had just taken in its second batch of candidates for the fellowship program. Virtually all the thoracic surgical patients are referred to the hospital. There are two referral clinic days in a week where cases are seen as an outpatient by surgical team constituting Consultant cardiothoracic and vascular surgeons, Fellows in the subspecialty training and General surgery residents attached to the unit of cardiothoracic and vascular surgery. Those who deserve and are considered fit for surgeries on provisional basis are admitted.

TAH has a total of 250 surgical bed capacity amongst which 2 wards are designated for elective adult cardiothoracic and vascular surgery patients. While a 3rd class is equally divided into 7 beds each

between females and males, Fifteen 1st and 2nd class beds are randomly given to either males or females with sum total of 29 beds. While generally it is understandable that analysis of hospital admissions is not a reliable method to study the disease patterns of a given community it may, however, despite its limitations, can give indicator information about the common diseases that require hospital admission. It can also provide background data for further comparison with similar studies elsewhere. There are few studies so far and data are scant on the patterns of admission be it to our hospital or in the Sub Saharan Africa at large.

The general Objective of this study was to evaluate disease patterns in cardiothoracic and vascular surgery practice at a tertiary teaching hospital in Ethiopia. The specific Objectives were to determine the epidemiology and disease patterns in cardiothoracic and vascular surgery and to evaluate the institutional experience and the implication on, training and service

Patients and Method

All patients seen at the cardiothoracic surgical referral clinic (i.e. elective cases) and decided to be admitted whether for the first time or readmission were included in the data collection. Prospective Data were collected in a designed format from July 2010 to June 2011 inclusive. Data were collected in terms of the following parameters: diagnosis, age, sex and pathology. The Adult age group is defined by those > 12 years. Age was further categorized in groups each of 10 years. Pathology was classified as neoplasm, infection, inflammation, congenital, degeneration, iatrogenic etc based on standard Pathology and surgical text books[1,2]. Neoplasm's were further divided in to benign, malignant, and unspecified based on clinical assessment, imaging or biopsy result.

Data collected were entered into a computer and analyzed using statistical tool SPSS software version 16. Data were summarized in form of frequency tables for categorical variables.
Means, median, mode, standard deviation and histograms were used to summarize continuous variables

Results

A total of 274 cases were decided to be admitted in this time period. Females were 145 accounting for 52.9 % while men were 129 accounting for 47.1%. (M:F sex ratio = 1:1.12) The aged ranged between 13 and 88 years with a mean age of 38 years (standard deviation of $\pm$1.4244) and median of 39 years,.

Neoplastic conditions were the leading surgical pathology for admission making 169 (61.7%) cases out of the 274. Malignancies are observed to contribute the majority of the neoplasm and all diagnosed cases as well by 123 (44.9%).Ninety eight Esophageal Cancer (35.7%) cases have been included to the admission making it the single most common diagnosis. Among the esophageal cancers 56 were females (57.1%), 42 (42.9%) were males. Among the patients diagnosed with esophageal cancer Sixty eight (70 %) of them were 39 years of age or above. Proximal Gastric and Gastro-esophageal junction cancer cases accounted for 4(1.45%) and 3(1.1%) respectively. One patient (0.4%) was admitted with the diagnosis of oesophageal polyp.

There were 13 cases of lung tumour (4.74 %) out of which 7 were cases of lung cancer with an additional 2 (0.7%) cases of lung-masses and 2 (0.7%) patients with lung teratoma with an additional two patients with endobronchial tumours (0.7%) (one carcinoid tumour) of as well were also decided for admission in this time period. Pleural-based mass accounted for 2 (0.7%) cases. A total of 17 (6.2 %) of the cases had mediastinal masses.

Nine (3.3 %) of cases presented with chest wall tumours including 6 (2.2%) cases of chest wall sarcoma, 2 (0.7%) patients with unspecified chest wall masses and one patient with Sternal Plasmacytoma (0.4%). Other rare lesions included one (0.4%) case each of neck mass, gluteal vascular mass and laryngeal cancer. Fourteen (5.1%) of carotid paraganglionomas were the leading

cause of vascular neoplasms followed by 2 (0.73%) cases of haemangiomas. There were 3 (1.1 %) cases patients with malignant pleural effusion.

Infection was the second major pathology patients presented with in 42 cases. Hydatid cyst of lung was diagnosed in 16 (5.84 %), empyema thoracis in 14 (5.10 %), Aspergilloma in 5(1.8 %), Constrictive pericarditis in 5 (1.8 %) Post TB destroyed lung in 1(0.4 %) and Post-TB-fibrosis 2(0.73 %) of the patients admitted. Degenerative diseases accounted for 33(12.04%) of the cases. There were 14 (5.1%) of achalasia cardia, 2 (0.7%) of vascular degenerative changes, 2(0.73%) had aneurysm and 15 cases (5.47%) of varicose veins were seen. There was one patient (0.4%) with lung bulla and one with para-oesophageal hernia. There were 11 cases (4.01%) of congenital conditions with patent ductus arteriosus (PDA) being the commonest (2.55%). Others included bronchogenic cyst (0.4%), pigeon chest (0.4%), cervical rib (0.4%) and vascular malformation (0.4%) account the remaining.

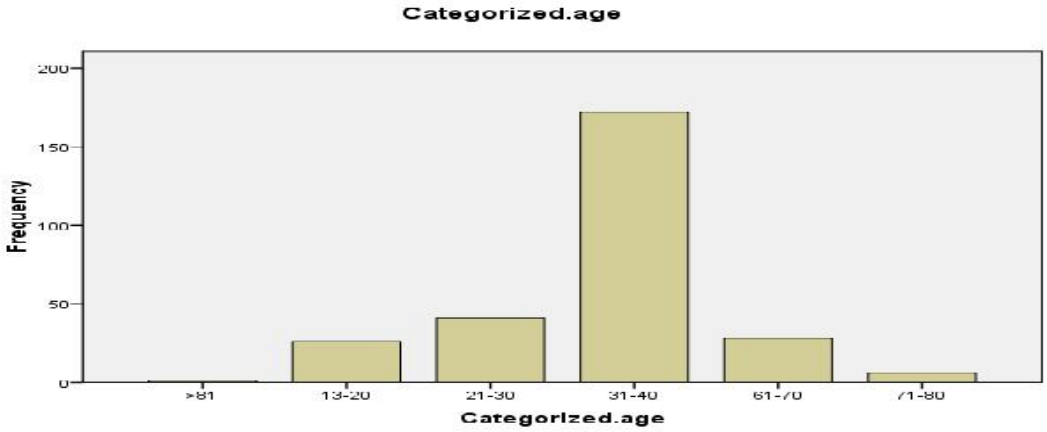

Figure 1. Categorized Age Group

Table1. Disease Distributions Requiring Admission into Cardiothoracic and Vascular Surgery

Pathology	**Frequency (n-274)**	**Percentage (100%)**
Neoplasm-malignant	123	44.9
Infection	44	16.1
Degenerative	33	12.0
Tumor otherwise unspecified	25	9.1
Neoplasm benign or low grade malignant	21	7.7
Congenital	11	4.0
Inflammation	8	2.9
Others	8	2.9
Autoimmune	1	0.4
Foreign body	1	0.4
Iatrogenic	1	0.4

Table 2. The Ten Top Diseases of Cardiothoracic and Vascular Surgical Admissions

Diagnosis	Frequency	Percentage
Esophageal Cancer	98	35.7
Mediastinal mass	17	6.2
Hydatid cyst of lung	16	5.8
Varicose Veins	15	5.5
Empyema thoracis	14	5.1
Carotid paraganglionoma	14	5.1
Achalasia cardia	14	5.1
Lung-cancer	7	2.5
PDA	7	2.5
Chest wall Sarcoma	6	2.2
Total	**208**	**75.8**

There were 2 (0.73%) cases with post-oesophagectomy anastomotic stricture and one patient with tracheal stenosis (0.4%). Five (1.82%) of the patients presented with benign esophageal strictures accounting the remainder of the inflammatory pathology. One patient (0.4%) had presented with signs and symptoms of myasthenia gravis. A single patient was foreign body in the shoulder was also decided to be admitted while one patient was admitted with post oesophagectomy leak. There were 8 cases which were difficult to categorize pathologically (cavitary lung lesion, vascular lesion, Esophageal varices and Arteriovenous fistula) accounting for 2.9 % of the 274 cases.

Discussion

The present study showed a slight female preponderance (F: M 1:1.12) with a reversal from the previous study[3] result M:F 1.4 : 1 for elective cases conducted in the same hospital for general surgical admissions from September1994 to August 1997.There were3968 admission in that study with 2314 male patients and 1653 female patients. The Chi square test corrected for two categories (Yates) gave ^{X2} = 4.88 with a *p value* < *0.05* which is statistically significant.

The missing age group of 41 – 60 years is attributable to the fact that most of the patients may not know their real age as it was evidenced by the fact that only 6 % of the population has a birth registry[4]. Another factor for the missed age group may be that some patients registration at the hospital medical records office are done by relatives who are unaware of the patients actual age .The unreliability of the age data makes it impossible to draw any conclusion regarding the age distribution.

The numbers of neoplasm cases being seen are increasing in proportion to the other non neoplastic pathologies. Comparing with a prior study in the same institution which reviewed the 5 years referral pattern of surgical patients abroad from September 2005 –August 2009[5] by applying the Chi square test and similar adjustment yielded. X^2= 13. 56 and *p value* < *0.001*. The number of malignant pathologies are also seem on the raise as it was evidenced with a significant change from the same study 1994 – 1997.Which has found that malignant cases accounted for 21.5 5 of the 5353 cases of surgical admission. Chi -square test result was X^2= 31.33 and *p value* < *0.001* which is highly significant departure from the null hypothesis that we have very strong evidence to conclude the fact that there is a shift in the paradigm of the disease patterns in the country.

The same study also found that esophageal cancer cases accounted for only 5.4% of the total surgical admission. However, in our study the number of esophageal cancer cases constituted the majority of the overall Cardiothoracic and Vascular Surgical admissions. There is a significant increment in the number of esophageal cancer cases with $X^2 = 28.08$ and $p\ value < 0.001$ adjusting the chi square test for double classification. Our study has also showed a M : F 1: 1.33 unlike the data from USA has been shown ,in Standard Thoracic surgery text books, that regardless of race, men are affected three to four times as often as women .The $X^2 = 54.6$ $p\ value < 0.001$[6].

The reason that oesophago-gastric Junction (GEJ) and Proximal gastric cancer are categorized as separate entity comes as result of that the data were collected based on the 2002 AJCC classification before the adoption of the 2010 AJCC classification which included Esophago-gastric Junction (GEJ) and proximal 5 cm of the stomach that extend into the EGJ or esophagus as esophageal cancers (the so-called Siewert III EGJ tumors)[7].

The tumors otherwise unspecified were those tumors which did not have preoperative tissue diagnosis to determine whether they are benign or malignant because either the sites are inaccessible ,since image guided tissue biopsies were not yet available at the time of their presentation or tissue needed excisional biopsy for definitive diagnosis.

The observation that infection remains an important pathology for admission is an understandable one as the country is developing and communicable diseases are still relevant. Most of the patients who presented with infectious pathology had diseases related to tuberculosis. Except for the 16 cases of Hydatid cyst of the lungs the remaining 36 cases that included empyema thoracis, constrictive pericarditis, pulmonary aspergiloma, destroyed lung and pulmonary fibrosis were a result of tuberculosis. These data goes in conformity with the last updated data by WHO which puts the prevalence of Tuberculosis per 100000 of the population to be 237caese compared to the global average of 170[4].

Adem A. et al[3] had also noted that the number of Cardiothoracic and vascular disease cases was low which might have been due to the fact that TAH lacked specialized services in these area, though the study failed to provide the number. In that study they have concluded that although TAH is a tertiary institution meant for managing special referred cases, the majority of admissions were simple or common problems that could have been managed in secondary level hospitals.

In our study we observed that there is an apparent shift from the previous study as there are 274 cardiothoracic and vascular surgical patients admitted in a single year unlike the rarity 14 years back. This change can reasonably be attributed to the fact that the institution has started the subspecialty training in the field even though other factors may contribute .for the last couple of years the country has seen a dramatic change in its overall growth which also manifested with the number of teaching and non teaching hospital which are capable of managing the simple and common problems. The availability and betterment of investigative modalities in the recent years increasing diagnostic capacity for otherwise difficult to diagnose neoplasms may be an additional factor in the raise of the number of malignant cases while not downplaying the increased awareness on the part of the medical community as well as the patient.

Conclusion

There is an observed change in the male to female ratio with female preponderance in the general cardio thoracic and vascular surgery admission and among the esophageal cancer cases. Neoplasms are leading causes of cardiothoracic admission surpassing non neoplastic in general and infectious cases in particular. Malignant cases are the number one pathologies for cardiothoracic and vascular surgical admissions. The number of cardiothoracic and vascular surgery cases has increased since the establishment of the cardiothoracic surgery fellowship program.

Recommendation

It may be right time that the institution and the country at large institute a Cancer registry. Further study is needed to have an impact in resource allocation at the institution.

Acknowledgment

We would greatly thank Dr Zebenay Bitew for his help on data entry for this study.

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