SACROCOCCYGEAL TERATOMA: A 4-YEAR EXPERIENCE AT KOMFO ANOKYE TEACHING HOSPITAL

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Abstract

Background: Sacrococcygeal teratoma (SCT) is the most common solid neoplasm in neonates with an estimated prevalence of 1 in 40000 live births. The purpose of this study is to review the clinical characteristics and determine the outcome of surgical treatment of neonates and children with SCT in our environment.
Methods: A retrospective review of all patients treated for SCT at the paediatric surgery unit between January 2006 and February 2010 was conducted and the results analysed.
Results: In all, 18 children were treated over the study period with a female preponderance of 3.5:1. Eleven patients (61%) presented by the 2nd week of life with a modal age of 5 days. The oldest child was 2.5yrs. Fifteen (83%) tumours were classified as Altman type I whilst 16 (88.9%) and 17 (94.4%) were respectively cystic and benign. Ultrasonography was the imaging investigative tool used. The largest tumour measured 35cm by 26cm. There was no correlation between age and size of tumour. Excision of tumour was by sacral approach via a chevron incision mostly as an elective procedure. Six (33.3%) were done as emergency procedures for various reasons. There was 89.9% surgical site infection with varying degrees of wound dehiscence. One child had a sigmoid colostomy on account of repeated wound breakdown from faecal contamination. Most patients do not comply with follow-up appointments.
Conclusion: Sacrococcygeal teratoma is mostly a benign tumour and prompt and complete surgical excision will provide good prognosis. Complex investigative tools are usually not necessary in the management of patients with this tumour. We recommend education of parents on the necessity to comply with follow-up appointments in order to capture long term sequelae.

Key words: Sacrococcygeal teratoma, coccygectomy, abdomino-sacral incision.

Introduction

Sacrococcygeal teratoma (SCT) is the most common solid neoplasm of newborns with a reported incidence of 1 in 35000-40000 live births[1,2]. The tumour arises from the Hensen's node which is made up of totipotent primitive cells[3]. Most patients present during the neonatal period with a sacral mass. However, the few with intrapelvic tumours usually present late outside the neonatal period. It has a malignant potential which parallels the age of the patient at presentation[4]. Complete resection of the tumour soon after birth provides an excellent prognosis[5,6].

In the western countries, tumour registries give detailed clinico-pathological features of this germ cell tumour. However, there is scarcity of reports from our part of the world and from Ghana in particular, where there is no tumour registry to provide information on tumours.

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The aim of this study was to review the clinical characteristics of sacrococcygeal teratoma seen in children at the Komfo Anokye Teaching Hospital over the study period.

Methods

The medical records of all children with SCT treated at the paediatric surgery unit between January 2006 and February 2010 were studied retrospectively. Clinico-pathological data were collected from the original medical records. Children with lumbo-sacral myelomenigoceles who were initially misdiagnosed as SCT were excluded from the study. The data was analysed for clinical characteristics and histopathological outcome.

Results

Eighteen children were seen over the study period with a sex distribution of 14 females to 4 males giving a ratio of 3.5:1. The ages of the children ranged between 2 days and 2.5 years with a modal age of 5 days. Eleven children (61.1%) presented within the first two weeks of life. Most of the tumours (15), representing 83.3% were Altman type I whilst there was one each of types II, III and IV (Figures 1 and 2). The tumours ranged in size between 5 cm ×6 cm and 35 cm× 25 cm. However, there was no correlation between tumour size and age of patients. Sixteen (88.9%) of the excised tumours were purely cystic lesions. Of the remaining

two one had some solid components comprising of cartilage and tuft of hair while the other was entirely solid and was from the 2.5 year old child.

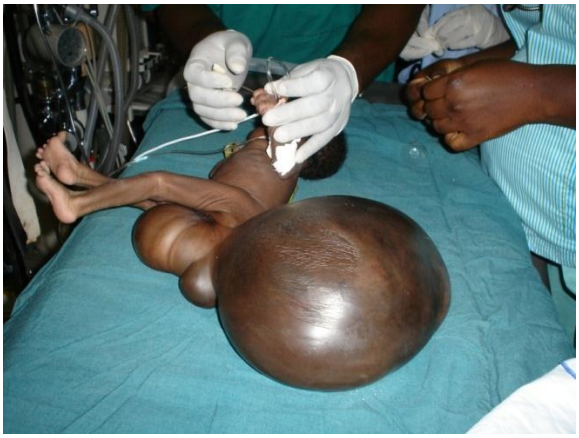

Figure 1: A huge Altman type I SCT

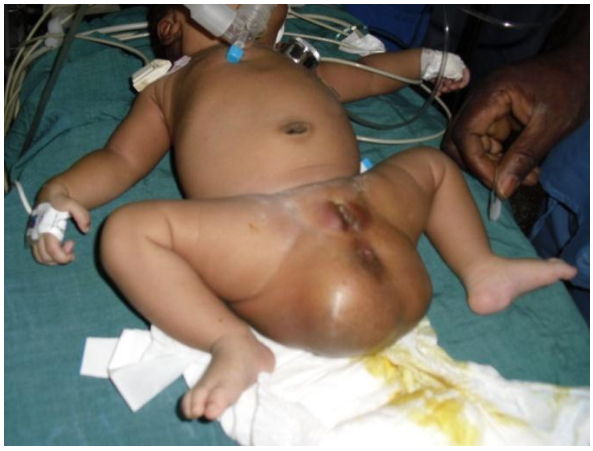

Figure 2: Altman type II SCT

Malignancy was found only in this one with entirely solid tumour which was an embryonal cell carcinoma. Ultrasonography was the sole investigative tool used in all the patients. Only 2 babies had α-fetoprotein assays done but all patients had their full blood count as well as grouping and cross-matching performed before surgery. The α-fetoprotein assays were normal for their ages. Three babies were haemotransfused with 20ml/Kg because of excessive bleeding from ruptured tumours or intraoperative bleeding.

All except one of the tumours were excised through a sacral approach using a chevron incision with the patients in the prone jack-knife position. The remaining tumour was excised using a combined abdomino-sacral incision. In six patients the excisions were carried out as emergency surgeries either because of complications or of an imminent threat of abandonment by the mothers. The remaining 12 were excised as elective surgeries. Surgical site infection with varying degrees of wound breakdown was the main post-operative complication and occurred in 16 children. One child had a sigmoid colostomy performed because of repeated wound break-down from faecal contamination. One child died from excessive bleeding post operatively. Only one child complied with follow-up appointments up to date and no long term sequelae have been noted yet.

Discussion

The earliest record of SCT was in the cuneiform tablet of the Babylonian Chaldeans between 625-539BC[3,5]. This neoplasm has been shrouded in mystery since then. The Chaldeans regarded this protuberance in the new born infant as an omen of prosperity rather than medical curiosity[5]. In certain African cultures, these babies are regarded as monsters, demons and babies from rivers, deities and sexual misconducts and as such a taboo to have such a baby[8, 9]. Such babies are subjected to all forms of inhuman treatment and become victims of infanticide soon after birth[8,10]. Today, much is known about this interesting tumour. It is known to be a germ cell tumour and considered as a displaced ovum or a *fetus-in-fetu*[5].

In our series, eighteen children were seen over four years. This incidence is on the higher side compared to the 33 children in 25years reported from Canada and two studies comprising 38 and 21 children, each study conducted over 18 years, in Nigeria[8,11,12]. This may be attributed to the introduction of the free maternal care and national health insurance. We still strongly believe the real incidence is far higher than this considering the fact that quite a significant number of home deliveries supervised by traditional birth attendants still occur in our part of the world. Such babies who are born with SCT are killed immediately after birth due to socio cultural beliefs[8, 10]. The female preponderance of 3.5:1 is consistent with literature reports. However, an equal sex distribution was found by Waklhu *et al.* in India[13]. Most of the babies had predominantly external tumours (Altman type I). This explains why 61.1% of our babies presented within 2 weeks of life and compares favorably with similar findings in Nigeria[8,12]. In contrast, studies from India showed Altman type IV tumours predominating and as many as two-thirds presenting outside the neonatal period[13].

Most of the tumours in our series were benign because they were entirely cystic which is consistent with the reports by Khanna and his colleagues [14] and Ein *et al*[11]. Malignancy was found in the only child with an entirely solid tumour, who was diagnosed at the age of 2 ½years. This child had an Altman type IV tumour which was likely to have accounted for the late diagnosis. Berry *et al*[15] observed a malignancy rate of 33.3% in tumours seen after 1 year of age whilst Donnellan and Swenson reported over 90% malignancies in infants over 2 months of age[16]. This consistency in the findings from other large series underscores the fact that risk of malignancy in SCT parallels the age of the

patient. When this malignant transformation takes place is as yet an unanswered question. Hence the tumour should be excised at the first opportunity no matter how small or apparently insignificant it appears. A finding of cartilage and hair in a SCT is not unusual in the literature[17, 18]. Other studies from different parts of the world have reported finding structures such as bones and teeth in these tumours. Indeed, a completely formed eye[19] and the lower trunk of a human body was found in a tumour in the Czech Republic[20].

For the purpose of investigating SCT, ultrasonography alone suffices and is readily available and harmless. In the western countries, prenatal diagnosis with a planned caesarean delivery and an immediate tumour excision is the current trend of management[18, 21]. Intrauterine intervention in fetuses with large tumours causing complications have been reported[22-25]. Although CT scan and MRI give better characterization of the tumour and its topographical relationship to other structures[2], these are usually not available and are expensive and do not change the eventual mode of treatment. Alpha-fetoprotein is a cardinal tumour marker used to monitor malignant change and recurrence and hence every effort should be made to assay for this marker in children with SCT. Unfortunately due to the lack of reagents in our laboratories this could not be done for the majority of the babies in our series.

The tumours were excised through a chevron incision at the sacral area. One tumour, which was Altman type IV, was however excised through a combined sacral and lower transverse laparotomy incision. The treatment for SCT is immediate surgical excision with coccygectomy. Excision without removal of the coccyx results in recurrence in over 30% of cases[2, 5]. Surgical approach depends on the size and topographic location of the tumour. The standard procedure is through a chevron incision from a sacral region. Intraoperative bleeding is minimized by meticulous haemostasis. To this end some authorities advocate control of the median sacral artery early at surgery[26] and there are reports of laparoscopic ligation of the median sacral artery for large and highly vascularised tumours before surgery is undertaken from some centres[27]. The former approach was employed in this study and this explains the low rate of haemotransfusion (16.7%). In our series, about a third of the surgeries were done as emergency sessions because of complications such as rupture of the tumour with bleeding or ulceration of the tumour (Figures 3 and 4). Another reason for performing urgent surgeries was imminent abandonment of babies by their mothers. This emphasizes the strong socio-cultural beliefs and stigma attached to this neoplasm in Africa. Other preoperative complications documented in the literature include bowel obstruction, urinary tract obstruction, hydronephrosis and hydrops fetalis[28,29,30].

The most common post-operative complication in our series was surgical site infection which was as high as 90%. This, generally, is due to the proximity of the surgical site to the anus. An infection rate of 18% and

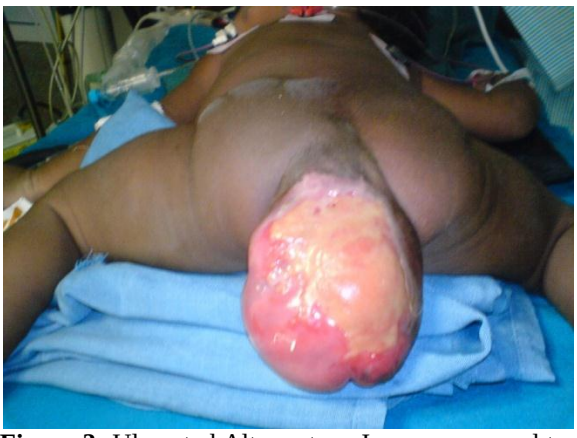

Figure 3: Ulcerated Altman type I sacrococcygeal teratoma

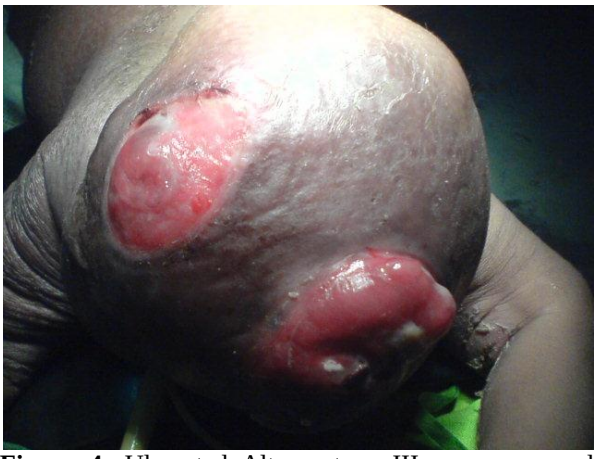

Figure 4: Ulcerated Altman type III sacrococcygeal teratoma

21% were reported from Canada[11] and Nigeria[8] respectively whilst only 2 of the babies in the series by Wakhlu *et al*[13] had surgical site infection. The unacceptably high rate of surgical site infection in our series can be attributed to congestion in our neonatal intensive care unit as well as lack of cooperation from mothers to nurse their babies in the prone position to minimize faecal contamination. One child underwent a sigmoid colostomy because of repeated surgical site infection with complete wound dehiscence. Chirdan *et al*.[8] did 3 colostomies in their study. Wakhlu and his colleagues[13] on the other hand performed preexcisional colostomies for large and obstructing tumours especially type IV tumours.

The only mortality in the study was due to excessive hemorrhage as a result of disseminated intravascular coagulation. This is considered one of the lethal complications associated with the excision of this tumour[31, 32]. Our mortality rate of 5.6% compares favorably with studies from other centres[12, 37]. Some series have reported post-operative functional sequelae such as faecal and urinary incontinence, paralysis and neurogenic bladder[33-36], but no such complications were observed in our study. This is likely because most of

the tumours in this series were predominantly external and so were excised with less perineal dissection and little disturbance to the presacral plexus of nerves. The high rate of non-compliance to follow-up schedules in our series also makes it difficult to have any meaningful assessment of these post-operative complications. Refusal to report for follow-up was also observed in a Nigerian study[12].

Conclusion

Sacrococcygeal teratoma is mostly a benign tumour. Complex investigations are not necessary in the management of babies with the tumour. Prompt and complete surgical excision with coccygectomy will provide good prognosis. However, compliance with follow-up appointments was very poor. We recommend education of parents on the necessity to fulfil follow-up schedules.

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