Original Paper

Sacrococcygeal Teratoma in Children- Experience in a Tertiary Military Hospital

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Abstract

Introduction: Sacrococcygeal teratoma (SCT) is the most common solid tumour found in neonates. Most SCTs are reported in neonates and children, but SCTs are found in adults also. SCTs occur more commonly in females than males. If not detected antenatally, clinical features at birth are prominent in most cases as a palpable or visible mass. Surgery is the principal mode of treatment. The standard treatment for SCT with malignant elements is surgery and chemotherapy.

Objective: To share the experience with sacrococcygeal teratoma, the most common tumour found in newborn in a tertiary military hospital.

Materials and Methods: This observational study was carried out in the Department of Paediatric Surgery, CMH Dhaka during the period of January 2010 to December 2015. During the study period a total of 6 neonates with SCT were admitted in this hospital that were included in the study.

Results: The age of the patients ranged from 3 days to 1 month. Out of them 5(83.33%) were female, 1(16.67%) was male with a female to male ratio of 5:1. Four neonates (66.67%) had type-I and 2(33.33%) had type II SCT. They were not diagnosed antenatally. All presented in neonatal period and were operated as elective cases. Five neonates (83.33%) were born by normal vaginal delivery and one (16.67%) was born by Caesarean section due to cervical dystocia. All patients had elevated alpha fetoprotein (AFP). Posterior sacral approach was sufficient in all the cases and histopathology revealed benign lesion. There was no perioperative death but 1 patient (16.67%) developed malignant recurrence within 1st year and 1 patient (16.67%) developed neurogenic bladder with bilateral hydronephrosis.

Conclusion: SCTs are rare tumours that occur most frequently in neonates. Antenatal diagnosis is possible in many cases in this modern era. Follow up of cases after surgery should not only the look out for tumour recurrence, it includes the management of possible secondary urinary and/or bowel incontinence.

Key-words: Sacrococcygeal teratoma (SCT), Neonates and children, Surgery and chemotherapy.

Introduction

Sacrococcygeal teratoma (SCT) is the most frequent germ cell tumour reported in neonate. It arises from totipotent stem cells, occurring in 1 in 20,000 to 40,000 live births[1,2]. The diagnosis of SCT may not be difficult initially but additional information on imaging studies is required in order to manage the patients adequately[3]. SCT, though histologically benign in most of the cases, has an alarming threat to recur either as a benign or malignant lesion during the 1st 2 years of life[4-6]. Around 35% of babies may have malignant elements[1]. Most SCTs are reported in neonates and children, but SCTs are also found in adults[7]. Antenatal detection is possible in about 50-60% of cases[8,9]. Most of the cases present between the 22nd and 34th week of gestation prenatally[10]. Most of the patients diagnosed postnatally usually have preferable prognosis, whereas the mortality and morbidities of the prenatally diagnosed SCT are high, with mortality rate of 30% to 50%[11]. All babies with the diagnosis of SCT should be delivered at a tertiary level centre where paediatric surgery expertise is available[12]. Patients with tumour larger than 10 cm in diameter will need Caesarean section delivery. Around 11 to 38% of the foetuses having SCT will have other abnormalities of the nervous, cardiac, urogenital, gastrointestinal or musculoskeletal systems[13]. A significant group of patients with SCT have problems with defaecation, urination, or a cosmetically ugly scar[14,15]. SCT develop more frequently in females than males with a ratio of 4:1. But malignant change is more frequent in males[16]. The preferred treatment option for SCT is complete surgical excision including the coccyx[17]. Follow up of cases after surgery should not only look for recurrence but include the management of possible urinary and/or faecal incontinence[18,19]. The mortality rate of SCT reported in neonate is less than 5%, but the mortality rate of SCT diagnosed in utero is 50%[13]. The objective of the study was to share the experience in terms of presentation, treatment and outcome of cases with SCT in a tertiary level military hospital.

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Materials and Methods

This observational study was carried out on all patients with SCT who were admitted and treated in Paediatric Surgery Department of CMH Dhaka during the period of January 2010 to December 2015. Patients with SCT irrespective of age, sex were included in the study. During this period, a total of 6 patients with SCT were admitted in the hospital. Particulars of all these patients were recorded which included age, sex, weight, type etc. Data were collected from detailed history, thorough clinical examination and relevant investigations like serum alpha feto protein, ultrasonography, X-ray, CT scan. All excised tissues were sent for histopathology.

All the patients underwent operative treatment as elective procedure. They were followed up at 01 month, 03 months and 01 year. During follow up clinical examination was done and serum AFP and USG of whole abdomen were done. All relevant clinical information, investigation results, operative procedures, post operative complications and follow up results were analyzed by SPSS. The parents were motivated for long term follow up.

Results

In this study 6 cases were studied as it is quite a rare disease. The age of the patients ranged from 03 days to 01 month and maximum patients 4(66.67%) presented in 1st week (Table-I). Out of them 5(83.33%) were female and 1(16.67%) was male (Table-II). So female to male ratio was 5:1. Four neonates (66.67%) had type-I and 2(33.33%) had type-II disease (Table-III). Five neonates (83.33%) were born by normal vaginal delivery and 1(16.67%) was born by Caesarean section due to cervical dystocia (Table-IV). None of the patients had antenatal diagnosis and all were diagnosed at birth. There was no perioperative death. Four patients (66.67%) had satisfactory outcome. One (16.67%), the only male neonate had a malignant recurrence after removal. One patient (16.67%) developed bilateral hydro-nephrosis with neurogenic bladder subsequently after operation (Table-V). Average hospital stay of cases with SCT in our study was 20 days. Excision biopsy revealed benign lesion in all the cases. None required emergency exploration to save life. All the patients had elevated AFP before surgery which subsequently came to normal except the one who had malignant recurrence.

Table-I: Distribution of patients by age (n=6)

Age in weeks	Patients (n)	%
<01 week	4	66.67
1-2 week	1	16.67
>2 week	1	16.67

Table-II: Distribution of patients by sex (n=6)

Sex	Patients (n)	%
Female	5	83.33
Male	1	16.67

Table-III: Distribution of patients by type of SCT (n=6)

Type	Patients (n)	%
Type-I	4	66.67
Type-II	2	33.33

Table-IV: Distribution of patients by mode of delivery (n=6)

Mode of delivery	Patients (n)	%
Normal vaginal delivery	5	83.33
Caesarean section	1	16.67

Table-V: Outcome of patients with SCT (n=6)

Outcome	Patients (n)	%
Satisfactory	4	66.67
Complication (neurogenic bladder)	1	16.67
Malignant recurrence	1	16.67

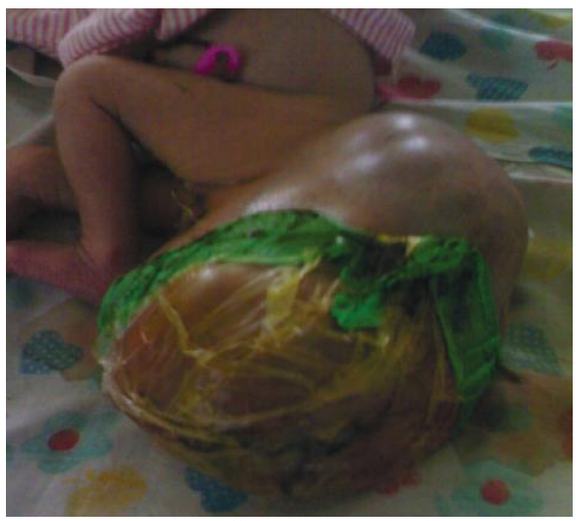

Fig-1: Huge Sacrococcygeal teratoma

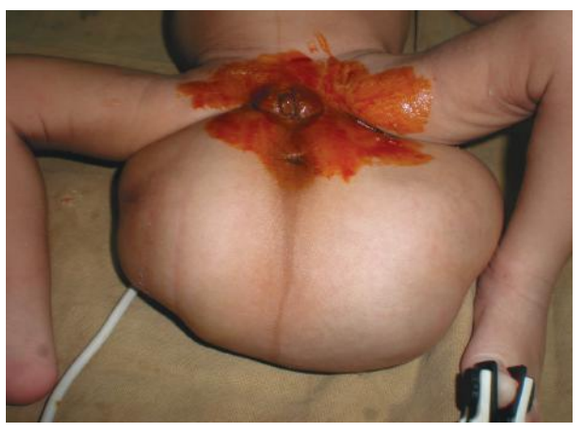

Fig-2: Huge Sacrococcygeal teratoma

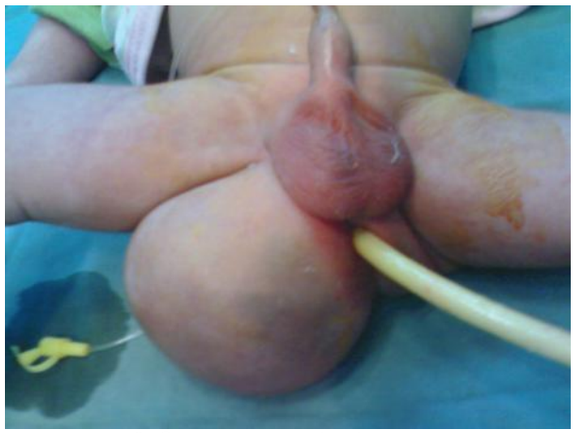

Fig-3: Sacrococcygeal teratoma in male

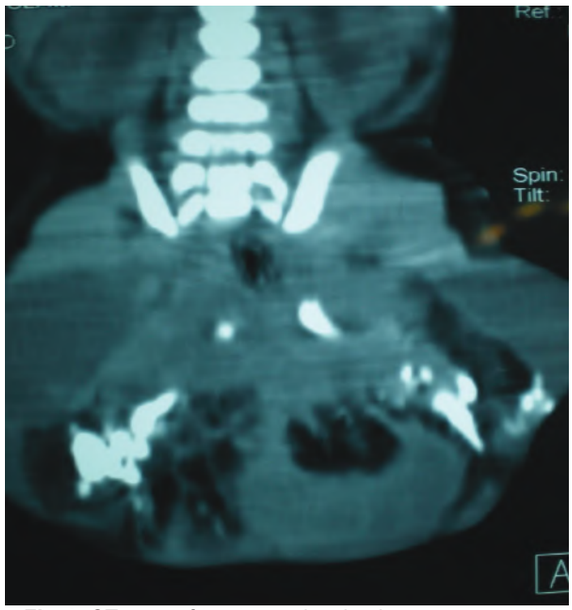

Fig-4: CT scan of teratoma showing bony components

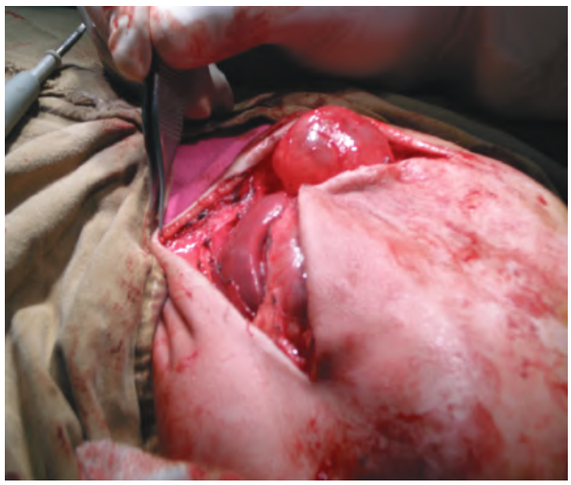

Fig-5: Sacrococcygeal teratoma on exploration

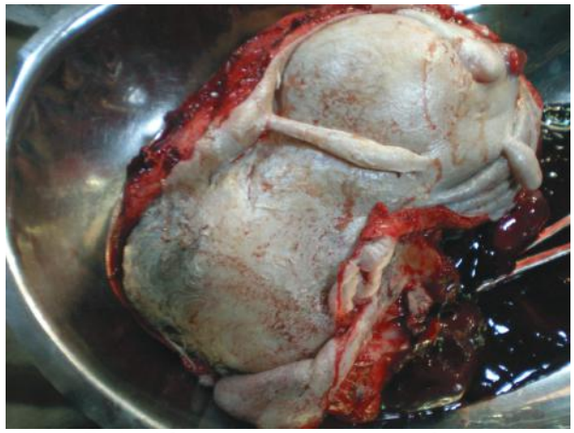

Fig-6: Excised Sacrococcygeal teratoma after operation

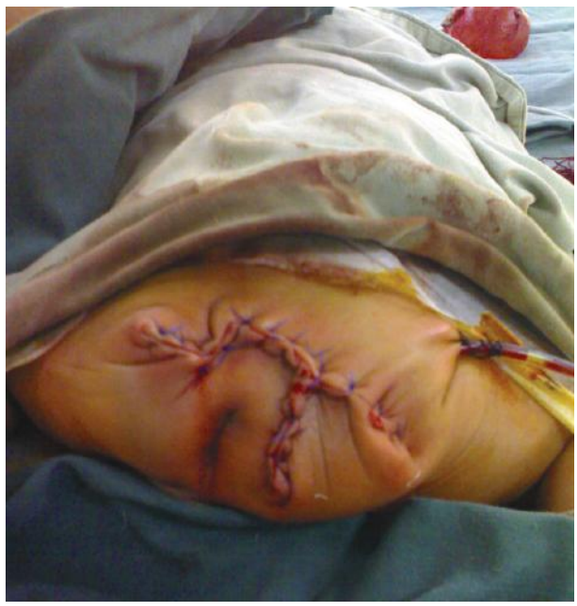

Fig-7: Wound closure after operation

Discussion

Sacrococcygeal teratomas are uncommon tumours found most frequently in newborns, although cases in adults are also reported[20]. SCT is the most frequent congenital extra-gonadal germ cell tumour. Unlike other teratomas, SCT may grow larger than the rest of the baby[7,21]. Usually overall survival of SCT is favourable, but recurrent lesions have poor outcomes[22]. Management protocol of most foetal SCT includes watchful waiting and monitoring rather than any intervention. Serial USG and MRI follow up of foetuses in utero shows that the Altman type can change with time[7]. The earlier the prenatal detection, the poorer the outcome, especially if the detection is due to an USG done for maternal reasons[2]. Cystic SCTs have a better prognosis. Prenatal diagnosis aids adequate parenteral counselling, planned delivery and quick surgical management[23]. SCT having

intraspinal extension is uncommon. It can result in paraplegia due to spinal cord compression[24]. Differential diagnosis of Sacrococcygeal teratoma includes chordoma, sacral lipoma, posterior meningocele, neuroblastoma, giant neurofibroma, extraspinal ependymoma, ependymoblastoma, rhabdomyosarcoma and rectal duplication[7,16,17,23].

Four types of SCT are described. Altman type-I: completely outside, Altman type-II: mostly outside, Altman type-III: mostly inside, Altman type-IV: completely inside[2,6,19,25]. Complete surgical removal is the treatment of choice for SCT. The preferred route for excision of small SCT is through perineum; large SCT may need an additional incision through the abdomen[7]. Surgical resection is possible in utero, in the neonatal period or later. Prematurity is linked with intraoperative and perioperative mortality[26]. In selected cases, laparoscopic mobilization of tumour allows surgeons to clearly identify and ligate the principal tumour-supplying vessels at the beginning of the surgical excision, thus prevent major haemorrhage[27].

SCT type-I and type-II tumours are usually readily diagnosed on prenatal USG[23]. But in this study, although all the six patients had either type-I or type-II lesions none had antenatal diagnosis. This could be because of the reason that USG of the foetus is not routinely and universally performed. Many of them did not have routine antenatal check up. The results are also machine and operator dependent. In this study all the patients presented with protruding mass (type-I and type-II) in the neonatal period. Majority (80%) of SCT are visible at birth as they are exophytic[28]. The incidence of malignancy ranges from 5 to 35% and it is a type called endodermal sinus tumour or yolk sac tumour[29]. Survival for malignant SCTs having metastases is improved with the use of modern cisplatinum-based chemotherapy[14,30]. In this study, female to male ratio was 5:1. Reported incidence[2] of female to male ratio is 4:1. Here four patients (66.67%) had type-I lesion, the commonest type. Dykes EH described 47% of babies to have type-I disease[23]. Prior to the advent of prenatal detection and hence Caesarean section 90% of babies diagnosed with SCT were born at term by normal vaginal delivery[7]. In this study five babies (83.33%) were born at term by normal vaginal delivery.

In this study, all the neonates were operated as elective case and all had elevated AFP before surgery which reduced to normal except one who had malignant recurrence. Outcome was satisfactory in 4 (66.67%) patients, one patient (16.67%) developed bilateral hydronephrosis with neurogenic bladder. In children with significant presacral components, there is high chance of neurogenic dysfunction following excision of the tumours[16]. One patient (16.67%), the male one developed malignant recurrence although initial histopathology of all six patients revealed benign lesion. Malignancy is unusual in the neonatal period but frequently found in male, particularly those with solid SCTs rather than complex or cystic tumours[23]. The incidence of malignant lesion correlates with the anatomical type (8% in type-I, but 38% in type-IV) and age of the baby at diagnosis[16]. A full term neonate with an SCT is likely to have an average stay of two to four weeks in the hospital[2]. In this study average hospital stay was 20 days. Even benign teratomas have a significant chance of recurrence requiring close follow up for around 3 years. As it is an uncommon tumour and the study was done in a single centre the number of cases in this study was relatively less.

Conclusion

SCT is relatively uncommon tumour affecting neonates, infants and children. Antenatal diagnosis when possible, timely referral and subsequent management at tertiary level centre is required for a favourable outcome. Poor management decisions, including interventions that are either premature or delayed can have dire consequences. Long term follow up is indicated for better outcome and to prevent complications.

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