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Childhood Sacrococcygeal Teratoma: A Histopathological Study

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Abstract

Background: Sacrococcygeal teratoma (SCT) is a relatively uncommon germ cell tumor and affecting neonates, infants as well as children with a female preponderance. Age is an important predictor of malignancy in SCT. Early diagnosis and management can provide better outcome. Histologically most of the cases are benign in nature. ***Aim:*** Since there have been paucity of sufficient studies in this field in Bangladesh. The present study was carried out to describe in various clinicopathological features and histopathological findings of SCT patients. ***Materials and Methods:*** The study included 66 patients of SCT operated from 2015 to 2021. A retrospective review of different clinical information and histopathological findings was done from the radiological and pathological records available in the Bangladesh Shishu Hospital & institute, Dhaka. ***Result:*** There were 44 females and 22 males with age ranging from newborn to 08 years. Forty two children (63%) presented in the neonatal age group. Most of the cases (39) have a size of 5-10cm in diameter. There were 66.66% cases of benign (mature), 10.60% immature and 22.72 malignant teratomas. ***Conclusion:*** Predominant cases of SCT are benign in nature histologically and have an excellent outcome after early diagnosis and surgery as well as reducing morbidity and mortality. Delayed presentation and the presence of malignant changes continue to be poor prognostic factors.

Keywords: Sacrococcygeal teratoma, children, mature, immature, malignancy, tumor marker.

INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common congenital tumor in the neonate, reported in approximately 1/35,000 to 1/50,000 live births [1]. It can be diagnosed antenatally or at birth with a prevalence of 3 to 4 times in female children compared to males [2, 3].

Though half of the pediatric teratomas are present at birth, they are infrequently associated with chromosomal or other congenital anomalies [3]. Most of them are benign in the neonatal age group, but the risk of malignancy increases with age. Blizzard reported the first surgical removal of SCT in 1841. Benign SCT has an excellent outcome after early surgery, but the incidences of malignancy increase if resection is delayed [4, 5]. Recurrence with poor survival and anorectal dysfunction are the main problems for patients [6].

In 1973, Altman classified SCT into 4 types based on the external component and intrapelvic/intraabdominal extension of a tumor (American Academy of Pediatrics Surgical Section classifcation) [7]. Type I: predominantly external with a minimal presacral component. Type II: external but with significant intrapelvic extension. Type III: apparently external but predominantly pelvic extending into the abdomen. Type IV: presacral with no external component. The present study was carried out to determine different clinicopathological features and histopathological evaluation of SCTs resected at a tertiary care children hospital in Bangladesh.

PATIENTS AND METHODS

The present retrospective study was carried out at the Department of Pathology of Bangladesh Shishu Hospital & Institute, Dhaka over a period of 06 years from 2015 to 2021. The medical records of 66 patients

operated for SCT were collected from the available data in the department. All the patients underwent routine hematological and biochemical investigations. Serum AFP levels were done in all patients older than 6 months at initial presentation. Ultrasound was done to look for solid and cystic nature along with the extent of mass and to rule out any other abnormality. CT scan and MRI were done to see for the extent of the mass and any associated anomaly. A detailed review regarding antenatal diagnosis, mode of delivery, age at presentation, sex, clinical presentation, site, diagnostic modalities and histopathological diagnosis was performed.

RESULTS

Out of a total 66 patient operated for SCT over a period of 06 years; there were 22 males and 44 females. The age ranged from newborn to 08 years. Forty-two cases presented in the neonatal period, 14 in neonatal to infancy period, 8 in 1-5 year age group and 2 in 05-08 year age group. Only two cases were diagnosed during the antenatal period. All of the children had been delivered through normal vaginal delivery except for six cesarean section.

Patients presented with the chief complain of midline irregular mass with variable consistency in the sacrococcygeal region, except for 08 children where the mass was located laterally in the gluteal region. Also, with the increase in the size of the mass there was progressive anterior displacement of the anal opening. The size of the mass varied from 05 to 26 cm. More than half of the cases had a mass of greater than10cm in dimension. During gross examination, surface ulceration of the mass was seen in 07 cases. In 08 cases (type III and IV) there was a mass in the hypogastric region which was extending into the pelvis. The cases who presented late (type III and IV) had associated urinary retention and constipation. An associated cardiac anomaly was present in 05 cases.

The tumors were classified according to Altman's classification. There were 15 cases of type I, 32 of type II, 11 of type III and 08 cases of type IV. All of the excised specimens were subjected to histopathological examination.

On serial sectioning, about 82% of the tumors were mixed types containing both solid as well as cystic areas, while the rest comprised solid and cystic equally.

On microscopic examination, tumors were classified into benign (mature), immature and malignant categories. The predominant component of mature teratoma was ectodermal tissue, mainly skin and adnexal structures along with mature fatty tissue and cartilage. Immature teratoma was mainly composed of immature neural tissue. Forty four (66.66 %) cases were benign, 7 cases (10.60 %) were immature while 15 cases (22.72%) were malignant teratomas.

DISCUSSION

SCTs are made up of tissues derived from ectoderm, mesoderm and endoderm. Although their early origin is still uncertain, they are thought to arise from the totipotential cells, a remnant of the primitive streak in the coccygeal region [8]. SCTs present at birth as a visible mass in the sacrococcygeal region. However, 14 of our children (21%) presented after the neonatal period. The causes of delayed presentation may include rural background, ignorance, low socioeconomic status and poor antenatal supervision. A female preponderance (2:1) was noted in our series which is consistent with previously published series (3-4:1) in the literature [9-11].

Close antenatal monitor is necessary for diagnosis as well as to look for complications and other anomalies [12]. Altman type I and II SCTs are commonly diagnosed by antenatal sonogram in the 24th-34th weeks of gestation by the presence of a heterogeneous, well-circumscribed exophytic mass at the caudal end of the fetus [13]. Even large type III and IV tumors can be diagnosed prenatally [14-16].

According to Altman's classification they can be small (2-5 cm), moderate (5-10 cm) and large (>10 cm). In our series only 12 cases were categorized as small while rest were large, 15 of which were more than 20 cm in diameter. Size is an important factor as some authors believe that larger tumors are more likely to have immature as well as malignant histology [7, 15]. However, we differ with their opinion as even larger tumors were mature on histology.

In our study, the most common type was Altman type II (48%), followed by I (22%), III (16%) and IV (12%), while others have shown a predominance of type I in their studies [17, 18].

On gross examination, about 82% of the tumors were of mixed type (solid and cystic), which is in concordance with the study by Keslar *et al.*, who recorded 62% mixed type tumors [11]. However, Aly *et al.*, had 40% each mixed and solid teratomas, while the rest were cystic in their study [19, 20].

SCTs are classified as mature, immature and malignant depending on the individual components. Mature teratomas are mainly composed of differentiated tissues and considered benign Fig. 1.

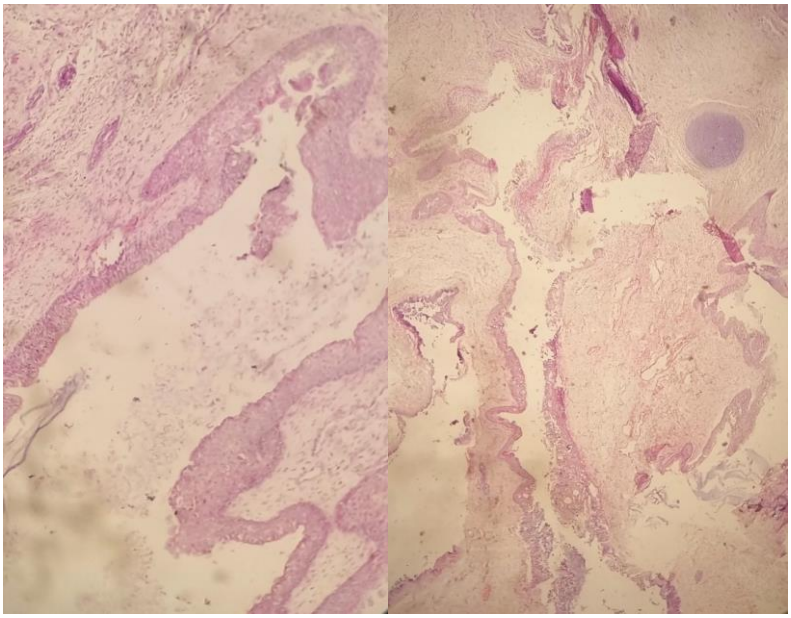

Fig. 1: Photomicrograph showing ectodermal derivatives as skin, mature cartilage and cystic spaces (H&E, low power)

Immature teratoma is characterized by the presence of immature non-malignant tissue [9]. However, some authors believe that differentiation into mature and immature does not correlate with the prognosis of SCT [11]. In our study, 10.60% of cases were immature teratoma which is in accordance with the studies by Aly *et al.,* (13.7%) and Sinha *et al.,* (11.8%) [17, 20].

However, Keslar *et al.,* had a higher proportion of immature cases in their study (32%) [11]. Teratomas with features of yolk sac tumor, choriocarcinoma or embryonal carcinoma are considered as malignant [9]. As reported in literature, risk of malignancy is associated with large tumor size (> 10 cm) in type III and IV due to delay in diagnosis and when the presentation is beyond the neonatal period [7]. However, the only size factor is not responsible for malignant potential as in neonatal age group most of our patients had large-sized (> 10 cm) tumors, but none was malignant [21]. In our study, out of 15 cases (22.72%) patients were malignant.

SCT is associated with congenital malformations in about 5-26% of cases [12]. The most commonly seen anomalies are anorectal and genital, which are seen in about 18-20% of cases [22]. There were 5 cases in our study have anomalies with predominant of anorectal malformation.

CONCLUSION

SCT constitutes a considerable part of the neonatal surgical problems. Most of the cases are benign (mature) in nature. Even large sized tumors, if excised in the neonatal period, have an excellent outcome. While this is a small series, our results are consistent with the literature.

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