

# Childhood sacrococcygeal teratoma: a clinicopathological study

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## Abstract

**Background:** Sacrococcygeal teratoma (SCT) is a relatively uncommon tumor affecting neonates, infants, and children with a female preponderance. Age is an important predictor of malignancy in SCT. Early antenatal diagnosis influences the management and provides a better outcome.

**Aim:** The present study was carried out to describe in detail various clinicopathological features and outcome of SCT patients; as many reports are available from the West, there is a paucity of literature on this entity from the Eastern region, especially India, which has a unique socioeconomic and demographic background.

**Materials and methods:** The study included 52 patients of SCT operated for 16 years from 2000 to 2015. A retrospective review of various parameters was done from the medical case records available in the Department of Pediatric Surgery (PGIMS, Rohtak, Haryana, India).

**Result:** There were 40 females and 12 males with age ranging from newborn to 13 years. Thirty-three children (63%) presented in the neonatal age group. There were 40 cases of benign (mature), 7 immature and 5 malignant teratomas. Four cases had a recurrence on follow-up. Out of 52 patients, 7 died while the others are doing well on follow-up.

**Conclusion:** A prenatal diagnosis of SCT is essential for reducing morbidity and mortality. Delayed presentation and the presence of malignant changes continue to be poor prognostic factors. Strict follow-up by clinical examination, ultrasound and tumor markers is mandatory to look for any recurrence.

## Keywords

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

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## 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].

Though half of the pediatric teratomas are present at birth, they are infrequently associated with chromosomal or other congenital anomalies. Most of them are benign in the neonatal age group, but the risk of malignancy increases with age. Prenatal diagnosis is becoming increasingly common.

Blizzard reported the first surgical removal of SCT in 1841. The landmark article detailing the still contemporary surgical technique for excision of SCTs was written by Robert Gross in 1951 [3]. He stressed that the entire coccyx should always be excised along with the neoplasm because little nests of neoplastic cells are commonly found in or immediately adjacent to the bone [4].

Benign SCT has an excellent outcome after early surgery, but the incidences of malignancy increase if resection is delayed [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 classification) [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 various clinicopathological features and outcome of SCTs treated at a tertiary care center.

## Patients and methods

The present retrospective study was carried out at the Department of Pediatric Surgery (PGIMS, Rohtak, Haryana, India) over a period of 16 years from 2000 to 2015. The medical records of 52 patients operated for SCT were archived from the medical records available 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 cystic nature, the extent of mass and to rule out any other abnormality. X-ray was performed to demonstrate calcification. CT scan and MRI were done to see for the extent of the mass and any associated anomaly. All patients were managed surgically. They were catheterized first and placed in prone position, then anal canal was packed to avoid any injury to the rectum. The posterior approach did a complete excision of the mass along with coccyx through an inverted chevron incision. In type III cases where the mass was extending into abdomen, the abdominal component of the tumor was mobilized per abdominally then the sacro-perineal component of the tumor was excised in the prone position. In cases where the tumor was hypervascular with the invasion of surrounding vital structures, debulking of the tumor was done. The wound was closed in layers with a drain in position.

Chemotherapy was given in cases with malignant teratomas.

The children were put on regular follow-up on 6 months basis with clinical examination including per-rectal examination and abdominal ultrasound to look for any local or pelvic recurrence. Serum hCG and alpha-fetoprotein were advised in all children older than 6 months at first presentation

A detailed review regarding antenatal diagnosis, mode of delivery, age at presentation, sex, clinical presentation, site, diagnostic modalities, surgical procedure, postoperative morbidity and mortality, histopathological diagnosis and follow-up was performed.

## Results

Out of a total 52 patients operated for SCT over a period of 16 years, there were 12 males and 40 females. The age ranged from newborn to 13 years. Thirty-three cases presented in the neonatal period, 10 in neonatal to infancy period, 5 in 1-5 year age group, 1 in 5-10 year age group and 3 children were

more than 10 years old. Because of poor antenatal supervision, only 4 cases were diagnosed during the antenatal period. All of the children had been delivered through normal vaginal delivery except for one cesarean section.

Patients presented with the chief complain of midline irregular mass with variable consistency in the sacrococcygeal region, except for 2 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 3 to 30 cm. About half of the cases had a vast mass of more than 15 cm (**Fig. 1**). Surface ulceration and infection of the mass was seen in 2 cases. In 5 cases (type III and IV) there was a mass in the hypogastric region which was extending into the pelvis. Cases who presented late (type III and IV) had associated urinary retention and history of constipation. An associated cardiac anomaly was present in 2 cases.

The tumors were classified according to Altman's classification. There were 13 cases of type I, 28 of type II, 8 of type III and 3 cases of type IV. All of the excised specimens were subjected to histopathological examination (**Fig. 2**).

On serial sectioning, about 70% of the tumors were mixed types containing both solid as well as

cystic areas, while the rest comprised solid and cystic equally.

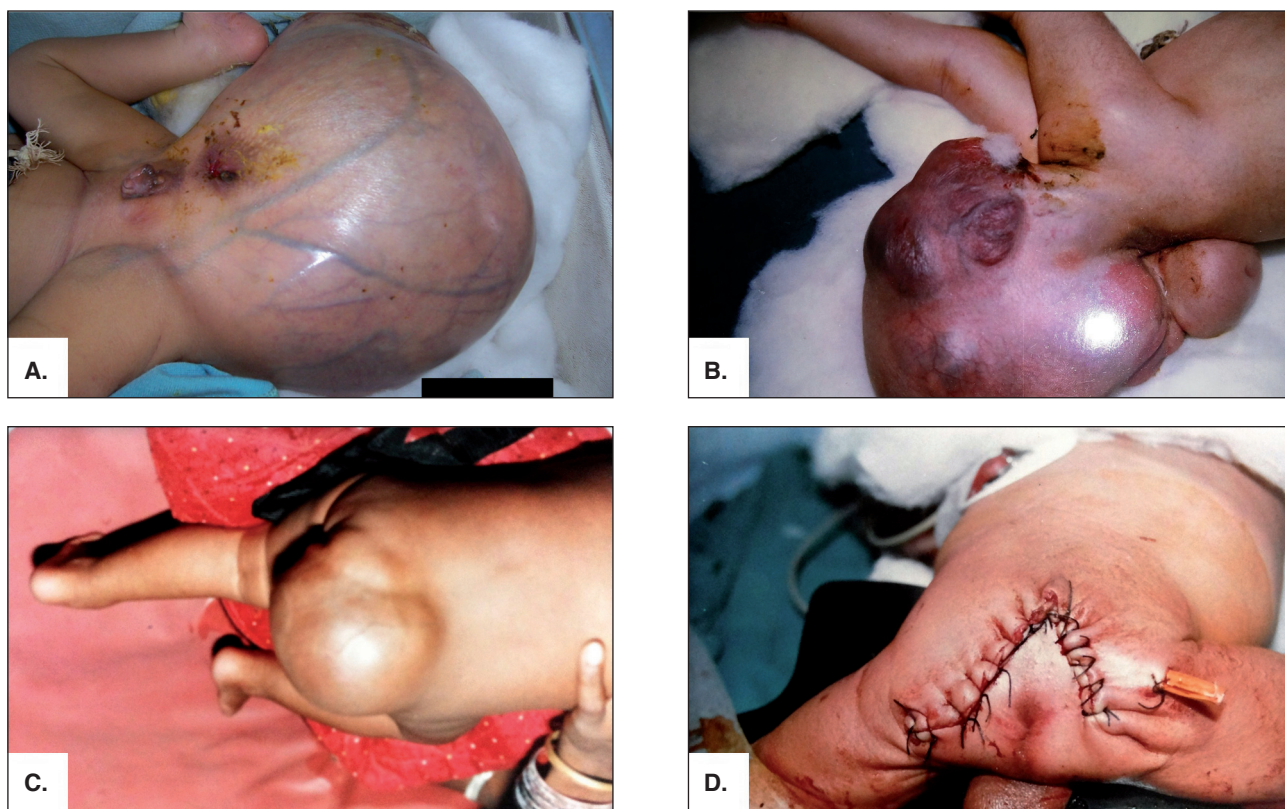
On microscopic examination, tumors were classified into good (mature), immature and malignant categories. The predominant component of mature teratoma was ectodermal tissue, mainly skin and adnexal structures. Immature teratoma was mainly composed of immature neural tissue. Forty (77%) cases were benign, 7 cases (13%) were immature while 5 cases (10%) were malignant teratomas.

All of the patients had an uneventful post-operative period, and the drain was removed on the 5<sup>th</sup> postoperative day. Wound infection was seen in 5 cases.

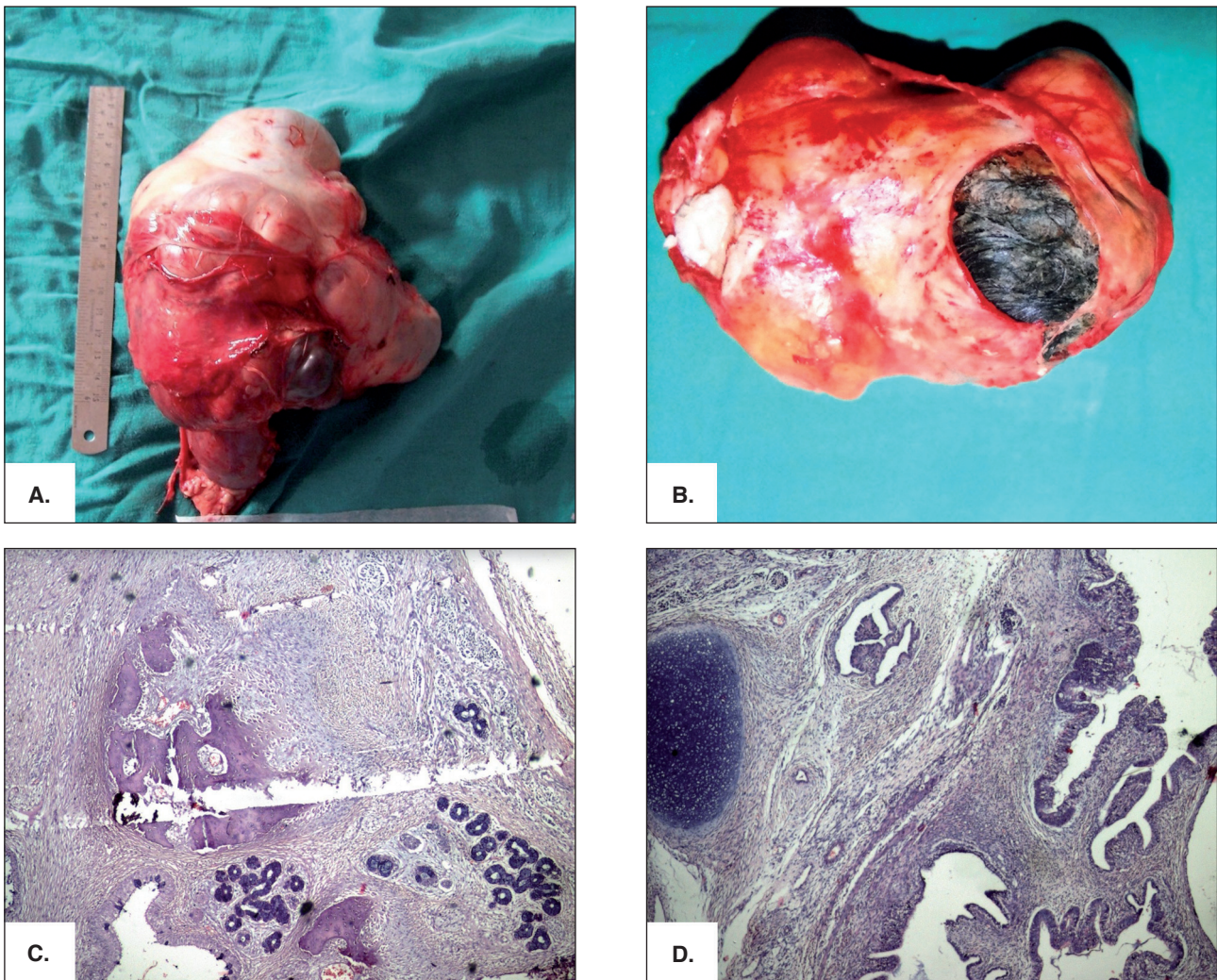
On follow-up till date, a total of 4 patients had recurrences which were found to be malignant on subsequent histopathological examination. A total of 7 children died, of which 2 were neonates. The rest of all children are well. None of the patients had a stool or urinary incontinence.

## 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 of Hensen's



**Figure 1.** Children with huge sacrococcygeal teratoma (SCT), preoperative (**A-C**) and postoperative (**D**).



**Figure 2.** Excised specimen of sacrococcygeal teratoma (SCT) (A, B) and microscopic photomicrograph showing an admixture of bone, cartilage, intestinal and respiratory epithelium (C, D).

node, a remnant of the primitive streak in the coccygeal region [8].

Majority of SCTs present at birth as a visible mass in the sacrococcygeal region. However, 19 of our children (37%) presented after the neonatal period. The causes of delayed presentation may include rural background, ignorance, illiteracy, unawareness, low socioeconomic status and poor antenatal supervision. A female preponderance (3.3:1) was noted in our series which is consistent with previously published series (3-4:1) in the literature [9-11].

Close antenatal supervision is necessary for diagnosis as well as to look for complications like placentomegaly, cardiomegaly, or nonimmune hydrops fetalis [12]. Altman type I and II SCTs are commonly diagnosed by antenatal sonogram in the 24<sup>th</sup>-34<sup>th</sup> 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]. The antenatal pickup was extremely poor in our patients (4/52), which may be because of poor antenatal supervision, as most of our patients are from rural background.

#### *Size*

According to Altman's classification they can be small (2-5 cm), moderate (5-10 cm) and large (> 10 cm). In our series only 6 cases were categorized as small while rest were large, 3 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 histology and may lead to greater intraoperative blood loss [7, 15].

However, we differ with their opinion as even larger tumors were mature on histology and were not associated with excessive blood loss.

### *Altman's classification*

In our study, the most common type was Altman type II (54%), followed by I (25%), III (15%) and IV (6%), while others have shown a predominance of type I in their studies [17, 18].

### *Surgical excision*

Complete excision of the tumor with coccygectomy is the treatment of choice. The recurrence rates reported in the literature without removal of the coccyx are as high as 37% [19].

Other factors responsible for recurrence are the failure to achieve complete resection of the tumor, tumor spillage, and failure to detect malignant components within the tumor. In our study coccygectomy was performed in all of the cases. On follow-up, 4 (8%) of our cases had a recurrence, of which 1 was a neonate. All of them were proved to be malignant on histopathology.

### *Gross*

On gross examination, about 70% of the tumors were of mixed type, 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 [20].

### *Histology*

SCTs are classified as mature, immature and malignant depending on the individual components. Mature teratomas are mainly composed of differentiated tissues and considered benign. 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, 13% 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].

### *Malignancy*

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 sole 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. Even a 13-year-old child presented with a substantial infected sacrococcygeal mass that was benign on histology [21].

In our study, out of 19 patients in the postneonatal period, 5 (26%) were malignant.

### *Associated malformations*

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 2 cases in our study, both having a cardiac malformation.

### *Complications*

Poor cosmesis was the most common complication mentioned in the literature following surgery [23]. Other include local wound infection, bowel, and urinary incontinence or temporary diarrhea. None of our patients had postoperative bowel or urinary incontinence. One had a rectal injury during operation, which was repaired by primary suturing.

### *Survival*

The survival rate of more than 95% has been documented in the literature [24]. A total of 7 (13%) children died, of which 2 were neonates. One neonate with cardiac anomaly died following operation, while in another child the cause might be tumor emboli. The other 5 were in post-infancy period, 3 malignant on the initial presentation, while 2 having a recurrence.

### **Conclusion**

SCT constitutes a considerable part of the neonatal surgical problems.

It must be diagnosed in the antenatal period and should be managed as early as possible to avoid the risk of malignancy. Even large sized tumors, if excised in the neonatal period, have an excellent

outcome, and intraoperative blood loss can be avoided with close supervision.

### Declaration of interest

The Authors declare that there is no conflict of interest.

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