# Sacrococcygeal Teratoma: A Report of 2 Cases with Review of Literature

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**ABSTRACT**

Teratomas are tumors derived from more than one germ cell layer. Teratomas range from benign, well-differentiated cystic to solid (mature) and malignant (immature) lesions. Arising from totipotential cells, these tumors typically are midline or para-axial and the most common location is the sacrococcyx in 57% cases. Sacrococcygeal teratoma are the most common type of germ cell tumors (both benign and malignant) diagnosed in neonates , infants and children younger than 4 years.We present 2 cases of sacrococcygeal teratoma with yolk sac differentiation, both in males aged 1 ½ years and 10 years.

**Keywords**: Sacrococcygeal teratoma, Coccygectomy, Totipotent cell, Young child.

**INTRODUCTION**

Teratomas are tumors derived from more than one germ cell layer. The word teratoma is derived from the Greek word teraton, meaning monster, used initially by Virchow in 1863. Teratomas range from benign, well-differentiated cystic to solid (mature) and malignant (immature) lesions. Arising from totipotential cells, these tumors typically are midline or para-axial. The most

common location is sacrococcygeal (57%). Because they arise from totipotential cells, they are commonly encountered in the gonads (29%). By far the most common gonadal location is the ovary, although they also occur somewhat less frequently in the testes.

Sacrococcygeal teratoma is the most common solid neoplasm in neonates, with an estimated prevalence of 1 in 35 000–40 000 live births [1-3]. Donnellan and Swenson reported over 90% malignancies in infants over 2 months of age [4]. They can be diagnosed prenatally by fetal ultrasound and 50–70% are detected during the first few days of life. 80% are diagnosed by the sixth month and fewer than 10% beyond the age of 2 years [5]. Reported cases of sacrococcygeal teratomas in adults are rare. They exhibit 2 peaks in incidence: the first at approximately 2 years of age which represents congenital neoplasms and the second in late adolescence [6]. It is a more common tumor in females, with a male: female ratio of about 1: 3–4 [5].

The tumor arises from the Hensen's node which is made up of totipotent primitive cells. Most patients present during the neonatal period with a sacral mass. However, few cases of intrapelvic tumors usually present late outside the neonatal period. It has a malignant potential which parallels the age of the patient at presentation. Complete resection of the tumor soon after birth provides an excellent prognosis.

Malignant germ cell tumors account for 3% of childhood neoplasms and yolk sac tumor is the most common histopathological subtype. Reviews of the literature by Ng et al [6] and Bull et al [7] found a total of 88 reported cases, with only 16 described as malignant or with malignant transformation. Here we discuss our experience with the extremely rare malignant extra-gonadal germ-cell tumor,

i.e., malignant sacrococcygeal teratoma with yolk-sac differentiation in two male children aged 1 ½ and 10 years along with the review of literature.

## CLINICAL SUMMARY

**Case No. 1:** A 1 ½ years old male child presented with a predominant presacral mass with a small external mass overlying the sacrococcygeal region and ulceration of overlying skin with inability to pass urine. On local examination suprapubic mass was felt. Proctoscopy revealed definite encroachment on the rectal lumen by an extrinsic mass located posterior and to the right of the rectum. Ultrasonography (USG) abdomen revealed a huge presecral mass with irregular margins extending into the pelvis and pushing the rectum antero-laterally. MRI abdomen revealed large dumbell shaped well delineated heterogenous solid cystic mass in the presacral space. There was no enlargement of retroperitoneal lymph nodes or the liver. Serum alpha fetoprotein (AFP) was found to be abnormally elevated. A clinical diagnosis of sacrococcygeal teratoma was made. The tumor was excised along with Coccyx in-toto through posterior approach dividing the sacrum and was sent for histopathological evaluation. 3-Drugs (VAC) Chemotherapy was started post–operatively i.e. 4-weeks after surgery. The gross specimen showed skin covered mass of sizes, 2.5cm x 2cm x 1.5cm and 5.5cm x 4.5cm x 10cm, with ulceration and variegated appearance showing solid and cystic areas, attached to the sacral bone but not to any other structure. Histopathological examination of the solid areas of the tumor revealed features of endodermal sinus tumour (yolk sac tumor), along with immature stromal and epithelial elements comprising of neuro-ectodermal tissue. The mature elements such as skin, cartilage, respiratory epithelium were seen in the lining of cystic areas of the tumour.

**Case No. 2:** A 10 years male child, came with the complaints of inability to pass urine and intestinal obstruction since 5 days, with a history of abdominal pain and constipation. Local examination revealed ulcerated skin overlying the sacrococcyx with a huge suprapubic mass. CT scan revealed a presacral mass extending into more than half of the pelvic area with significant mass-effect on adjacent organs. No invasion of the spine was seen. No lesions were seen on chest radiograph. He was operated upon, removing the tumor in one piece along with coccyx after dividing the sacrum through posterior approach. Gross specimen showed a soft tissue mass of size 6 cm x 3.5 cm x 4 cm, along with bony pieces and piece of ulcerated skin of size 4cm x 3.5cm. (Figure 1)

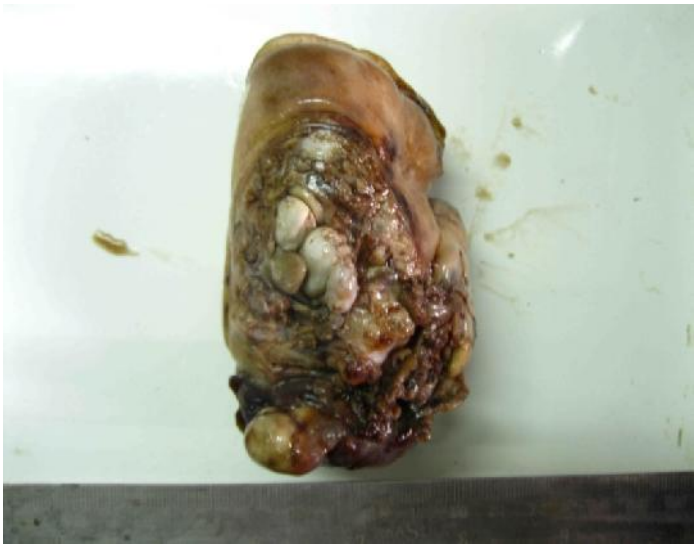

**Figure 1:** Gross specimen showed a globular firm tissue mass of size 6 cm x 3.5 cm x 4 cm, along with bony pieces and ulcerated skin.

Histopathologic examination revealed skin, immature neuroepithelium and fibro-muscular stromal elements with mature cartilage, intestinal goblet cell and pancreatic acini. (Figures 2 and 3) Areas with yolk sac differentiation was also present.

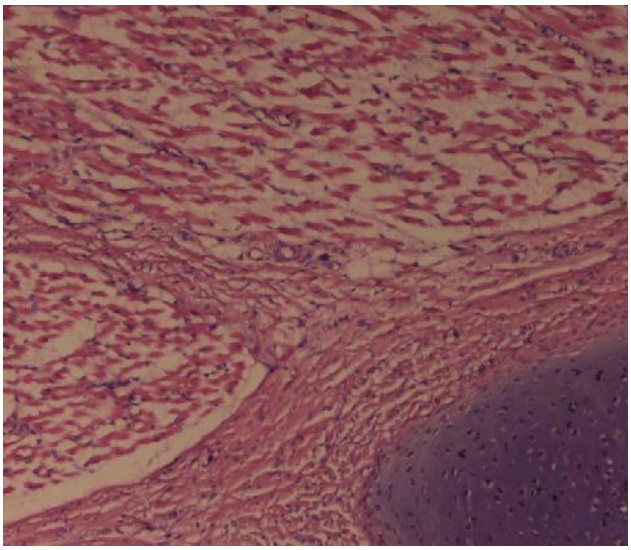

**Figure 2:** Histopathologic examination revealed fibro-muscular stromal elements with mature cartilage. Hematoxylin and Eosin x 40X.

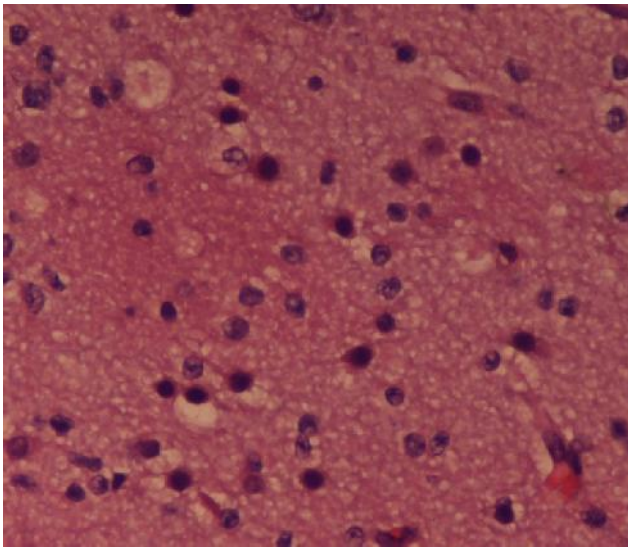

**Figure 3:** Microscopic examination revealed mature neuroepithelium. Hematoxylin and Eosin x 40X.

The histopathological findings confirmed both of these tumor as malignant sacrococcygeal teratoma with yolk sac differentiation, thus confirming the clinical diagnosis. Both the cases were classified as type 3 sacrococcygeal teratoma as per Altman classification.

## DISCUSSION

Sacrococcygeal area is one of the most common sites for teratoma, like other teratomas sacrococcygeal teratomas are included in category of non-seminomatous germ cell tumors. Teratomas are divided into immature and mature tumors depending on the amount of immature elements in them, mostly neuroepthelium. Grossly sacrococcygeal teratoma can be cystic, solid, or of mixed solid and cystic consistency. The solid tumors are more vascular and associated with complications as hydrops, urinary obstrution, still births and distocia.

Microscopically Sacrococcygeal teratoma present in three distinct forms: (1) entirely mature adult type tissues which are clearly benign, (2) a mixture of mature and embryonic tissue, the latter often being of neuralor endodermal sinus (yolk sac) origin and having a malignant potential, (3) tumor consisting partly or entirely of frankly malignant tissues.

ALTMAN with his associates classified the extent of sacrococcygeal teratoma, based on the treatment strategy followed by American Academy of pediatrics [8]: Type 1: Predominantly external tumor, attached to coccyx, with presacral component (45.8%), with No metastasis. Type 2: Tumor having external mass along with significant presacral pelvic extension (34%), With metastasis (6%). Type 3: Tumors visible, with predominant pelvic and intrabdominal mass (8.6%), With metastasis (20%). Type 4: No visible external tumor, entirely presacral mass (9.6%), With metastasis (8%).

About 80% of sacrococcygeal teratoma are of type 1 & 2 with minimal chance of metastasis, and better prognosis as compared to type 3. Type 4 have highest chance of metastasis and worst prognosis.

Gross examination of Yolk Sac Tumors (YST) typically reveals a mass that is predominantly solid and is soft, white, gray, or pale yellow. Cystic degeneration as well as necrosis and hemorrhage are often present. Microscopic patterns of YST are numerous [9]. Several different patterns are usually admixed. They are characterized by the intermingling of epithelial and mesenchymal elements in a specific organoid fashion. Microcystic, glandular-alveolar and papillary formations are common. Many of the cystic spaces are lined by flattened, endothelium-like layer of cells. The stroma can be quite cellular, spindle shaped, and reminiscent of smooth

muscle. Perivascular schiller-duval bodies, which are almost always present in sizable samples but may be absent in limited material, as in biopsies, are the most distinctive features of yolk sac tumor. Periodic acid Schiff positive hyaline intracytoplasmic and extracytoplasmic droplets are consistently seen in yolk sac tumors. Immunohistochemically, these tumors characteristically stain for alpha-fetoprotein.

Most sacrococcygeal teratomas present at birth as sizable masses extending outward from the sacrum and coccyx with varying degrees of involvement of one or both buttocks. Although the majority of sacrococcygeal teratomas are asymptomatic mass, Patients with sacrococcygeal yolk sac tumor present most often with complaints of constipation or buttock swelling. The perianal pain and altered bowel patterns were presumed to be secondary to pressure effects from the tumor. Fecal or urinary symptoms secondary to the obstructions are present in case of large mass. Associated congenital anomalies are common in children with sacrococcygeal teratoma. These associated anomalies include anorectal anomalies, spinal dysraphism, and limb abnormalities.

Sacro-coccygeal yolk sac tumors can be external or internal i.e presacral and intra-pelvic or intra-abdominal extension. Tumors that are predominantly external have a lower malignant potential than prescral that are always malignant. For infant with external masses in the sacro-coccygeal area, the differential diagnosis must include meningomyelocoele, chordoma, neurogenic tumors, lipoma, vestigial tail and hemingioma etc [10].

In infants without external masses, a careful rectal examination is essential in the evaluation of these infants. Presacral or intra-abdominal extension of tumors occurs in 27% of patients. Symptoms of urinary

tract or colonic obstruction are associated more often with malignant than benign tumors. Rarely, invasion of the lumbo-sacral plexus or spinal cord may result in lower extremity weakness and pain.

In Sacro-coccygeal lesions when malignancy is suspected, abdominal and chest x-rays plus sacral film are taken to look for tumor invasion or metastasis [11]. IVP, Barium-enema, USG & CT scanning of the abdomen are obtained in patients who have malignant lesion. Patients with sacro-coccygeal lesions may have an elevated AFP which can indicate malignancy and also should predict recurrence [11, 12].

Complete surgical excision should be attempted in malignant lesion. Since complications with sacro-coccygeal lesions usually involve haemorrhage, control of the tumor vasculature is important. Failure to excise malignant lesions completely had always resulted in death from the disease. The outlook is improving with the use of combination chemotherapy and long-term responses are being noted. Adjuvant VAC chemotherapy (vincristine, actinomycin-D and Cyclophosphamide) has been most extensively used in children with endodermal sinus (yolk sac) tumors [13].

The incidence of re-growth of the tumor at the primary site is extremely high in the absence of radiation. However, in a more common yolk sac tumor of extra gonadal origin, the role of radiotherapy is less certain. Metastatic lesions may require palliative treatment with local radiation [13].

Follow up is integeral part of management of sacrococcygeal teratoma in view of potential for recurrence. All patients must be followed up for at least three years because most of the recurrence occur within this period. Frequent clinical examination, serum AFP level estimation and diagnostic imaging are required to detect early recurrence. Malignant recurrence are treated by re-excision along with combination

chemotherapy and radiotherapy with 40% survival rate [12].

An interesting clinical observation is that the large majority of sacrococcygeal teratomas present at birth are benign, whereas tumors in the same general location discovered after the age of 2 months are often malignant. This has been taken by some to indicate that a malignant transformation has supervened in that short period, we doubt that this is what happened with the 1st case (i.e a 1 ½ years) we are reporting here. The newborn with sacrococcygeal teratoma (SCT) has an excellent prognosis depending on the timing of diagnosis, malignant potential of the tumour and the ease of surgical resection. Although prenatal diagnosis is possible in some cases, this was not possible in our patient since the mother did not attend antenatal care, and therefore our cases presented later as they developed symptoms due to mass effect.

The majority of sacrococcygeal tumors are benign, teratomas [10]. These tumors however have the potential for malignant degeneration. Malignancy is usually limited to a single element, a yolk sac tumor also known as endodermal sinus tumor (EST) [13]. This tumor may less commonly be present in "pure" form [13]. Both the cases of sacrococcygeal teratoma presenting to the department of pediatric surgery had predominant presacral (intrabdominal) with external component and were classified as type 3 sacrococcygeal teratoma as according to the Altman classification, also both of these cases showed yolk sac differentiation along with immature neuroepithelial component in them which was very much confirmed by the histopathological examination. Although about 75% of cases are seen in females, we reported this in two male patients. The exact reason for female preponderance is not fully understood.

## CONCLUSIONS

Infants and young children present with a palpable mass in the sacropelvic region have a greater likelihood of being malignant**.** There is growing body of evidence documenting complete responses of metastatic and primary disease to chemotherapy with long term survival. Therefore, it appears reasonable to conclude that all patients even those with completely resectable tumors should receive adjuvant chemotherapy.

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