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Sacrococcygeal Teratoma: A Retrospective Study Of 23 Surgically Treated Cases At A Tertiary Care Centre

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Abstract
Aim: *To study the cases of sacrococcygeal tumors in our institution*
Materials and methods: *23 cases of sacrococcygeal tumors presented over a period of 5 years and complete excision with coccygectomy was attempted in all. Follow up was done for all cases.*
Results:*Female to male ratio was 2.28:1.The median age at presentation was at 8 days. Altman type 1 was the most common (69.5%) and type 3 was the least common (4.3%) type seen. The antenatal detection rate was poor (5/23). Complete excision with coccygectomy could be done in 21 of the 23 cases.*
There were two recurrences in the completely excised group. One patient presented to us with a recurrence, after being operated once in neonatal age with complete excision. The patient had malignant yolk sac tumor recurrence which was re excised but succumbed to widespread metastatic disease even after receiving chemotherapy. Another patient after undergoing complete excision in the neonatal age group presented with bowel obstruction to another surgeon. Recurrence was detected intraoperatively and a colostomy was done. The patient was then treated with re-excision by us and after two attempts at re excision, complete excision of the recurrent tumor could be achieved and the patient had no further recurrences.
In the incomplete excision group one patient with type 3 tumor was given post operative chemotherapy and developed no recurrences and another patient with type 4 tumor was lost to follow up.
One patient was diagnosed with malignant yolk sac tumor on histopathology after complete excision and was started on post operative chemotherapy but the treatment had to be terminated after one cycle due to DIC. Patient was kept on watchful follow up without further chemotherapy and developed no recurrences.
The serum alpha feto protein level was followed and decreased in an age appropriate pattern in all but one case. The patient had malignant yolk sac tumor recurrence with a serum alpha feto protein level of 4950 ng/ml and finally succumbed to the tumor.
Mature teratoma was the most common sacrococcygeal tumor seen on histopathology (82.6%).
Median follow up was 32 months and wound infection was the most common complication seen (6/23).
There was 1 mortality in the series. The overall survival rate was 95%.
Conclusion: *Sacrococcygeal teratomas are highly curable malignancies. The recurrence rate is low with complete excision and coccygectomy. Even after a diagnosis of malignancy on histopathology, close follow up without adjuvant chemotherapy is a viable option after complete excision and coccygectomy as observed in one of our cases. Close follow up is mandatory in all cases irrespective of the completeness of resection and pathology of the tumor as recurrence can develop in any case and its early detection greatly improves survival.*
Keywords*: Complete excision, Sacrococcygeal tumors, Recurrence*

I. Introduction

Sacrococcygeal teratoma (SCT) occur in 1 in 35,000 live births, are the most common germ cell tumors in infants and newborns, and more commonly affect girls.[1]The female-to-male predominance is 3:1 [2]. Approximately 80% of these tumors are diagnosed within the first month of life[1]. Sacrococcygeal teratoma is the most common tumor in the newborn, even after including stillbirths [3] The histiogenesis of these tumours is thought to be related to incomplete migration of germ cells from yolk sac endoderm to the urogenital ridge in pre sacral tumors or to multi potentent embryonal cells associated with the primitive streak (hensons node) in post sacral presentations[2].

Four types of sacrococcygeal teratomas have been defined on the basis of the abdominopelvic extent and the presence or absence of external extension by Altman et al. [8]

Type 1: predominantly external (46.7%)
Type 2: external with intrapelvic extension (34.7%)
Type 3: visible externally but predominantly pelvic and abdominal (8.8%)
Type 4: Completely internal, this type is also known as presacral or retro rectal teratoma (9.8%)

SCTs can be diagnosed prenatally because of the widespread use of routine obstretic ultrasonography, especially when the examination is performed in the second trimester[4]. Although most teratoma do not adversely affect the foetus or fetal life, the presence of a large solid vascular tumor is associated with significant mortality rate both intrauterine and perinatally[5]. Perinatal mortality is usually related to prematurity or tumor rupture with exanguination or both. Dystocia during vaginal delivery is associated with tumor rupture and is an avoidable obstretic nightmare. Repeated ultrasonographic assessment of tumor size important because the fetus should be delivered by Caesarian section if the tumor is larger than 5 cm or larger than the fetal bi parietal diameter[6].

Most sacrococcygeal teratoma are seen as a visible mass at birth. Although many neonates do not have symptoms, some require intensive care because of prematurity, high output cardiac failure, disseminated intravascular coagulation and tumor rupture or bleeding within the tumor [5,7]. Lesions with a very large intrapelvic component may cause urinary obstruction. Physical examination should always include a rectal examination to evaluate for any intrapelvic component[4]. The most helpful imaging studies consist of plain anteroposterior and lateral radiograph of the pelvis and spine to look for calcifications in the tumor and for spinal defects. Also ultrasonography of the abdominal pelvis and spine is helpful to evaluate the intrapelvic component and rule out meningocele[4]. Further preoperative studies are unnecessary in most newborns. Doppler studies, CT scan and MRI can also be done and these may also provide clues as to the vascular supply of the tumors.

The diagnosis of purely intrapelvic teratoma is often delayed[8]. Children have constipation, urinary retention, abdominal mass or symptoms of malignancy such as failure to thrive.[4]

Risk of Malignancy increases with the more hidden types of sacrococcygeal teratomas
(38% in type 4 vs 8% in type 1) and also increases in older infants and children of age > 2 months [8].

The SCTs seen at birth are usually Altman Type 1 and 2 (87%) [8]. Rarely Type 3 can also be seen in neonates [9]. Type 4 is typically seen later in life as there is no external component [8].

We describe our experience with 23 Sacrococcygeal tumors and review the pertinent literature.

II. Materials And Methods

A retrospective analysis was conducted on 23 cases of sacrococcygeal tumors treated in our hospital between June 2009 and July 2015. The general information, age at presentation, antenatal diagnosis, clinical features, tumor markers, histological diagnosis and post operative complications of all cases in our series were assessed.

III. Results

23 patients with sacrococcygeal mass presented between June 2009 and July 2015. Clinical assessment included a detailed history and physical examination, including a digital PR (per rectal) examination in all. Imaging studies included lateral sacral radiographs and abdominal and chest radiographs. USG (ultrasonography) abdomen was also done in all cases. A few cases also needed CECT (contrast enhanced computerised tomography) abdomen and pelvis also to determine the extent of tumor. Baseline and follow up serum AFP (alpha feto protein) were done.

16 of 23 children presented in neonatal age. Two children presented between 1month to 1 year and 5 children presented between 1 and 3 years of age. The median age of presentation was 8 days. There were 16 female and 7 male patients. Female to male ratio was 2.28:1. Five patients were diagnosed on antenatal ultrasound. No associated congenital malformations were noted.

Most of the patients presented with a visible external mass, four patients presented with constipation and urinary retention, two patients had tumor rupture with bleeding. 16 patients were type 1(69.5%), 2 were type 2(8.6%), 1 was type 3(4.3%) and 4 were type 4(17.3%).

All the patients were counselled for surgery. Complete excision with coccygectomy was attempted in all patients by a posterior approach in prone position, using a chevron incision. Complete excision with coccygectomy could be successfully done for 21 patients (91.30%) All patients of type 1 and type 2 underwent complete excision and coccygectomy. In one patient of type 4 out of four and one patient of type 3 out of one complete excision could not be done and incomplete excision with coccygectomy was done.

Histopathology of 19 tumors showed mature teratoma, 2 were immature teratoma, 1 was malignant yolk sac tumor and 1 was osteosarcoma. One patient with immature teratoma had recurrence as malignant yolk sac tumor.

In the completely excised group there were two recurrences. One child after being operated with complete excision and coccygectomy in the neonatal age, presented with bowel obstruction at 3 years of age and was operated upon by another surgeon where a intraoperative diagnosis of presacral mass was, colostomy was done and patient was referred to us. Complete excision of SCT was attempted but follow up imaging by CT scan showed significant presacral mass (type 4 Sacrococcygeal teratoma) and patient was taken again for complete excision of mass and 3 months later colostomy was closed.

The case series had one mortality. The patient, whose presumably complete excision was done, came back after 2 years with complaints of constipation and urinary retention. Patient had increased AFP levels (4950ng/ml) and on per rectal examination a mass could be palpated posteriorly. Histopathology of previous surgery reported immature teratoma. On CT scan pre sacral mass with infiltration of posterior wall of rectum extending to the anterior rectal wall was seen. Patient was started on chemotherapy and then surgical excision of the mass was done. The tumor had to be shaved off the wall of the rectum. This second histopathology report showed malignant yolk sac tumor. The patient later developed multiple local and metastatic recurrences and died.

In the incomplete excision group one patient with type 3 SCT was given chemotherapy post resection and patient had no recurrences on follow up. Another patient presented at the 3 years of age with urinary retention and type 4 sacrococcygeal teratoma. Laparotomy was done because of large size of the tumour but only a partial excision with coccygectomy could be performed. Histopathology showed immature teratoma. Patient was given chemotherapy. However the patient was lost to follow up.

One male patient presented on day one with a sacrococcygeal mass. Complete excision and coccygectomy was done. Histopathology report showed malignant yolk sac tumor and chemotherapy was started. Patients developed severe neutropenia and went into disseminated intravascular coagulation after first cycle of chemotherapy. Chemotherapy was discontinued and patient was followed closely without any further treatment. Patient did not develop any further recurrences and is now 5 years old with no complaints.

Another patient in whom histopathology showed osteogenic sarcoma was not given chemotherapy and was kept on close follow up. Currently after 2 years patient is free of recurrence. The duration of follow up ranged from 1 month to 71 months. The median follow up was 32 months. One patient was lost to follow up. The serum alpha feto protein level was followed and decreased in an age appropriate pattern. In one patient, 2 years post resection, serum alpha feto protein was found to be raised (4950ng/ml). A mass could be palpated on per rectal examination and the histopathology showed a malignant yolk sac tumor.

One post operative intestinal obstruction and six wound complications were noted making wound infection the most common complication in the series. Two patients have constipation and need laxatives. There were no urinary complaints or complaints of lower limb weakness on follow up.

IV. Discussion

Teratomas are the most common histological subtype of childhood germ cell tumor arising in extra gonadal locations [2]. Teratomas are composed of multiple tissues foreign to the organ or site in which they arise [4]. Sacrococcygeal teratomas are the most common tumors in newborn [3].

A female preponderance of 2.28:1 was noted in our series which is slightly lower as compared to the series of 398 cases by Altman et al. in 1974. The reason for this female predilection is unknown.[8]

Both type 1 and type 4 tumors had a higher incidence and type 2 and type 3 had a lower incidence as compared to that reported by Altman et al [8].

Type	Our series	Altman et al
1.	69.5%.	46.7%
2.	8.6%.	34.7%
3.	4.3%.	8.8%
4.	17.3%.	9.8%

This probably due to the smaller sample size of 23 cases as compared to 398 cases reviewed by Altman et al.Congenital anomalies are observed in 18% of cases , with musculoskeletal and CNS defects being the most common[10]. However no associated congenital abnormalities were noted in our series.

Type 1 and 2 SCTs are commonly diagnosed by prenatal sonogram in the 24th – 34th weeks of gestation [11]. The presence of a heterogeneous, well circumscribed exophytic mass at the caudal end of the fetus is pathognomonic. Even large Type 3 and 4 tumors can be diagnosed prenatally [12]. A close antenatal observation is necessary to look for complications. The presence of placentomegaly, cardiomegaly, or non-immune hydrops fetalis is indicative of a poor outcome [13]. Fetal MRI may provide additional anatomical information [14, 15]. Antenatal pickup was poor in our series (5/23)

The most common approach used is sacral approach [16]. However, in large tumors and especially in type 3 and type 4 SCTs, one should not hesitate to take the sacro-abdominal approach [16]. All our cases were

excised with a sacral approach except one which was a case of type 4 teratoma in a child of three years of age presenting with urinary retention. However even with a laparotomy approach complete excision of the tumor could not be achieved.

The ideal surgery to be performed is complete excision of the tumor with coccygectomy. However this is not always possible in older infants and children due the presence of malignancy in many of these cases. In such cases initial biopsy and neoadjuvant chemotherapy followed by definitive resection after decrease in tumor size is the best approach. In the CCG/POG intergroup study no survival difference was noted between initial and delayed resections. [17]. In our series complete excision and coccygectomy was attempted in all cases but could be done successfully in 21/23 cases i.e. 91.30% of cases. One of the 21patients with complete excision was diagnosed with a presacral mass one month following first resection on CECT abdomen pelvis and was operated upon again and complete excision was done in the second surgery.

In one patient of type 3 SCT, after partial excision chemotherapy was given and patient did not develop any recurrence. Another patient of type 4 SCT with incomplete resection was lost to follow up.

The incidence of different histopathological grades is reported as benign (grade 0) - 75%, immature (grades 1-3) - 11.8%, and malignant - 13.2% [18]. The incidences of histopathological grades were similar in our series. 19 cases were mature teratomas (82%), two were immature teratomas (8.6%), one was malignant yolk sac (4.3%) and one was osteosarcoma(4.3%). The chances of malignancy increase when the tumor size is >10 cm ,in Altman Type 3 and 4 due to delay in diagnosis, with the presence of solid areas and when presentation is beyond the 2nd month of life [8].

It is important to remember that even completely excised mature neonatal SCT has a startling potential to recur (11-22%) either as a benign or malignant tumour during the first 3 years of life [16]. In our series there was one recurrence two years after a presumably complete excision of an immature teratoma. The tumor recurrence was a malignant yolk sac tumor and the patient developed further recurrences even after complete excision of the first recurrence and succumbed to the disease.

The strategy of close follow up without chemotherapy for completely resected teratoma with malignant foci has also been evaluated in literature. Paediatric Oncology Group/Children's Cancer Group Intergroup Study by Marina et al. reported only 4 recurrences in 24 patients of teratoma with malignant foci treated with complete surgical excision and kept on follow up without chemotherapy [19]. One patient in our study was diagnosed with malignant yolk sac tumor on histopathology examination of a completely respected sacrococcygeal tumor. The patient was given chemotherapy with BEP (Bleomycin, Etoposide, Cisplatin). The patient developed neutropenia and DIC after the first cycle. No further chemotherapy was given and patient was kept on follow up. No recurrences have been detected to date.

The complications seen in our patients are comparable to that reported in literature with wound infection being the commonest due to proximity of the surgical site to the anus [20]. Neuropathic bladder and lower limb weakness have also been described as complications but these were not noted in our series.
Bowel constipation has been described as a long term complication which was seen in two patients in our series. These patients need laxatives to relieve constipation.

An excellent overall survival of sacrococcygeal teratoma has been reported (95%) [21]. As noted previously this series also had one mortality and survival of 95.65%.

5. Figures

Figure 1: Preoperative image of a sacrococcygeal tumor presenting as an externally visible mass

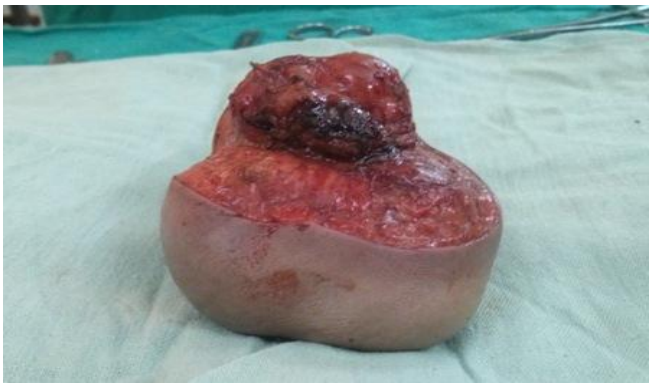

Figure 2: Resected specimen of the Tumor shown in Fig 1

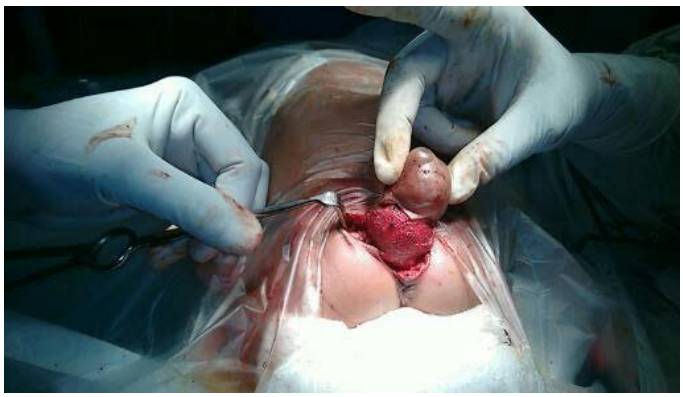

Figure 3: Intraoperative image of a sacrococcygeal tumor dissected from its surrounding structures

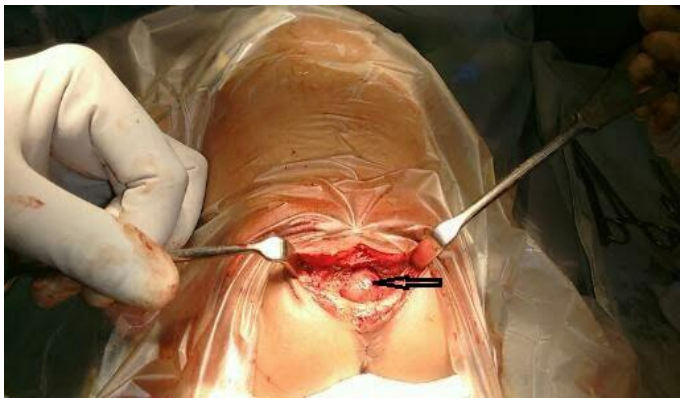

Figure 4: Tumor in fig 3 has been excised without any residual mass. The arrow points to the ballooned rectum

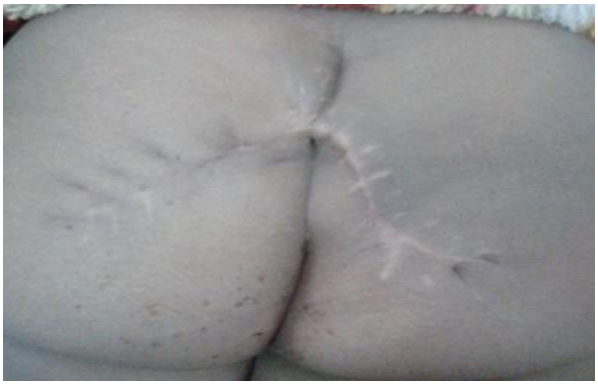

Figure 5: Post operative scar of a chevron incision made for sacrococcygeal tumor excision

V. Conclusion

Most sacrococcygeal tumors present in the neonatal age with a predominantly external component. Older children can present with constipation and urinary retention. Per rectal examination and ultrasonography can clinch the diagnosis in such cases. Complete excision of the tumor with coccygectomy improves the chances for recurrence free survival. Even after a diagnosis of malignancy on histopathology, close follow up without adjuvant chemotherapy is a viable option after complete excision and coccygectomy as observed in one of our cases. Carefully follow up is mandatory in all cases for early detection of recurrence and its timely and appropriate treatment. With rigorous treatment and follow up protocols, extremely high survival rates are attainable.

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