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Original Article

Surgery Section

Clinicopathological Profile of Sacrococcygeal Teratomas: A Retrospective Observational Study from North India

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ABSTRACT

Introduction: Sacrococcygeal Teratomas (SCTs) are the most common extragonadal germ cell tumours during the neonatal period. Although they are typically benign, these tumours carry a significant risk of malignant transformation if not treated promptly. Early diagnosis through clinical evaluation, laboratory tests, and radiological investigations, followed by surgical excision, is crucial in preventing malignancy and ensuring better outcomes.

Aim: To comprehensively analyse SCTs by examining the demographic distribution, clinical features, radiological characteristics, and histopathological profiles of affected patients.

Materials and Methods: This retrospective descriptive observational study was conducted in the Department of Paediatric Surgery of Pandit Bhagwat Dayal Sharma, Postgraduate Institute of Medical Sciences, Rohtak, Haryana, India, from June 2018 to May 2022 involving 11 patients diagnosed with SCT. These patients underwent thorough clinical evaluations, laboratory investigations, and radiological assessments. All patients were treated using standard posterior surgical excision techniques. Postoperative follow-up included both short- and long-term monitoring, focusing on wound infections, recurrence, and metastasis. The data were analysed using Statistical Packages for Social Sciences (SPSS) software version 25.0. Continuous variables were presented as mean±Standard Deviation (SD) or median and range, depending on the distribution. Categorical variables were expressed as frequencies and percentages.

Results: Out of 11 patients, there were nine females and two males. The median age at presentation was 18 days, ranging from one day to three years. The majority of tumours belonged to Altman type 1a7 (63.6%), while types 2 and 3 each constituted two cases (18.2%). Clinical presentation predominantly involved a lower back mass Since birth in 10 (90.9%) cases. Histopathological Examination (HPE) revealed mature teratomas in 9 (81.8%) cases, with immature teratoma and yolk sac tumour each accounting for 9.1% (1 case each). Notably, early presentations correlated with mature teratomas, whereas delayed presentations were associated with yolk sac tumours. One patient with a Type 3 lesion experienced recurrence and metastasis during the follow-up period.

Conclusion: The SCTs are predominantly found in neonates, with a strong female predominance. Early detection generally results in a favourable outcome, while delayed diagnosis is associated with an increased risk of malignancy, recurrence, and poorer prognosis. Careful follow-up is essential, particularly for patients with higher-risk tumour types and later-stage presentations.

Keywords: Extragonadal, Germ cell, Malignancy, Neonatal

INTRODUCTION

The SCTs remain the most common extragonadal germ cell tumours diagnosed during the neonatal period, with an incidence of approximately 1 in 40,000 live births [1]. These tumours are thought to originate from the aberrant migration of primordial germ cells, which fail to reach their gonadal destination during early embryogenesis and instead settle in extragonadal sites, such as the sacrococcygeal region [2]. Although SCTs are often diagnosed at birth due to the visible mass in the lower back, advances in prenatal imaging, particularly foetal Ultrasonography (USG) and Magnetic Resonance Imaging (MRI), have enabled earlier detection and more detailed assessment of these tumours, allowing for more informed prenatal counselling and timely postnatal intervention. Despite significant improvements in early detection, prompt diagnosis and management are still critical, as SCTs have a propensity to turn malignant if not treated early [3].

Recent advances in SCT management have focused on refining surgical techniques and improving long-term outcomes. The posterior approach remains the gold standard for excising benign SCTs; however, more complex lesions, especially those with significant intrapelvic or intra-abdominal extension, may require combined abdominoperineal approaches to achieve complete resection [4]. Furthermore, advancements in perioperative care and anaesthetic techniques have minimised perioperative morbidity, even in cases requiring extensive surgery [4]. In malignant SCTs, cisplatin-based chemotherapy regimens have significantly improved survival rates, particularly in cases where tumours are resected incompletely or metastases are present [5]. Modern protocols now often involve multidisciplinary teams, including paediatric oncologists, surgeons, and radiologists, to tailor treatment plans based on the tumour's stage and histopathology.

Diagnostic work-ups for SCTs continue to centre on both laboratory and radiological assessments. Alpha-fetoprotein (AFP) remains a key biomarker used to monitor the presence of germ cell tumours, as it is typically elevated in patients with SCT [6]. Advances in imaging technologies, such as Contrast-enhanced Computed Tomography (CECT) and MRI, have also contributed to more accurate staging of SCTs, particularly for evaluating the degree of intrapelvic and intra-abdominal extension [7]. These imaging modalities help guide surgical planning and provide critical information for assessing the tumour's malignant potential.

While much progress has been made in the understanding and management of SCTs, gaps remain in the literature regarding optimal treatment timing and the long-term outcomes associated with different surgical approaches, particularly in cases of malignant transformation or recurrence. By adding to the existing literature, present study aimed to analyse SCTs by examining the demographic distribution, clinical features, radiological characteristics, and histopathological profiles of affected patients.

MATERIALS AND METHODS

The present retrospective descriptive observational study was conducted in the Department of Paediatric Surgery at Pandit Bhagwat Dayal Sharma, Postgraduate Institute of Medical Sciences, Rohtak, Haryana, India, from June 2018 to May 2022. Data were collected from the department's admission and discharge registers, as well as Operating Theatre (OT) records.

Inclusion and Exclusion criteria: A total of 11 patients diagnosed with SCT during this period were included in present study. There were no exclusion criteria.

Study Procedure

The SCTs were classified according to the Altman classification system [8], which categorises SCTs into four types based on their anatomical characteristics:

- Type 1: Predominantly external with minimal presacral involvement.
- Type 2: Predominantly external with significant intrapelvic extension.
- Type 3: Small external component with significant pelvic extension into the abdominal cavity.
- Type 4: Entirely presacral with no external presentation.

The parameters studied included demographic data such as age and sex, clinical presentations including the presence of a lower back mass, constipation, or abdominal mass, and radiological features such as tumour characteristics on USG and CECT. Additionally, histopathological features were assessed, with tumours being classified as mature teratoma, immature teratoma, or yolk sac tumour.

Treatment protocol: After thorough clinical, laboratory, and radiological evaluations, patients underwent surgical excision of the mass via a posterior approach. Histopathological Examination (HPE) was performed on all excised specimens to determine the tumour type. Postoperatively, patients were followed up for both short-term outcomes (such as wound infection) and long-term outcomes, including recurrence, functional status, and metastasis. Long-term follow-up included clinical examinations, serial serum AFP levels, and radiological assessments.

STATISTICAL ANALYSIS

The data were analysed using SPSS software version 25.0. Continuous variables were presented as mean±SD or median and range, depending on the distribution. Categorical variables were expressed as frequencies and percentages.

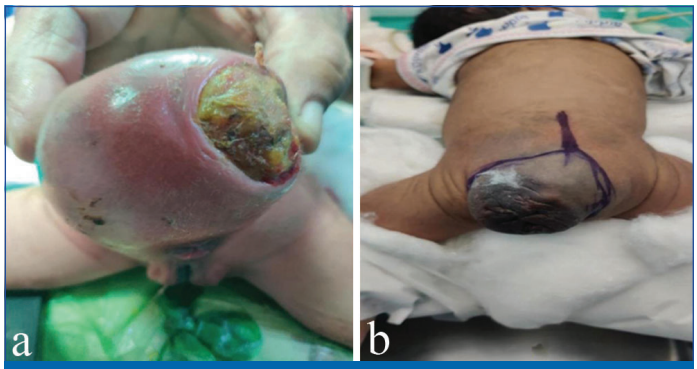
RESULTS

Among the 11 patients diagnosed with SCT, there was a marked female predominance, with a female-to-male ratio of 4.5:1 (9 females and 2 males). A total of 7 (63.6%) patients presented during the neonatal period (within the first month of life), with a mean age of 9.28 days (range: 1-28 days). Three children presented within the first year of life, and one child presented at three years of age. The median age of presentation across all cases was 18 days (range: 1 day to 3 years) [Table/Fig-1].

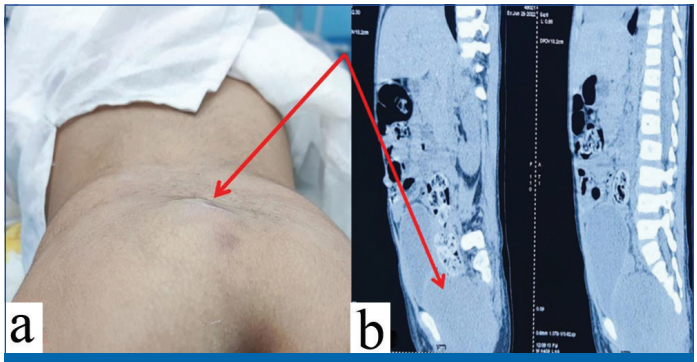
Parameters	Findings (N=11)
Age (years)	<1 month-7 (63.6%)
	<1 year-3 (27.3%)
	1-3 years-1 (9.1%)
Gender	Female-9
	Male-2
	F:M=4.5:1
Clinical presentation	Mass in lower back since, birth-10 (90.9%)
	Constipation-1 (9.1%)
Altman types	Type 1-7 (63.6%)
	Type 2-2 (18.2%)
	Type 3-2 (18.2%)
	Type 4-0
Histopathological Examination (HPE) findings	Mature teratoma-9 (81.8%)
	Immature teratoma-1 (9.1%)
	Yolk sac tumour-1 (9.1%)
Wound infection	2 (18.2%)
Recurrence / Metastasis	1 (9.09%)
Mortality	1 (9.09%)

[Table/Fig-1]: Summary of patients' demographic, clinical, radiological and histopathological findings.

Clinical presentation: The predominant clinical presentation was a lower back mass since birth, observed in 10 of the 11 cases (90.9%) [Table/Fig-2a,b]. One child, who presented at the age of three years, had constipation as the primary symptom. Additionally, this child had a palpable abdominal mass, which was later classified as Altman Type 2 SCT [Table/Fig-3a,b].



[Table/Fig-2a,b]: Clinical pictures of patients with type 1 Sacrococcygeal Teratoma (SCT).



[Table/Fig-3a,b]: Clinical and radiological picture of patient with type 2 Sacrococcygeal Teratoma (SCT) (indicated by red arrow).

Radiological findings: All patients underwent USG followed by CECT of the pelvis and abdomen for detailed assessment. The largest tumour measured 10×8×6 cm and was classified as Type 2, while the smallest tumour, classified as Type 3, measured 6×3×3 cm.

Serum AFP levels: Serum AFP levels were within normal limits (<12 ng/mL) in nine patients. However, two patients had elevated AFP levels (>3000 ng/mL), both of whom were later diagnosed with malignant SCTs-one with an immature teratoma and the other with a yolk sac tumour.

Surgical management and outcomes: All patients underwent tumour resection, with the majority (9 out of 11) achieving complete excision without intraoperative complications. Two patients required colonic diversion: one due to an iatrogenic rectal wall tear during surgery and the other due to metastasis.

Histopathological Examination (HPE): Histopathology revealed-

- Mature teratoma in 9 (81.8%) cases
- Immature teratoma in 1 (9.1%) cases

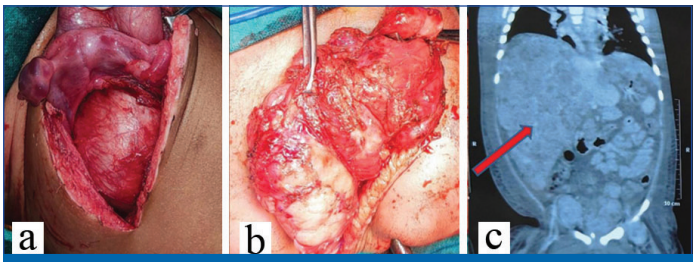
- Yolk sac tumour in 1 (9.1%) case.

Patients who presented earlier were more likely to have mature teratomas, while delayed presentations were associated with more aggressive histologies, such as yolk sac tumours [Table/Fig-4].

HPE (N=11)	<1 month	1 month to 1 year	>1 year
Mature teratoma	6	3	Nil
Immature teratoma	1	Nil	Nil
Yolk sac tumour	Nil	Nil	1 (at 3 years age)

[Table/Fig-4]: Agewise distribution of Histopathological Examination (HPE) findings.

Postoperative and long-term follow-up: Postoperative wound infections were observed in 2 (18.2%) cases, both of which were managed conservatively with daily dressing and cleaning. During long-term follow-up, one patient with a yolk sac tumour and liver metastasis was lost to follow-up. Another patient, initially diagnosed with a Type 3 SCT and treated with combined abdominal and sacral excision [Table/Fig-5a,b], experienced recurrence and widespread metastasis one year after surgery [Table/Fig-5c]. This patient, despite aggressive treatment, succumbed to metastatic disease. The remaining nine patients are doing well, with no evidence of recurrence or functional impairment during follow-up.



[Table/Fig-5]: a,b) Intraoperative images of type 3 SCT operated by combined abdominal-sacral approach and c) Contrast-enhanced CT image of the patient with type 3 SCT with recurrence and liver metastasis (solid red arrow).

DISCUSSION

The SCTs are the most common germ cell tumours in the paediatric age group, with a particularly high incidence in the neonatal period. In our study, the female-to-male ratio was found to be 4.5:1, which aligns with existing literature on the demographics of SCTs [9]. Most cases in our study (63.6%) presented during the neonatal period, consistent with the literature, where approximately 80% of SCTs are diagnosed either prenatally or shortly after birth [10,11]. Notably, no antenatal diagnosis was made in our cohort, which may reflect the lack of antenatal ultrasound facilities in certain rural or underdeveloped areas.

The most common clinical presentation of SCT is a visible mass in the lower back, as observed in 90.9% of our cases. This is consistent with the typical presentation of SCTs, which are predominantly external tumours, classified as Altman Type 1 [12]. In our study, the majority of tumours were classified as Type 1 (63.6%), which corresponds with findings from previous studies [2,3,5]. Types 2 and 3, which involve significant pelvic and abdominal components, were less common, but one case of Type 3 required a combined abdominosacral approach for complete resection. The present case underscores the complexity of managing tumours with extensive intrapelvic and abdominal components, which may increase the risk of recurrence and metastasis due to incomplete resection.

The surgical principle for achieving a complete cure in SCTs hinges on the total resection of the tumour along with the coccyx. Failure to remove the coccyx has been associated with a recurrence rate of 30-40%, with a higher risk of malignant transformation [13,14].

In our study, all patients except one underwent complete tumour resection. Unfortunately, the present patient experienced recurrence and metastasis 11 months after the primary surgery, likely due to microscopic residual disease at the time of the initial operation. This aligns with the literature, where recurrence rates have been reported to range from 10-15%, particularly in cases of incomplete resection and malignant histology [14].

The HPE revealed that the majority of tumours in our cohort were mature teratomas (81.8%), with only one case each of immature teratoma and yolk sac tumour. This finding is consistent with existing literature, where most SCTs are benign at the time of initial presentation [15]. Previous studies have also demonstrated that delayed presentation is associated with a higher likelihood of malignant transformation [16,17].

The treatment of SCTs largely depends on histopathology. Cisplatin-based chemotherapy has been shown to improve outcomes in patients with malignant SCTs, particularly those with metastases or incomplete resections [18]. In our study, one patient with a yolk sac tumour required neoadjuvant chemotherapy after a biopsy revealed malignancy; however, this patient was unfortunately lost to follow-up.

A significant finding in our study was the lack of antenatal diagnosis in all cases, which could be attributed to limited access to antenatal care, especially in rural and underserved areas. Advances in prenatal imaging, including USG and foetal MRI, have dramatically improved the early detection of SCTs, allowing for timely planning of surgical interventions and multidisciplinary care [19].

Several studies have highlighted the benefits of early antenatal detection in improving outcomes [20]. In contrast, delayed diagnosis, particularly in low-resource settings, have been associated with larger tumours, more frequent malignant transformations, and higher rates of recurrence. Addressing disparities in antenatal care and expanding access to prenatal imaging in underdeveloped regions could potentially reduce the number of late-stage presentations and improve overall survival rates for SCT patients.

Future research should focus on large-scale, multicentre studies to further elucidate the prognostic factors influencing recurrence and metastasis. These studies should also examine the long-term outcomes of patients with SCT, particularly those with malignant transformations, and evaluate the efficacy of different treatment modalities, including multimodal chemotherapy protocols.

Limitation(s)

The present study has small sample size with limited number of 11 patients that restricts the generalisability of the findings, and its a single centre study, the results may not reflect the broader population or diverse clinical settings. The retrospective design may introduce bias and limit data completeness, particularly regarding long-term outcomes and one patient was lost to follow-up, affecting the completeness of the recurrence data.

CONCLUSION(S)

The SCTs are the most common germ cell tumours in neonates, typically exhibiting a predominantly benign course if, treated early. Early presentation and complete excision, including coccygectomy, are essential for favourable outcomes. However, delayed presentation, malignant histology, and incomplete resection are associated with poor prognosis and higher rates of recurrence and metastasis. Continued advances in antenatal screening and multimodal treatment, as well as large-scale studies, are necessary to optimise management strategies and improve outcomes in patients with SCTs.

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