International Journal of Contemporary Pediatrics
Jhalani A et al. Int J Contemp Pediatr. 2024 Apr;11(4):388-392
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: https://dx.doi.org/10.18203/2349-3291.ijcp20240725

Original Research Article

Sacrococcygeal teratoma: an experience from a high-volume tertiary institute in North India

Ajay Jhalani, Vinita Chaturvedi, Neeraj Tuteja, Atul K. Khare*

Department of Pediatrics Surgery, SMS Medical College, Jaipur, Rajasthan, India

Received: 27 January 2024
Revised: 06 March 2024
Accepted: 11 March 2024

***Correspondence:**
Dr. Atul K. Khare,
E-mail: mgmatulkhare92@gmail.com

ABSTRACT

Background: Sacrococcygeal teratoma (SCT) is the most common tumour in the newborn. The majority is present in neonates as a sacral mass; however, some may be present late with varied clinical presentation. The study aims to evaluate the clinical presentation and management of patients with SCT in our high-volume tertiary institute in North India.
Methods: This is an observational study of infants and children treated between May 2021 to April 2022 in the department of pediatric surgery, SMS Medical College, Jaipur (a high-volume tertiary institute in North India). Data collected included antenatal diagnosis, mode of delivery, age at diagnosis, clinical presentation, Altman classification, surgical approach, histopathology and complications. Functional results were evaluated clinically and radiologically.
Results: Twenty-one patients (M: F=1:3.2) with a median age of 40 days (range: 1 day to 5.8 years) with SCT were managed at our centre. Nearly, two-thirds of the tumors were either Altman type 1 or 2. Yolk sac tumour was present in 2 (9.5%) patients, while the rest had either mature or immature teratoma. Tumours were removed through a posterior sagittal approach (chevron incision). In five patients, an abdominoperineal approach was used. Early complications were surgical site infection (n=5; 23.8%), superficial wound dehiscence (n=2; 9.5%), complete wound dehiscence (n=1; 2.4%), and urinary tract infection (n=1; 4.7%). Late complications were urinary dribbling or poor stream (5/21; 23.8%) and faecal soiling (n=3; 14.2%).
Conclusions: Most of the sacrococcygeal tumours are benign, and the incidence of malignancy increases with age. Morbidity due to associated malformation and treatment may persist in these patients, especially like urinary complications stream (one-fifth) and faecal incontinence (one-seventh), as seen in our series. A proper long-term follow-up is needed for the management of late complications.

Keywords: Sacrococcygeal teratoma, Tumour, Fecal incontinence, Exploratory laparotomy, Complications, Malignancy

INTRODUCTION

Sacrococcygeal teratoma (SCT) is the most common tumour in the newborn with a reported incidence of 1 in 35,000 to 40,000 births.[1] Approximately 10-20% of the tumours are malignant, and this incidence increases significantly if diagnosed in later life. The sacrococcygeal region is the most common site of teratomas in infants and children, the second most common being the gonads. The origin of the tumour is usually described as the totipotent cells of the primitive knot or Hansen's node, embryonic entities that contribute to the gonadal ridge and eventually end up in the coccygeal region.[2] SCT can be diagnosed antenatally or later postnatally.

The majority are present in neonates as a sacral mass; however, some may be present late. It can be benign or malignant. Benign tumours are treated with simple excision with coccygectomy. The malignant tumours

require a multidisciplinary approach including surgery and chemotherapy. The complexity of the sacral neuroanatomy and its close relationship to pelvic organs adds additional challenges for treatment.[3] Neurological dysfunctions, uncontrollable intraoperative haemorrhage, difficulty in achieving complete resection, and high recurrence rate is the most important difficulty faced by surgeons.

This study focuses on the clinical presentation and management of patients with SCT managed over a period of one year in a tertiary health care centre.

METHODS

This is an observational study of infants and children treated between May 2021 to April 2022 in the department of pediatric surgery, SMS Medical College, Jaipur (a high-volume tertiary Institute in North India).

Sampling

A total of 21 patients with SCT were managed during the period of study at our center. The data were analyzed for relative and absolute frequency and compared by the chi-squared test with Fischer's correction, when necessary. Continuous variables were summarized by measures of central tendency and compared by student's t-test, or by non-parametric test when with non-normal distribution. Statistical package for the social sciences (SPSS) 16.0 statistical package was used. The study was designed according to the guidelines and norms regulating research involving human beings and approved by the research ethics committee of the two participating centers.

Data collected included gender, race, place of birth, prenatal or postnatal diagnosis, results of tests used for diagnostic confirmation, histopathological report, serum tumor marker levels, the presence of early or late surgical complications, frequency of recurrence, postoperative sequelae and deaths, and the maintenance of outpatient follow-up. The diagnosis was made based on clinical examination and is confirmed by radiological investigations (ultrasonography and contrast-enhanced computed tomography (CT).

The management protocol included upfront resection of the tumour. Surgical excision was performed by local (inverted V-shaped incision or Chevron incision) or abdominoperineal approach. Post-operative complications were noted.

Patients who had already been discharged from the outpatient clinic were contacted by telephone, in order to obtain updated information of all cases.

Inclusion criteria

All patients with SCT presented to the institute during the study period were included.

Exclusion criteria

Patients with other sacral pathology were excluded.

RESULTS

A total of 21 patients with SCT were managed during the period of study at our centre. There were 16 females and 5 males with a male-to-female ratio of 1:3.2. The median age of presentation was 40 days (range 1 day-5.8 years). Out of which 11 children presented in less than 1 month of age (mean 8.8 days, range 1-29 days), 6 children presented between 1 month to 1 year of age (mean 81 days, range 32-359 days), and 4 children presented with more than 1 year of age (mean 2.6 years, range 1.5-5.8 years).

The mean age of the mother at the time of delivery was 27 years (19-33 years). History of antenatal diagnosis of sacral mass (Figure 1) was present in 5 of 21. All these 5 patients presented early; 4 presented in the neonatal age group and the other 1 shortly after that. None of these patients underwent any fetal intervention.

Mode of delivery was vaginal in 17 of 21 patients and caesarean section was performed in 4 of 21 patients.

Out of 21 patients, 14 patients presented with a sacral mass (66.6%). 5 patients presented with abdominal mass (23.8%). 2 patients presented with chronic constipation (9.5%) (Table 1).

These patients were further classified into four subtypes according to the Altman classification: Altman type 1 - n=9/21 (42%), Altman type 2 - 5/21 (23.8%), Altman type 3 - 4/21 (19%), and Altman type 4 - 3/21 (14.2%) (Table 1).

Out of the 5 patients with a palpable abdominal mass, 2 patients were graded as Altman type 3 tumours and 3 patients Altman type 4. Out of 21 patients, 16 patients (76.2%, Altman type 1: n=9, Altman type 2: n=5, Altman type 3: n=2, and Altman type 4: n=0) were operated by posterior sacral approach (inverted V-shaped incision or Chevron incision) and 5 patients (23.8%, Altman type 1: n=0, Altman type 2: n=0, Altman type 3: n=2, and Altman type 4: n=3) patients were operated by combined abdominoperineal approach (Figure 1 and Table 1).

Out of 21 patients, histopathology of mature teratoma was found in 15 (71.4%) patients, immature teratoma in 4 (19%) patients, and yolk sac tumours in 2 (9.5%) patients. Mature teratoma was seen more in younger age groups: 9/11 children less than 1 month of age, 4/6 children between 1 month to 1 year of age, and 2/4 children more than 1 year of age. Immature teratoma was found in 2/11 children less than 1 month of age, 1/6 children between 1 month to 1 year of age, and 1 child more than 1 year of age (Table 2).

Table 1: Demographics, clinical presentation, Altman classification and surgical approach of patients.

Demographics	No of patients	Percentage
Age		
<1 month	11	52.3
1 month to 1 year	6	28.5
>1 year	4	19.0
Sex		
Male	5	23.8
Female	16	76.2
Clinical presentation		
Sacral mass	14	66.6
Abdominal mass	5	23.8
Chronic constipation	2	9.5
Altman type		
Altman 1	9	42
Altman 2	5	23.8
Altman 3	4	19
Altman 4	3	14.2
Surgical approach		
Posterior sacral	16	76.2
Altman 1	9	42
Altman 2	5	23.8
Altman 3	2	9.5
Altman 4	0	0.0
Abdominoperineal	5	23.8
Altman 1	0	0.0
Altman 2	0	0.0
Altman 3	2	9.5
Altman 4	3	14.2

Table 2: Histopathological diagnosis.

Parameters	Age <1 month (n=11)	1 month <age <1 year (n=6)	Age >1 year (n=4)
Mature teratoma (n=15)	9	4	2
Immature teratoma (n=4)	2	1	1
Yolk sac tumour (n=2)	0	1	1

In patients with yolk sac tumour histopathology, there was no child with age less than 1 month of age, 1/6 (0.16%) was between 1 month to 1 year of age and 1/4 (25%) was more than 1 year of age.

Early complications included surgical site infection (n=5; 23.8%), superficial wound dehiscence (n=2; 9.5%), complete wound dehiscence (n=1; 2.4%), and urinary tract infection (n=1; 4.7%). The patient with complete wound dehiscence underwent a diversion colostomy. In the early follow-up, urinary dribbling or poor stream was seen in 5 patients (5/21; 23.8%). Faecal soiling was seen in 3 patients (14.2%). In our series, there was no incidence of rectal injury.

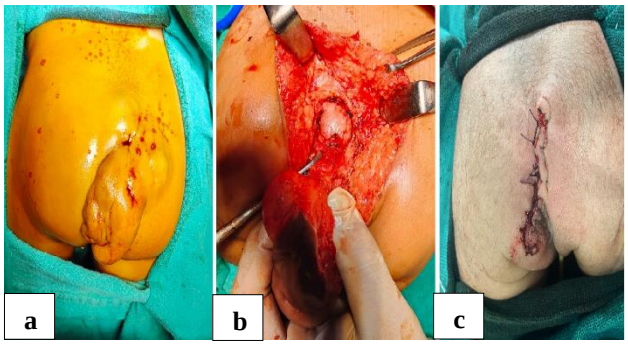

Figure 1: (a) Preoperative picture of a patient with sacrococcygeal teratoma, (b) intraoperative picture, and (c) postoperative picture.

Late complications included an ugly-looking scar of secondary healing (n=3; 14.2%), keloid formation (n=1; 4.87%), and persistent urinary dribbling (n=3; 14.2%). Neither faecal incontinence nor improper gait were present in any of the patients.

DISCUSSION

SCT are the most common germ cell tumors in the pediatric age group. It arises from the totipotent cells from Hansen's node or primitive germ cells.[4] The reported incidence is approximately 1 in 40,000 live births with a male-to-female ratio of 1:3-1:4.[5] The male-to-female ratio in our series is 1:3.2 which is like the rate quoted in literature.

A history of antenatal diagnosis was available in 5/21 patients; none of these patients had any perinatal complications. A literature review revealed that the cases with antenatally diagnosed cases have a high risk of perinatal complications and fetal death from high-output cardiac failure.[6,7] Only 2 of these 5 patients underwent caesarean section for indications related to tumour size. The patients in this series are majority from a community with low socioeconomic status. They have limited resources and struggle to afford basic requirements. The significance of a prenatal diagnosis in cases with SCT, however, cannot be overstated. The arrest of a vaginal delivery may demand an urgent caesarean section. Apart from tumour size other complications are placentomegaly, fetal hydrops, and cardiac failure. The literature suggests that small tumours of size <5 cm in diameter can be managed by vaginal delivery; caesarean section should be performed for large tumours. In our study, five patients were delivered vaginally and had tumour sizes of more than 5 cm (5.4, 5.7, 6, 6.2, and 7.1 cm, respectively). Apart from labour dystocia, the ratio of tumour volume to fetal weight has an effect on prognosis with mortality of 30%–50% in cases diagnosed antenatally.[8]

The patients have been classified by the system given by Altman. Two surgical approaches were used: posterior sacral (inverted V-shaped incision or Chevron incision) and combined abdominoperineal approach. The Altman system cannot be used to predict or correlate with the surgical approach of choice.

The posterior sacral approach provides excellent exposure to resect local as well as pelvic masses. However, this approach has limitations such as the inability for handling tumours extending beyond the sacral promontory, leaving an ugly-looking scar and may result in posterior displacement of the anus giving a pulled-up appearance.[9]

On histopathology, most of the neonatal tumours are mature or immature teratomas.[10] We have observed the same trend in our series. All 11 neonates had either mature (n=9) or immature teratoma (n=2).

In our series, yolk sac tumour was found more commonly in cases (25% of cases) presenting after 1 year of age. In the American Academy of Pediatrics survey, the incidence of malignancy was 7-10% in patients operated on at the age of <2 months but 48-67% if they were treated after 2 months of age.[1] However, in the series by Niramis et al the incidence of malignancy was only 2.4% in patients who underwent surgery at the age of <1 year and 73.3% in the patients operated on after 1 year of age.[11]

The most common complication in the early postoperative period is surgical site infection or dehiscence; this may be related to the proximity to the anus. The rate of wound dehiscence in our series was 14.2% although wound dehiscence rates as high as 90% have been reported in literature.[12]

Urologic and anorectal dysfunction after surgery for SCT have been reported with a variable incidence. Several factors play crucial roles which include the Location of the tumour, local invasion, the extent of resection, compression injury, and surgical complications. Partridge et al have reported urological complications in 33% of patients and anorectal dysfunction in 29% of patients.[13] Ozkan et al have reported neurogenic bladder in more than three-fourths of the patients.[14] In our series, we observed very low rates of such complications. However, our sample size is small, and the data are based on the history given by patients regarding voiding behaviour and related urological symptoms, clinical examination, and sonography with the measurement of post-void residue. Neurodynamic evaluation was performed in our study.

The natural history of the disease and its anatomic location demands long-term follow-up for functional consequences such as gait abnormalities, sexual dysfunction, self-perceived body image, and psyche including self-confidence and self-acceptance. It is observed that in a developing country like ours despite all counselling and advice there is a lack of regular follow-up which limits the availability of data on long-term results. This may be related to illiteracy, excessive working hours to earn, and lack of an alternative support system.

CONCLUSION

Most of the SCTs are benign, and the incidence of malignancy increases with age. Late presentation may also be seen in SCTs. Associated anomalies are not uncommon There was no procedure-related mortality. Morbidity due to associated malformation and treatment may persist in these patients, especially like urinary complications stream (one-fifth) and faecal incontinence (one-seventh), as seen in our series. Concomitant surveillance of urologic and anorectal dysfunction is also essential. A proper long-term follow-up is needed for the management of late complications.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

REFERENCES

1. Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics surgical section survey 1973. J Pediatr Surg. 1974;9:389-98.
2. Ein S, Mancerk Y, Adeyemi S. Malignant sacrococcygeal teratoma endodermal sinus, yolk sac tumour in infants and children: a 32-year review. J Pediatr Surg. 1985;20(5):473-7.
3. Beddis I, Noblett H, Mott M. Effective chemotherapy for metastatic malignant sacrococcygeal tumour. Med Pediatr Oncol. 1984;12(4):231-2.
4. Hirose S, Farmer DL. Fetal surgery for sacrococcygeal teratoma. Clin Perinatol. 2003;30:493-506.
5. Schropp KP, Lobe TE, Rao B, Mutabagani K, Kay GA, Gilchrist BF, et al. Sacrococcygeal teratoma: The experience of four decades. J Pediatr Surg. 1992;27:1075-8.
6. Hedrick HL, Flake AW, Crombleholme TM, Howell LJ, Johnson MP, Wilson RD, et al. Sacrococcygeal teratoma: Prenatal assessment, fetal intervention, and outcome. J Pediatr Surg. 2004;39:430-8.
7. Bond SJ, Harrison MR, Schmidt KG, Silverman NH, Flake AW, Slotnick RN. Death due to high-output cardiac failure in fetal sacrococcygeal teratoma. J Pediatr Surg. 1990;25:1287-91.
8. Akinkuotu AC, Coleman A, Shue E, Sheikh F, Hirose S, Lim FY, et al. Predictors of poor prognosis in prenatally diagnosed sacrococcygeal teratoma: A multi-institutional review. J Pediatr Surg. 2015;50:771-4.
9. Abou Zeid AA, Mohamed MH, Dahab MM, Gad Allah MA, Zaki AM. Sacrococcygeal teratoma excision: A vertical rather than transverse wound closure. Ann Pediatr Surg. 2017;13:207-12.
10. Rescorla FJ. Pediatric germ cell tumors. Semin Pediatr Surg. 2012;21:51-60.

11. Niramis R, Anuntkosol M, Buranakitjaroen V, Tongsin A, Mahatharadol V, Poocharoen W, et al. Long-term outcomes of sacrococcygeal germ cell tumours in infancy and childhood. Surg Res Pract. 2015;2015:398549.
12. Amoah M, Boateng N, Abantanga FA. Sacrococcygeal teratoma: A 4-year experience at Komfoanokye Teaching Hospital. Postgrad Med J Ghana. 2012;1:15-9.
13. Partridge EA, Canning D, Long C, Peranteau WH, Hedrick HL, Adzick NS, et al. Urologic and anorectal complications of sacrococcygeal teratomas: Prenatal and postnatal predictors. J Pediatr Surg. 2014;49:139-42.

Cite this article as: Jhalani A, Chaturvedi V, Tuteja N, Khare AK. Sacrococcygeal teratoma: an experience from a high-volume tertiary institute in North India. Int J Contemp Pediatr 2024;11:388-92.