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Prognostic Appraisal of Sacrococcygeal Teratoma- Authors Experience and Review of literature

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ABSTRACT

Background: Sacrococcygeal teratoma is associated with malformations which may have a bearing on the outcomes of these patients. The study aimed to present its varied clinical presentation, management, and outcome analyses in our geographical region.

Methods: A prospective descriptive observational study was carried out on a cohort of paediatric patients with sacrococcygeal teratoma. All consecutive patients admitted and managed by the authors in a tertiary care teaching institute from 2011 to 2024 were studied. Patients were followed up for at least one year after surgical intervention.

Results: There were 53 patients (F: M=1:2.18). The median age at presentation was 12 days. Forty (74.47%) patients presented with swelling lower back. The patients were Altman I - 23/53 (43.39%), Altman II - 17/53 (32.08%), Altman III - 5/53 (9.43%), and Altman IV - 8/53 (15.09%). Associated major malformations were present in 9 (16.98%) patients; anorectal malformation- 4 (7.55%) patients; 3 (5.66%) were consistent with Currarino syndrome. Tumour excision and coccygectomy were performed in 52 (98.11%) patients. Histopathology revealed mature teratoma in 41/52 (78.85%), immature teratoma in 9/52 (17.31%), and malignant tumours in 2/52 (3.85%) patients. Early postoperative complications were seen in 11/52 (21.15%) patients, including 3 (5.77%) deaths. Early survival (within 3 months) was 48/53 (90.57%). Late complications (within 1 year) were seen in 8/35 (22.86%) patients.

Conclusion: Most of the sacrococcygeal teratomas are benign (mature); immature and especially malignant tumours are often seen around and beyond infancy. Mortality is due to associated major malformations, sepsis, sclerema, and tumour recurrence.

Keywords: *associated malformations, complications, Currarino syndrome, sacrococcygeal teratoma.*

1. INTRODUCTION

The sacrococcygeal teratomas (SCT) are rare embryonal tumour.[1,2] It has an incidence of one in every 35,000–40,000 live births with a female-to-male ratio of 2:1 to 4:1.[1,3-5] SCT is also the most common congenital neoplasm in neonates. SCT may be associated with other malformations which may have a bearing on the survival and final outcomes of these patients.[5,6] In addition to the malignant transformation and associated malformations, postsurgical complications like large scars, and bowel and bladder continence affect the long-term outcomes of these patients.[8] In a gentle attempt to evaluate the factors which are associated with the morbidity and mortality of these patients in our geographical area, the present study was contemplated. Also, there is a paucity of literature on the factors which are associated with outcomes in SCT, from the industrialized nations.[4,5] The study aimed to prospectively evaluate varied clinical presentation, management, and outcome analyses of patients with sacrococcygeal teratoma in our geographical region.

2. METHODS

A prospective descriptive observational study was carried out on a cohort of paediatric patients with SCT. All consecutive patients admitted and managed by the authors in a tertiary care teaching institute from 2011 to 2024 were studied. The ethical approval of the study was taken with IEC No. 394MC/EC/2022, dated-16/05/2022.

There were a few previously published reports from the authors that have been included in the study.[9-13] A detailed clinical examination and radiological evaluation (ultrasonography with or without computed tomography [CT] or Magnetic Resonance Imaging [MRI]) was performed. The data collected included antenatal diagnosis, clinical presentation, physical extent of mass, levels of alpha-fetoprotein, surgical approach, histopathology, recurrence, and outcomes. The patients were sub-classified into four subtypes as per Altman's classification (I to IV), based on its external component and intra-pelvic / intra-abdominal extension of the tumour.[14]

The tumours were also classified into three groups/categories (A to C) based on Benachi et al. for prognostication.[15] Group A: tumour smaller than 10 cm or avascular tumours moderately vascularized with gradual growth; Group B: tumour greater than 10 cm with rapid growth and marked vascularity linked to heart failure and Group C: tumour ≥ 10 cm with gradual growth, cystic predominance, and moderately vascularization.[15] Additionally, we have divided group A into A1 (≤5 cm) and A2 (>5cm) depending up on its size.

The management practice included upfront resection for tumours with the absence of signs of local or distant spread and the presence of age-appropriate AFP levels. Preoperatively, good vascular access was obtained in all patients. An adequate supply of blood products were made available. Steps were taken to prevent hypothermia, especially in neonates by cotton wrapping and high operating room temperature, Lap pads over the tumour and blood warmers.[16] Surgical excision was attempted by sacral/perineal approach (in the prone jack-knife or frog position) presenting with type I and II by chevron incision or posterior sagittal incision. The abdominoperineal approach was reserved for type III or type IV tumours.

Histological grading was done as per Gonzalez-Crucci classification:[5,17] Adjuvant chemotherapy (total four cycles PEB regimen: with Cisplatin, Etoposide and Bleomycin) was initiated 2 weeks postoperatively following the histopathology report.

The follow-up was performed monthly for 3 months, 3 monthly for 1 year and yearly thereafter. It consisted of a complete clinical examination; functional outcomes included bowel and bladder continence, and cosmetic outcomes. Laboratory evaluation (AFP levels) and ultrasonography abdomen were performed for detection of tumour recurrence. Statistical analysis was done using SPSS Statistics for Windows, version x.0 (Chicago, Ill., USA).

3. RESULTS

There were 53 paediatric patients with SCT; 36 girls and 17 boys [Table 1]. The median age at presentation was 12 days (range 1 day–11 years, mean 412 days). Thirty-one patients presented in the neonatal period (Figure 1) with a median 2 days (mean 4.7 days, range 1–30 days).

Table 1: Demographic details and investigations undertaken in our study

Demographic Data	**Frequency n = 53**	**Percentage (%)**
Sex distribution (M: F Ratio = 1:2.18)		
• ***Female***	36	67.92%
• ***Male***	17	32.08%
Age Distribution		

• ***≤ 1 month***	31	58.49%
• ***> 1 months ≤1 year***	7	13.21%
• ***>1 year***	15	28.30%
Radiological investigation		
• ***Ultrasonography abdomen***	53	100%
• ***CT abdomen***	20	37.74%
• ***MRI abdomen***	8	15.09%

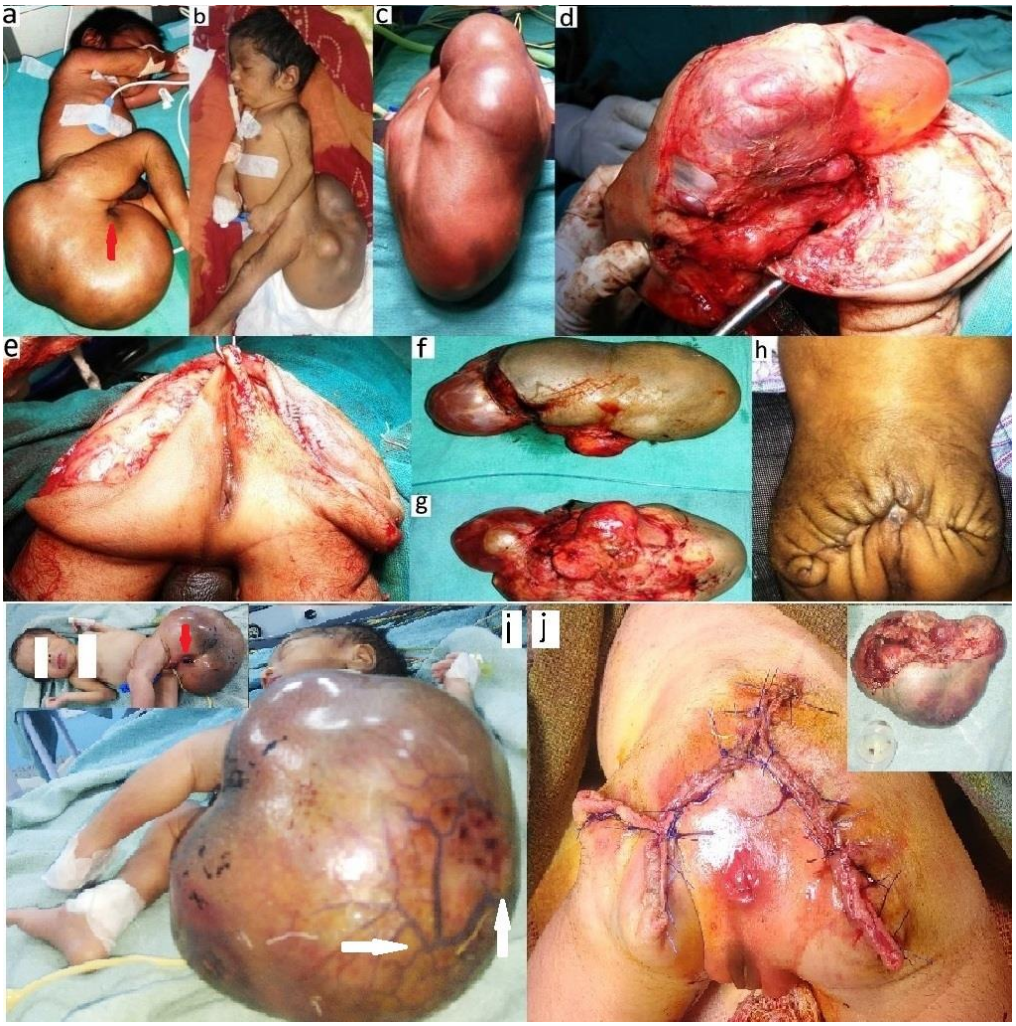

Figure 1: *Clinical photographs* show (a, b and c) a male neonate with a giant sacrococcygeal teratoma (SCT); anus is pushed anteriorly (red arrow). (d) Intra-operative photograph shows dissected SCT typically Altman type I and Benachi type C arising from the coccyx; (e) wound closure in progress after excision of the mass, and (f and g) excised mass with coccyx. (h) Ugly scar following tumour excision. *Clinical photographs* show (i with inset image) a 2 day old neonate with a large, highly vascular sacrococcygeal teratoma (SCT) 150 x 130 x 120 mm in size (giant SCT), typically Altman type I and Benachi type B; anus is pushed anteriorly (red arrow) and superficial veins are prominent (white arrows). (j with inset image) Intra-operative photographs show wound closure after excision of the mass and the anus is placed slightly posteriorly; excised mass with coccyx in the specimen container.

Seven children presented within the 1st year of life beyond the neonatal period (median 8 months, mean 199 days, range 45–330 days), and 15 children (Figure 2) presented at or beyond the age of 1 year (mean 3 years and 8 months, median 26 months, range 1 year–11 years).

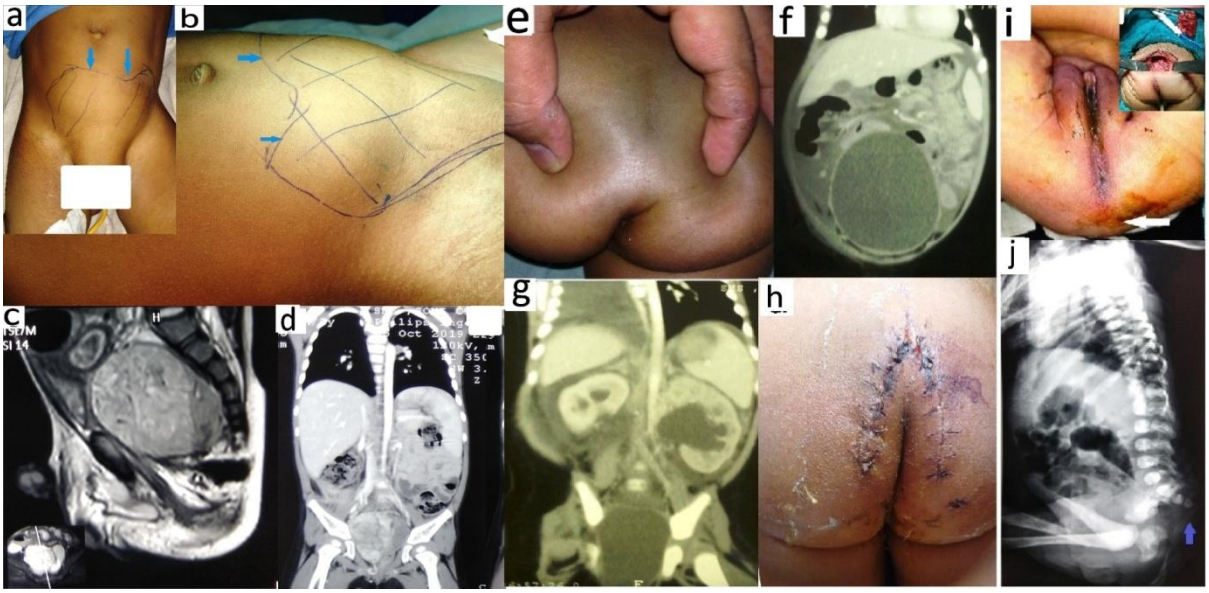

Figure 2: *Clinical photographs* show (a and b) a 2 year old female child with a large pelvic mass extending in the suprapubic region (blue arrow) having Altman type IV and Benachi type B SCT. *Radiological Images* of a 2-year old male child with Altman type IV sacrococcygeal teratoma: (c) Magnetic Resonance Imaging (MRI) sagittal image show a large pelvic tumour arising from the sacrococcygeal region. (d) Contrast Enhanced Computed Tomography (CECT) coronal section images reveal a heterogenous density tumour limited to the pelvic cavity. *Clinical photograph* shows (e) a female infant with sacral swelling. Contrast Enhanced Computed Tomography (CECT) abdomen and pelvic images (f and g) show a large pelvic cyst arising from the sacrococcygeal region with Altman type III and Benachi type A causing bilateral hydrouretronephrosis. (h) Postoperative scar of chevron incision. *Clinical photographs* show (i with inset image) a 2 day old female neonate with sacrococcygeal teratoma (white arrow) and absent anal opening; tumour bed following excision of tumour along with coccyx. Lateral radiographs show absent lower sacral vertebra (indigo arrow) with posteriorly positioned sacrum due to presence of tumour.

History of antenatal diagnosis was available in 10/53 (18.87%). Patients with the antenatal diagnosis presented early in the neonatal age, except one (1.89%) patient. Mode of delivery was vaginal in 30 (56.60%) patients, caesarean section was done in 10 (18.87%) and data unavailable in 3 patients (5.66%).

Forty (74.47%) patients presented with chief complaints of swelling lower back (sacral mass) as shown in Figure 3.

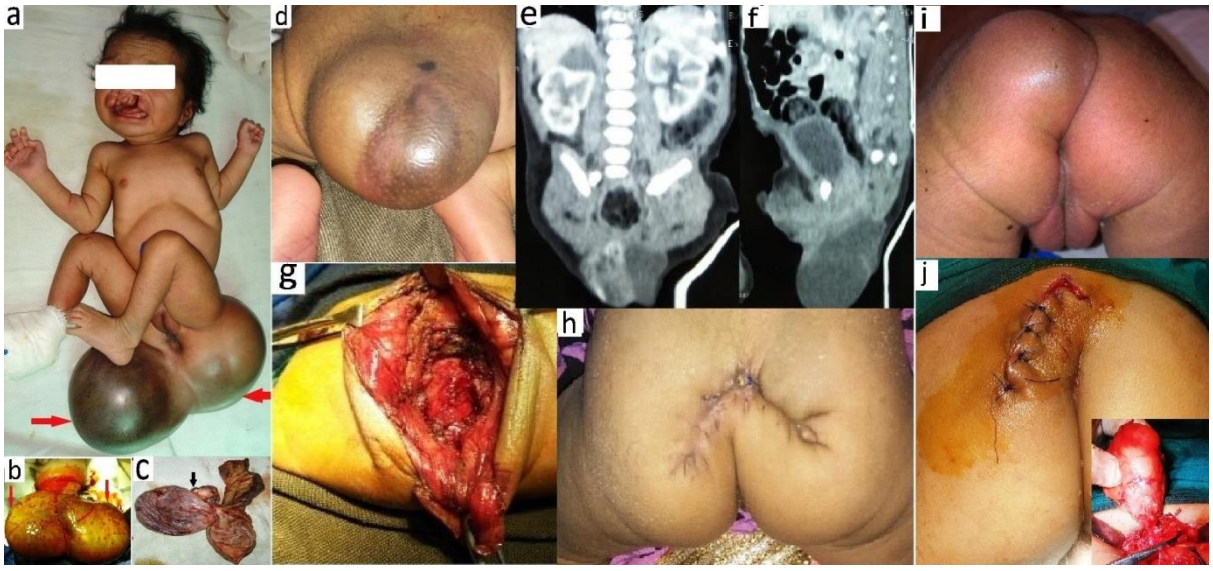

Figure 3: *Clinical photographs* show (a and b) a female neonate with cleft lip and palate having a giant sacrococcygeal teratoma (SCT) typically Altman type I and Benachi type C involving both the buttocks (red arrow); anus is pushed anteriorly.

(c) Excised tumour along with coccyx (black arrow). *Clinical photograph* show (d) an infant with Altman type I and Benachi type A2 sacrococcygeal teratoma; (e and f) Contrast Enhanced Computed Tomography (CECT) coronal section images reveal a heterogenous density tumour with almost whole of it situated externally and arising from the coccyx. (g) Intra-operative photographs show tumour bed following excision of SCT along with coccyx; (h) postoperative scar of chevron incision. *Clinical photographs* show (i) a female neonate with sacral swelling (situated more on left side); Intra-operative photographs show (j with inset image) skin closure of truncated chevron incision and dissected sacrococcygeal teratoma Altman type I and Benachi type A1 arising from the coccyx.

In 12/53 (22.64%) patients, more than one complaint was present [Figure 2]. Varied clinical presentation included: lower urinary tract symptoms (urinary dribbling, dysuria, frequency and hesitancy) in 5 (9.43%), absent anal opening in 4 (7.55%; Figure 2), pain abdomen in 3 (5.66%), suprapubic large mass in 3 (5.66%; Figure 2), swelling anal region in 2 (3.77%), perianal pain and discharge in 1 (1.89%), retention of urine in 1 (1.89%), bilateral hip swelling in 1 (1.89%), gluteal swelling in 1 (1.89%; Figure 3), swelling left lumbar region in 1 (1.89%), feeling of mass coming out from rectum in 1 (1.89%), frothing from the mouth in 1 (1.89%), difficulty in walking in 1 (1.89%), and loss of appetite in 1 (1.89%). Six (11.32%) patients had severe constipation at the time of presentation.

Out of the 7 (13.21%) patients with a palpable tumour per abdomen, 5 (9.43%) patients were Altman IV (Figure 2) and only 2 (3.77%) were Altman type III (Figure 2); tenderness was present in one (1.89%) patient.The concomitant mass palpable per abdomen with sacral mass was present in 2 (3.77%) patients with Altman III tumour (Figure 2).One (1.89%) had suprapubic mass with an associated vague lump in the left gluteal region and anus pushed anteriorly as a slit-like opening.

Ultrasonography was performed in all patients, an additional Computed Tomography (CT) abdomen in 20 patients and an Magnetic Resonance Imaging (MRI; Figure 2) in 8 [Table 1]. No intra-abdominal secondaries were detected in any of the cases at the initial presentation.

Table 2: Distribution of sacrococcygeal teratoma patients into Altman's and Benachi classification

Benachi type	**Altman's type**	**Frequency N=53**	**Percentage**
Type A1	Type I	10	55.56%
N=18	Type II	7	38.89%
(33.96%)	Type III	0	-
	Type IV	1	5.56%
Type A2	Type I	8	30.77%
N= 26	Type II	10	38.46%
(49.06%)	Type III	3	11.54%
	Type IV	5	19.23%
Type B	Type I	1	33.34%
N= 3	Type II	0	-
(5.66%)	Type III	1	33.34%
	Type IV	1	33.34%
Type C	Type I	4	66.67%
N= 6	Type II	0	-
(11.32%)	Type III	1	16.67%
	Type IV	1	16.67%
Total	Type I	23	43.39%
N=53	Type II	17	32.08%
	Type III	5	9.43%
	Type IV	8	15.09%

The distribution of patients as per Altman's classification is enumerated in Table 2. The classical shape of the SCT was present in 48 (90.57%) patients, while dumbbell-shaped tumour (Altman's type II) was appreciated in 3 (5.66%) patients; two (3.77%) were pedunculated.

On radiological evaluation, 46 (86.8%) patients were mixed (solid-cystic) type, while 7 (13.21%) were cystic type. Calcification inside the tumour was seen in 6 (11.32%) patients. SCTs were sub-classified into types A to C as per Benachi et al. [Table 2] with most patients in group A – 44 (83.02%; Figure 3). Out of 53, there were 3 (5.66%) highly vascular tumours (group B; Figure 1), while three (5.66%) had little vascularity. In group C there were 6 (11.32%) patients (Figure 3). Among group A, A1 constituted 18 (33.96%), and A2 – 26 (49.06%) patients.

The patients were divided into 4 groups depending upon the percentage of tumor weight/weight of the patient [Table 5]. Percentage <10% were present in 48 (90.57%) patients, ≥10% to <20%- 1 (1.89%, immature teratoma), ≥20% to <40%- 3 (immature teratoma -1 [1.89%] and mature teratoma – 2 [3.77%]), ≥40%- 1 (1.89%, immature teratoma).

Table 3: Clinical presentation, management and outcomes of patients with associated major malformations and atypical presentation in our series

Clinical details	**Case 1** **Gupta et al[9]**	**Case 2** **Gupta et al[9]**	**Case 3** **Gupta et al[12]**	**Case 4** **Gupta et al[10]**	**Case 5** **Gupta et al[11]**	**Case 6** **Gupta et al[13]**	**Case 7**	**Case 8**	**Case 9**
Age	2 days	3 days	2 days	2 days	26 days	3 year	2 days	2 days	1 day
Sex	Female	Female	Female	Female	Male	Female	Female	Female	Female
Weight (grams)	2700	2500	1940	2600	2400	12000	3000	2700	3800
Familial history	None	None	None	None	None	None	None	None	None
Chief complaints and examination findings	Sacral swelling and absent anal opening	Sacral swelling and absent anal opening	Sacral swelling and absent anal opening	Sacral swelling and absent anal opening	Sacral swelling and swelling in the left lumbar region	Anal discharge, Itching, vague perineal pain	Bilateral hip swelling	Giant sized Sacral swelling	Sacral swelling and Suprapubic lump
Altman's classification	Type II	Type II	Type II	Type I	Type I	Type IV	Type IV	Type I	Type III
Benachi Classification	A (A1)	A (A1)	A (A1)	A (A2)	A (A1)	A (A1)	A (A2)	B	C
Size of tumour (External component)	1.5 × 1.2 cm	2 x 2 cm	5 x 4.5 cm	6 X 6 cm	4 x 3.5 cm	Nil	Nil	15 x 13 x 12 cm	<1cm
Size of tumour (Internal component)	2.6 × 2.1 × 3.8 cm	2 x 2 cm	4 x 4 cm	Tumour attached to coccyx	Tumour attached to coccyx	3 x 2.5 x 3 cm	7.7 x 5.9 x 6.7 cm	Tumour attached to coccyx	8.8 x 5 x 11.7 cm
Consistency of Sacrococcygeal teratoma on Ultrasound	Solid-cystic mass	Solid-cystic mass	Multi-Septate cystic mass	Solid-cystic mass	Solid-cystic mass	Not identified Intraoperative finding	Solid-cystic mass	Solid-cystic mass	Solid-cystic mass

ARM	Vestibular fistula	Cloacal Malformation	Vestibular fistula	Vestibular fistula	No	No	No	No	No
Vertebral defects	Partial sacral agenesis (sacral defect with absent lower sacral vertebrae)	Partial sacral agenesis (sacral defect with absent lower sacral vertebrae)	Partial sacral agenesis (absent lower S4, S5 sacral vertebrae) Multiple thoracic vertebral anomalies	No	Hemivertebra and partial fusion of lumbar spine	No	No	No	No
Other associated malformations / conditions	-	-	Fracture shaft of femur	EA with TEF (Gross type C), Multicystic Dysplastic Kidney and increased Cardio-thoracic ratio	Right renal agenesis, spinal dysraphism of lower cervical region Lumbar hernia, Congenital Talipes Equinus Varus (CTEV), Natal teeth	Anal duplication	Cleft lip and Cleft palate	Sclerema Bilateral Hydroureteronephrosis SFU Grade 3	Syndromic face Bilateral Hydroureteronephrosis SFU Grade 3, uterus displaced anteriorly, ureteral compression
Rectum and bowel	Rectum anteriorly displaced	Rectum anteriorly displaced	Rectum anteriorly displaced	NAD	NAD	NAD	Rectum and bowel anteriorly displaced	Rectum anteriorly displaced	Rectum and bowel anteriorly displaced
CT findings	Presacral mass: 2.6 x 2.1 x 3.8 cm and external component: 1.5 x 1.2 cm	Not done	Hypodense multi-septated sacrococcygeal mass with pelvic component	Not done	Heterogeneously enhancing solid-cystic, with soft-tissue density mass arising from the coccyx	Not done	Not done	Not done	Large heterogeneous density lesion pelvis and presacral region up to umbilical region

Approach	Perineal approach, chevron incision	Perineal approach, Chevron incision	Perineal approach, Chevron incision	Not done	Perineal approach, Chevron incision	Perineal approach, posterior sagittal incision	Abdomino-perineal approach, Pfannenstiel incision and post sagittal midline	Perineal approach, Chevron incision	Abdomino-perineal approach, Pfannenstiel incision and Chevron incision
Procedure for SCT	Tumor excision with removal of Coccyx PSARP	Diverting colostomy Tumour excision with removal of Coccyx	Tumour excision with removal of Coccyx	-	Tumour excision with removal of Coccyx	Excision of duplication tract, tumour excision with removal of Coccyx	Tumour excision with removal of Coccyx	Tumour excision with removal of Coccyx	Tumour excision with removal of Coccyx
Histopathology of specimen	Mature teratoma	Mature teratoma	Mature teratoma	-	Immature grade I teratoma	Mature teratoma	Mature teratoma	Mature teratoma	Mature teratoma
Outcome	Unfavourable (sepsis)	Favourable	Favourable	Unfavourable (sepsis→ Thoracotomy for EA with TEF)	Favourable	Favourable	Favourable	Unfavourable (sepsis)	Favourable

Associated major malformations (anomalies) with atypical presentation were present in 9 (16.98%) patients [Table 3]. Anorectal malformation (ARM) was present in 4 (7.55%) patients, out of which three 3 (5.66%) was also having sacral defect (partial sacral agenesis) as shown in Figure 2. One patient had cleft lip and palate [Figure 3]. Presence of isolated hydrouretronephrosis (due to compression by the tumour) with SCT was not included as malformation.

Serum AFP levels could be evaluated in 19 (35.85%) patients. The values were elevated (inappropriately raised for age) in 6 of 19 (31.58%) patients (range 350–79,860 ng/ml). On presentation, laboratory evaluation revealed anaemia – 6 (11.32%), polymorph leucocytosis – 5 (9.43%), thrombocytopenia (low platelet 88,000 mm^3) – 1 (1.89%), deranged coagulation profile (INR) – 2 (3.77%), and deranged renal functions – 2 (3.77%) patients. Among the neonates, physiological jaundice was present in 16/31 (51.61%).

Neoadjuvant chemotherapy (2 cycles of PEB) was administered to 2 patients. Excision with coccygectomy was performed in 52 patients [Table 4]. In the former (sacral approach), only one (1.92%) Altman type IV with anal duplication (diagnosis suspected intraoperatively) was managed classically with posterior sagittal incision. The tumour was excised along with anal duplication and coccygectomy. Thirty-six patients (69.23%, Altman 1) were operated by posterior sacral approach with chevron (inverted V-shaped) incision) as shown in [Figure 3]. In one (1.92%, Altman 1) SCT with left gluteal swelling, a modified (truncated) chevron incision was given [Figure 3; Table 4]. In 3 patients chevron incision was small and skin flaps were created; wound closure could be performed in the midline [Figure 4 and Figure 5].

Table 4: Management, complications and outcomes of patients in our study

Management details	**Frequency n = 52**
Posterior sacral (perineal) approach • *Chevron (inverted V-shaped) incision)* • *Posterior sagittal incision* • *Modified (truncated) chevron incision*	38 (73.08%) • 36 (69.23%) • 1 (1.92%) • 1 (1.92%)
Combined abdominosacral approach • *Pfannenstiel and posterior sagittal incision* • *Pfannenstiel and chevron incision* • *Abdominal approach with mini posterior sagittal incision*	14 (26.92%) • 6 (11.54%) • 5 (9.62%) • 3 (5.77%)
Complications	**Frequency n = 52**
Intraoperative Complications • *Rectal rent managed by repair and diverting sigmoid colostomy* • *Intrapelvic tumour rupture managed expeditiously by suctioning and irrigation.*	2 (3.84%) • 1 (1.92%) • 1 (1.92%)
Early postoperative complications (within one month) • *Wound infection* • *Prolonged serosanguinous drain output* • *Complete dehiscence with faecal fistula managed by* diverting colostomy • *Mortality*	11 (21.15%) • 5 (9.62%) • 2 (3.84%) • 1 (1.92%) • 3 (5.77%)
Late Complications (During follow-up)	**Frequency n = 35**
Complications (within one year) • *Ugly scar of secondary healing* • *Faecal incontinence* • *Mortality*	8 (22.86%) • 5 (14.29%) • 1 (.86%) • 2 (5.71%)
Mortality and Aetiology	**7/53 (13.21%)**
Preoperative mortality • *sepsis and sclerema* *Postoperative mortality* • *faecal fistula followed by sepsis* • *sclerema and sepsis* • *unknown cause* *Mortality (within 3 months)* • *malignant recurrence* *Mortality (within 1 year)* • *malignant recurrence*	1/53 (1.89%) • 1/53 (1.89%) 3/52 (5.77%) • 1/52 (1.92%) • 1/52 (1.92%) • 1/52 (1.92%) 1/52 (1.92%) • 1/52 (1.92%) 2/52 (3.85%) • 2/52 (3.85%)

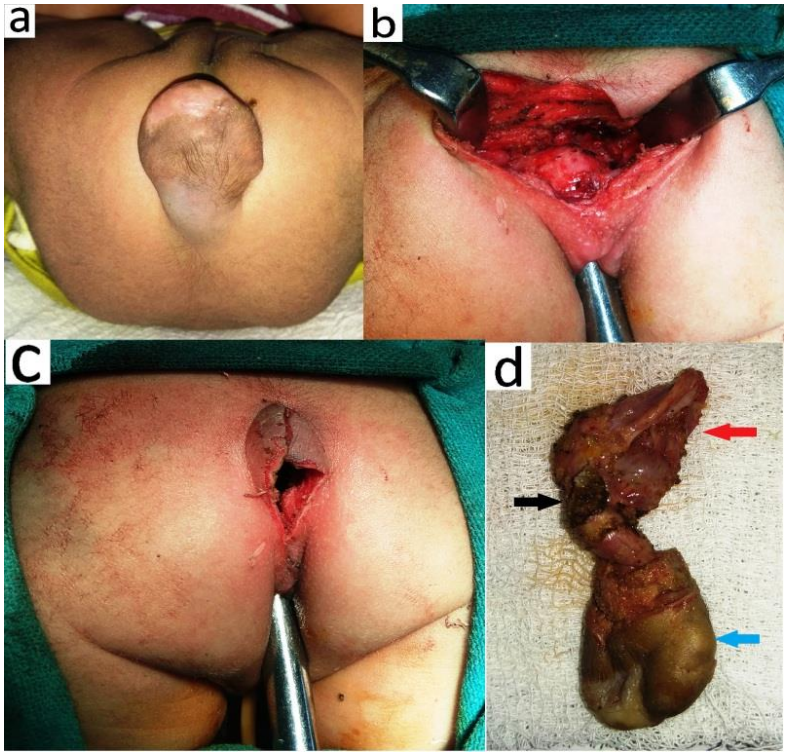

Figure 4: *Clinical photograph* shows (a) a 15 day old female neonate with a small pedunculated sacrococcygeal teratoma (SCT) with Altman type II and Benachi type A. (b and c) Intra-operative photographs show intact rectal wall is clearly seen with anal dilator in-situ following excision of the tumour by using small chevron incision; (c) midline approximation of wound. (d) Excised mass having external (blue arrow) and small internal component (red arrow) with coccyx (black arrow).

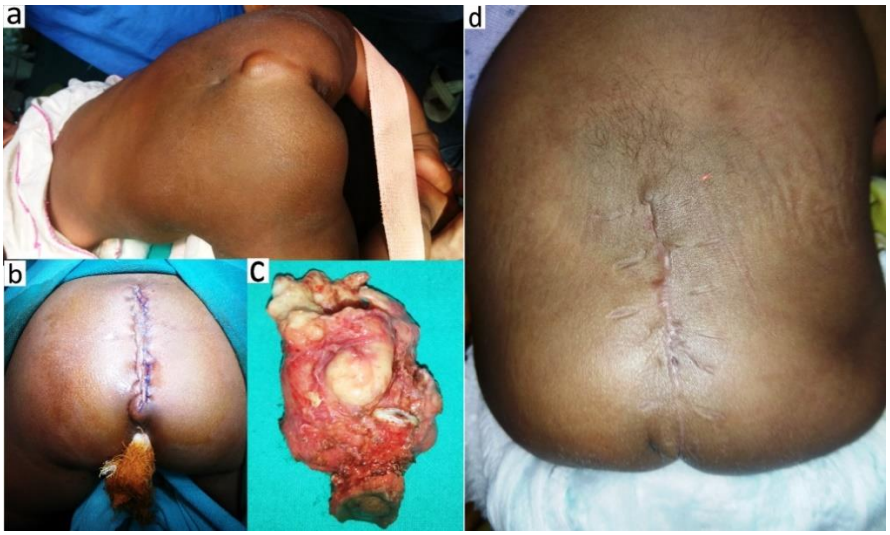

Figure 5: *Clinical photograph* shows (a) a child placed in prone jack knife position with Altman type II sacrococcygeal teratoma. (b) Intra-operative photograph showing wound closure in mid-line following tumour excision using a small chevron incision. (c) Excised mass with coccyx (black arrow) and portion of overlying skin (white arrow) for histopathological evaluation. (d)Postoperative midline scar.

In 14 (26.92%) patients combined abdominosacral approach was used [Table 4]. Out of these, 3 (5.77%) tumours were completely excised by initial abdominal approach (midline incision- 1 (1.92%), infraumbilical transverse incision- 2 (3.84%), but were positioned prone for coccygectomy via the posterior sagittal incision.

The histopathological diagnosis was consistent with mature teratoma in 41/52 (78.85%), immature teratoma in 9/52 (17.31%), and yolk sac tumour (with Schiller duval bodies, and hyaline globules) in 2/52 (3.85%) patients. In 9 patients with immature teratoma: grade 1–3 (5.77%), grade 2–1 (1.92%), and grade 3–2 (3.84%). In 3 (5.77%), patients with immature

teratoma, immature grade was not recorded (two patients were on neoadjuvant chemotherapy). Yolk sac tumour was present in two females aged 16 months (Altman's type III and Benachi group B) and 2 years (Altman's type II and Benachi group A1).

Four (7.69%) patients with immature grade 1 (all 3 neonates) and grade 2 (11-month female) teratomas were simply followed without chemotherapy. Two patients with grade 3 (3-year-old female with Altman's type IV and female neonate with Altman type II) were subjected to adjuvant chemotherapy. Higher grades of immaturity and (malignancy) yolk sac tumour histopathology were seen beyond infancy except in one out of 6 (11.54%) patients.

Additional procedures were performed in 4 (7.69%; 4/52) patients along with tumour excision and coccygectomy. The procedures included excision of the duplicated anal tract (1), diverting sigmoid colostomy with the repair of rectal rent (1), PSARP with left transverse diverting colostomy in a patient with Currarino syndrome (1), and diverting sigmoid colostomy as tumour was adherent to sigmoid mesocolon and colon.

The complications are described in Table 4. Early postoperative complications (within one month) were seen in 11/52 (21.15%) patients; postoperative mortality in three patients were present with associated major malformations [Table 3]. Survival was 48 of 53 (90.57%) patients within 3 months duration. A patient with immature grade 2 teratoma (an eleven-year-old female with Altman type III tumour) had recurrence within 6 weeks and succumbed at 3 months following surgical intervention.

Only 35 patients could be followed up for one year after surgical intervention. Late complications (within one year) were seen in 8/35 (22.86%) patients [Table 4]. In one older patient, faecal incontinence with presence of soiling and inability to hold back defecation was managed with laxatives and bowel management programme. None of the patients had urinary dribbling or motor abnormalities.

4. DISCUSSION

SCT is the most common congenital tumour in the neonatal period and comprises 70% of all teratomas in children.[5,14,17] SCT arise due to the failure of migration of multipotent epiblast cells from the Hensen's (primitive) node of primitive streak. This may result in persistence in the sacrococcygeal region as a teratoma.[9,17] Its association with ARM was earlier hypothesized to be due to obstruction by tumour preventing descent of the urorectal septum to the cloacal membrane, resulting in high-type ARM.[18] The theory of defective mesenchymal precursor cells (proliferation, migration, and differentiation) could explain the presence of associated malformations including low-type ARM with SCT.[9,19]

The number of cases in our study and gender representation were similar to earlier reports in the literature.[4,7,20] SCT has diverse clinical presentations depending on the age, location and pressure effects of the tumour.[4-6] Sacral exophytic mass was the clinical presentation in most series including the present study.[4,7,20] One case in our series with anal duplication had an atypical presentation with anal discharge, along with itching and vague perineal pain.[5] The reasons for late presentation beyond 1 year in 28.30% of patients on diagnosis, and treatment were due to conglomeration of factors including poor socioeconomic factors, geographical barriers, local myths and illiteracy.

The distribution of tumour as per Altman's classification is roughly parallel to the previous literature with the exception of type IV > type III (present study).[14] Approximately 40% of the tumours were solid, 40% were mixed and only 20% were cystic (13.21% in our study).[21] A size of tumour ≥ 10 cm (largest dimension) should be considered a giant SCT. In our series, Benachi group B and group C together (giant SCT) constituted 16.98% (9) patients [Table 5]. Also, if the percentage of tumor weight/ weight of the patient ≥20% is considered as a giant SCT. Then, as per this criterion, at least 7.69% (4) patients could be considered giant SCT [Table 5].

Table 5: Giant sacrococcygeal teratoma in our series as per size and percentage of tumour weight/weight of the patient

Giant SCT	Benachi type	Frequency (Percentage)		Altman	Frequency (Percentage)
Size of tumour ≥10 cm (N=9)	• Type B • Type C	• 3 (5.66%) • 6 (11.32%)		• Type I • Type II • Type III • Type IV	• 5 (9.43%) • 0 • 2 (3.77%) • 2 (3.77%)

Percentage of tumour weight/weight of the patient ≥20% to <40% (N=3)	• Type C	• 3 (5.66%)		• Type I	• 3 (5.66%)
Percentage of tumour weight/weight of the patient ≥40% (N=1)	• Type B	• 1 (1.89%)		• Type I	• 1 (1.89%)

The presence of calcification in the cyst may be present in one-third to 50% of all teratomas.[22,23] The yield was low (11.32%) in our study. Calcification is more frequent in benign than malignant teratomas.

Ultrasonography (USG) is a reliable diagnostic modality during the antenatal period and for the postnatal diagnosis, surgical planning and monitoring of disease.[24-26] A CT scan is required for surgical management.[14] Antenatal diagnosis in our study (18.87%) was comparable to a previous study (7/41; 17.07%) from India.[4] One (1.89%) patient was diagnosed with type III SCT with bilateral hydroureteronephrosis on antenatal evaluation in the early third trimester, while 9 (16.98%) were diagnosed in the late trimester or before the delivery was planned.

SCT may be associated with vertebral (80%), anorectal, and urogenital anomalies (12%).[27,28] The incidence of associated major malformations in our study (16.98%) was similar to other reports in the recent literature (15% to 25%).[5,21] Anorectal malformation (ARM) was the most common association in our series which is in coherence with the literature.[18,29] The incidence of vertebral abnormalities in our study was less (4; 7.55%) as compared to one of the previous study.[30]

The syndromes associated with SCT are Currarino syndrome, Sotos syndrome, Weaver syndrome and Beckwith-Wiedemann syndrome.[31] The chromosomal abnormalities associated with SCT are 1, 3, 10, 12 and 13 chromosomes.[31] Currarino syndrome is a malformation complex and has a triad of presacral mass, sacral defect (partial sacral agenesis) and anorectal malformation. The presacral mass could be teratoma in up to 40% patients.[32-34] ARM is usually of the low type (anorectal stenosis), but high types have been reported i.e. vestibular fistula and persistent cloaca (as in our study) and recto-urethral fistula.[34,35] The presacral teratoma i.e. Altman's type IV SCT is classical of Currarino syndrome. In our series, all 3 (5.66%) patients had Altman's-type II SCT (a variant of Currarino syndrome).[10] About half of these specific neurenteric malformation cases are familial with autosomal dominant inheritance. HLXB9 gene (causative gene in 30% of cases of Currarino syndrome) is telomeric to the sonic hedgehog (SHH) on 7q36; loss of SHH may be contributory to its associated malformations.[9]

Complete surgical resection with coccygectomy within the first few days of life (ideally within two months) remains the primary management.[24,36] The delayed surgical intervention beyond 2 months of age is associated with a higher rate (48%-67%) of malignancy and recurrence.[4,14,24,36] Sacral route with Chevron incision (inverted V-shaped) was used for the majority of tumours as it provides an excellent exposure to SCT Altman's type I and type II.[37] Its drawbacks were ugly scars and some degree of posterior displacement of the anus (as seen in one of our cases; Figure 1).[36,37]

The posterior sagittal incision is associated with better cosmetic outcomes as it uses the median-raphe and results in lesser disfigurement. It has been recommended for smaller-sized lesions and those located more in the midline.[38] The combined abdominosacral approach is used for (i) type III tumours extending beyond the sacral promontory or tumors palpable per-abdominally and type IV tumours.

Grading of SCT does not draw a parallel with prognosis, as against ovarian teratoma where grading has direct correlation with prognosis.[7] Percentage of mature teratoma in similar studies has been reported to be between 53.4% to 80% (78.85% in our series), while immature teratoma is between 12% to 30.8% (17.31% in present study).[4,5,7,17,20,39] Malignant tumours range from 12% to 19.5% of cases (3.85% in present study).[4,5,7,17,20,39]

On histopathology, most of the neonatal SCT's are invariably mature or immature type and less than 10% of the cases are malignant in the neonatal period.[4] Out of 31 neonates in the present study, histopathology was available in 30; 25 had mature teratomas, while immature teratoma were present in 5. Immature grade 1 (3), grade 3 (1) and grade not mentioned (1). Malignant tumours reported are yolk sac tumour (as in our series), choriocarcinoma or embryonal carcinoma.[1,38-40]

Out of 15 children ≥1year in our study: mature teratoma (10), immature teratoma (3) and malignant tumour with yolk sac histopathology (2) were present. The percentage of malignancy in the present series, especially among the children beyond infancy was less as compared to other studies.[4,14]

Recurrence in SCT is attributed to incomplete tumour resection, incomplete en bloc coccygectomy with the tumour, tumour spillage or rupture, and failure to detect intra-tumoral malignant components on histopathological evaluation. In patients with an entirely removed coccyx, recurrence may be as high as 20% of initially benign tumours over the first three years of life, and 40% in the form of malignant tumours.[24] In the present series, recurrence was appreciated in an eleven-year-old female with an Altman type III tumour with initial grade 2 teratoma (histopathology) in the benign subgroup.

We second the observation of Ein et al. that immature teratoma is a grey zone between benign and malignant tumour and the risk of malignant recurrence in immature teratoma group is high.[22]

The wound-related complications (10.42%; 5/48) were invariably due to the location of tumour, proximity to anus, ischemia of the skin flaps and type of the tumour.[4] It was in concurrence with previously published studies.[4] Anorectal dysfunction including faecal incontinence, constipation or postprandial tenesmus on clinico-radiological evaluation may be present in 29% of patients.[32,41] In our series, anorectal dysfunction was appreciated in 6.25% (3/48) cases. Long term evaluation of anorectal functions in adults showed faecal incontinence in 12%, but normal bowel habits in only 27% of patients.[42] Urologic dysfunction (urinary dribbling/ incontinence/ neurogenic bladder) was not present in our study. Urinary incontinence can be prevented by avoiding deep pelvic facial planes.[11]

The prognosis is multifactorial and it depends on the time of diagnosis, the malignancy potential, and the quality of surgery. Neonatal mortality with SCT is around 16% as per the previous literature. It increases if the tumours are associated with complications like haemorrhage (most common cause of mortality), and high output cardiac failure (vascular tumours with marked arteriovenous shunting).[39,43] The presence of coagulopathy due to the release of thromboplastins into the bloodstream after tumour trauma at time of delivery may result in haemorrhage.[47] SCT with associated major malformations have high mortality rate (3/9; 33.33%) in our study. Survival rates are excellent in absence of major associated malformations. The mortality rate for tumours >10 cm is 18%.[24,25,43] It was 11.11% (1/9) in our study with 33.33% (1/3) in Benachi type B tumors.

Malignant transformation is an important cause of mortality with late presentation, especially beyond infancy. The type IV has the highest and type I lowest risk of malignancy. Comprehensive clinico-radiological follow-up along with determination of AFP level (3 monthly) to detect any recurrence within 3-5 years postoperatively is mandatory.[24,40]

5. CONCLUSION

Antenatal screening must be done for all pregnancies for early diagnosis of congenital malformations, especially SCT. Atypical presentation like lower urinary tract symptoms (including retention of urine), abdominal pain, suprapubic lump, anal duplication (with perianal pain and discharge) may be present. Associated major life threatening malformations (16.98%) with SCT are anorectal malformations and esophageal atresia. Benachi classification guides about the vascularity and size of the tumor; Altman classification guides the surgical approach. Histopathological grading of immature tissue in SCT does not correlate directly with prognosis and a thorough search for the malignant component, especially yolk sac tumour is mandatory. The mortality is due to sepsis following faecal fistula, sclerema in highly vascular tumour >10 cm (Benachi group B tumour), associated malformations and malignant recurrence. Major postoperative complications like urinary and bowel incontinence and also motor abnormalities are unusual owing to improve surgical techniques and postoperative management. A comprehensive long-term close follow-up for recurrence of malignant tumours and distant metastasis must be performed with clinico-radiological evaluation and serial AFP monitoring.

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