Sacrococcygeal teratoma: A new grading system

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Learning objectives

- MRI is a superior modality for the preoperative assessment of sacrococcygeal teratoma (SCT) by delineating the size, extension, and consistency of the mass.

- Our proposed grading system includes the following criteria: tumor size (T1, T2, T3); level of deep extension of the tumor (coccygeal, lower sacral, upper sacral, abdominal); tumor consistency (cystic 1, 2, 3, or solid 1, 2).

- Tumors with deep extension (proximal to the level of S3) were associated with marked compression and distortion to surrounding pelvic structures and may require a combined abdomino-perineal approach for excision.

- Pure cystic tumors (C1) were associated with low vascularity, easy dissection, benign pathology, but may cause diagnostic confusion with meningeoceles.

- A special subtype of predominantly cystic tumors (C3) was characterized by irregular internal cyst wall thickening and immature pathology.
Background

Sacroccocygeal teratoma (SCT) is considered the most common tumor in the neonatal period but may also present later during infancy or childhood [1]. Excision of the tumor is usually feasible, and the prognosis is generally good [2]. Most tumors are benign [3]; however, the risk of recurrence and the occasional presence of malignant foci are major concerns [1,4].

Based on the American Academy of Pediatrics surgical section survey (1962 - 1972), Altman et al proposed their famous classification for SCT in 1973 [5]. Despite its wide spread acceptance, the Altman classification has been criticized for being descriptive rather than a practical classification with limited impact on the prognosis [6]. Usui et al have shown 'predominantly' solid tumors to be associated with higher risk of mortality [7]. The tumor size is another important factor that has been used in predicting the prognosis of antenatally diagnosed cases [8,9].

Enormous development has involved the cross-sectional imaging techniques since Altman introduced his classification for SCT in 1973. This might have been expected to share in the evolution of the original classification by disclosing more anatomical and structural details. However, to the best of our knowledge, little contribution has been published in this field. Here, we tried to spot light on maximizing the role of preoperative imaging in guiding pediatric surgeons to the prognosis and challenging situations they may encounter during managing these cases. This was achieved by studying a group of patients of SCT who were managed at our hospital, while correlating their preoperative imaging to the operative and pathological findings.
Findings and procedure details

The medical records of patients with SCT, who were managed at our hospital during the period 2009 through 2016, were retrospectively reviewed. These included demographic data, preoperative investigations, operative details, and pathology reports of excised specimens. Only cases of SCT with available preoperative cross-sectional imaging studies (MRI and/or CT scans) were included in the study. Pre-sacral dermoid cysts associated with ano-rectal and vertebral anomalies as a part of the Currarino triad were not included, as we believe these cases represent another entity of developmental cysts or hamartomas rather than true neoplasms [10].

The preoperative cross-sectional imaging studies of cases of SCT were retrieved (either from saved electronic copies, or more recently from our hospital Picture Archiving and Communication System 'PACS' database) and were re-examined by two of the authors (SAM, AAA). The preoperative imaging features were correlated to the operative and pathological findings.

MRI technique: MRI pelvic examinations were carried out using 1.5 Tesla magnet (Philips, Achieva; The Netherlands). Examinations were performed under sedation or general anesthesia. The following sequences were obtained in axial, coronal, and sagittal planes: T2WI, T1WI, and fat suppressed T2WI.

Operative procedure: Mostly, the sacral (perineal) approach was used to excise the sacrococcygeal masses either via the classic 'chevron' incision [11], or the vertical posterior sagittal approach [12]. Sometimes a combined abdomino-perineal approach was needed to excise tumors with deep pelvic or intra-abdominal extension.

The study included 24 cases of SCT (20 females and 4 males). Their age at presentation ranged from day one to 36 months. Their preoperative cross-sectional imaging included: MRI examinations in 13, CT scans in four, and both imaging modalities were available in seven cases.

Pathological reports of excised specimens were available in 13 cases: benign mature teratoma in eight, immature teratoma in three, and malignant histology was found in two cases.

The following were identified as relevant preoperative imaging features to be included in our proposed grading system: tumor size (T1, T2, T3); level of deep...
extension of the tumor (coccygeal, lower sacral, upper sacral, abdominal); tumor consistency (cystic 1,2,3, or solid 1,2).

First: tumor size

We categorized patients according to the maximum tumor dimensions in two perpendicular planes into three grades: T1, T2, and T3 Fig. 1 on page 12.

Second: deep extension of the tumor

The level of tumor extension in the cranial direction (deep extension) was measured in relation to the near-by vertebral column (number of the corresponding vertebral body) Fig. 2 on page 12. Four main categories could be identified: 1) coccygeal; 2) lower sacral (S 3, 4, 5); 3) upper sacral (5 1, 2); and 4) lumbar (abdominal). As expected, the last

Fig. 1: Grading of SCT according to the size of the tumor. (a and d) Grade T1: less than 5 cm. (b and e) Grade T2: between 5 and 10 cm. (c and f) Grade T3: more than 10 cm.

References: Abouzeid AA et al. (2017) preoperative grading of sacrococcygeal teratoma: A road map to successful resection J Neonatal Surg 6:75-
two categories were more challenging in their management. Many surgeons would agree that pre-sacral tumors extending above the body of S3 may need a combined abdomino-perineal approach during excision [13,14]. Moreover, SCTs with deep extension (upper sacral/lumbar) were found to be associated with marked compression and distortion of the pelvic organs (sometimes causing hydrocolpus and hydrenephrosis Fig. 3 on page 13.

![Fig. 2: Mid sagittal MRI of 4 cases of SCT demonstrating the different grades of deep tumor extension in relation to the vertebral column (an asterisk * is marking for the first sacral vertebra S1). a) Tumor extension opposite to the coccyx in a one-day-old female. b) Tumor extension up to S4 (lower sacral) in a 25-month-old female. c) Tumor extension up to S1 (upper sacral) in a 12-month-old female. d) Tumor extension up to T12-L1 (abdominal) in a 25-day-old female.](image)

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**Third: tumor consistency**

We could identify two main categories: predominantly cystic, and predominantly solid tumors.

- The predominantly cystic tumors were further sub-classified into: a) purely cystic (C1); b) multi-compartmental cyst (C2) displaying variable signal intensities and presence of fatty components Fig. 4 on page 14; and c) the last subtype (C3) with its characteristic irregular internal cyst wall thickening caused by the in-growth of solid tumor component Fig. 5 on page 15. The later subtype (C3) was always associated with immature pathology.
**Fig. 4**: Differentiation between two different subtypes of predominantly cystic SCT by MRI. (a and b) Sagittal T2 and T1 WI of a one-day-old female with purely cystic subtype of SCT (C1); note the homogenous hyperintense signal in T2, hypointense in T1, and thin capsule. (c and d) Sagittal T2 and T1 WI of a 12-month-old female with complex multi-compartmental cyst (C2) displaying variable signal intensities and presence of fatty components.

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**Fig. 5:** Subtype C3 of the predominantly cystic SCT; the cyst cavity is marked by an arrow that is pointing to its characteristic irregular internal wall thickening. a) CT scan of a 10-month-old boy (coccygeal mass). b) MRI axial T1WI of a 2-month-old girl (coccygeal mass). c) MRI coronal T2WI of a 25-day-old female; note the marked intra-abdominal extension of the tumor, with a fungating internal wall thickening (black arrow)

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The purely cystic subtype (C1) was associated with low vascularity, easier dissection, and benign pathology. However, the purely cystic appearance of the tumor and the proximity to the vertebral column provoked some confusion about the diagnosis [Fig. 3 on page 13](#). The typical multi-cystic appearance of the tumor, absence of vertebral bony defects, absence of spinal cord anomalies (tethered cord), and lack of continuity with the dural sac were important differentiating signs from meningeoceles [Fig. 3 on page 13](#).
Fig. 3: Pure cystic type of SCT (C1). a) Three-day-old female presenting with a cystic mass at the lower back misdiagnosed initially for meningeocele. (b,c,and d) MRI study, sequential sagittal T2WI, showing the intra abdominal extension of the mass. The curved black arrow in (b) is pointing to cystic out-pouching through the neural foraminae. The cystic out-pouching appears to be separable from the dural sac (straight black arrow in c). The white arrow in (d) is pointing to normal termination of the cord in the mid-sagittal plane with intact vertebral bodies down to the coccyx. Note the vaginal distension (*) and bladder compression by the mass effect of the cyst.

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- On the other hand, the predominantly solid tumors may be subclassified into S1 or S2 according to the absence or presence of poor prognostic imaging
criteria respectively. By poor prognostic imaging criteria, we mean signs of local tumor invasion Fig. 6 on page 15 and/or distant metastasis Fig. 7 on page 15.

**Fig. 6:** Seven-month-old boy presenting with a gluteal swelling (a). Mid sagittal MRI (b) showing the deep extension of a predominantly solid mass, and the axial section (c) showing local tumor invasion by breaching the capsule (white arrows).

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Fig. 7: CT scan of a 3-year-old girl presenting with metastasizing malignant sacrococcygeal tumor. a) Predominantly solid sacrococcygeal mass. b) Tumor invasion of bony sacrum. c) Abdominal cuts showing liver metastasis. d) CT chest showing pulmonary metastasis.

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Conclusion

Solid SCT are thought to carry poorer prognosis [7,15]. This can prove to be true from two aspects. Large tumors (T3) with significant solid components are associated with high vascularity. In small babies, this may be associated with circulatory compromise, besides the risk of bleeding complications [6]. The other poor prognostic aspect with solid tumors is related to the risk of malignancy [15].

On the other hand, predominantly cystic SCT were not always benign as it might be expected [15]. In the preoperative imaging studies, we could identify a special subtype (C3) with its characteristic irregular internal cyst wall thickening caused by the in-growth of solid tumor component. This subtype was always associated with immature pathology, which would necessitate more caution during dissection to achieve complete excision and avoid spillage.

Finally, MRI represents a superior modality in the preoperative assessment of cases of SCT that can provide a grading system indicating for the prognosis and degree of expected surgical challenge.
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