ORIGINAL ARTICLE

Preoperative Grading of Sacrococcygeal Teratoma: A Roadmap to Successful Resection

Amr Abdelhamid Abouzeid[1],* Shaimaa Abdelsattar Mohammad[2], Mohammed Elsherbeny[1], Nehal A Radwan[3], Osama El-Naggar[1]

1 Department of Pediatric Surgery, Ain-Shams University, Egypt

2 Department of Radiodiagnosis, Ain-Shams University, Egypt

3 Department of Pathology, Ain-Shams University, Egypt

ABSTRACT

Purpose: to include the most relevant preoperative imaging features of sacrococcygeal teratoma (SCT) in a grading system that would provide guidance to surgeons during excision of such rare tumors.

Patients and Methods: The medical records of patients with SCT, who were managed at our hospital during the period 2009 through 2016, were retrospectively reviewed. Only cases of SCT with available preoperative cross-sectional imaging studies (MRI and/or CT scans) were included in the study. The preoperative imaging features were correlated to the operative and pathological findings.

Results: The study included 24 cases of SCT (20 females and 4 males). Their age at presentation ranged from day one to 36 months. The following were identified as relevant preoperative imaging features to be included in our proposed grading system: tumor size, level of deep extension of the tumor, and tumor consistency. Among predominantly cystic SCT, a special subtype C3 could be identified with its characteristic irregular internal cyst wall thickening caused by the in-growth of solid tumor component. These cases were always associated with immature pathology.

Conclusion: MRI is a useful modality in the preoperative assessment of cases of SCT by providing a grading system that can indicate for the prognosis and degree of expected surgical challenge.

Key words: Teratoma; MRI; Prognosis; Grading

INTRODUCTION

Sacrococcygeal teratoma (SCT) is a rare tumor with a reported incidence of 1 per 35,000 - 40,000 live births [1]. However, SCT is considered the most common tumor in the neonatal period with a characteristic female predominance [2,3]. Based on the American Academy of Pediatrics Surgical Section survey (1962-1972), Altman et al. proposed their famous classification for SCT in 1973 [3].

SCT has a tendency to grow enormously reaching a huge size. The tumor mass is usually seen displacing, distorting, but not invading the surrounding pelvic organs. The aim of surgery is to excise the sacrococcygeal mass completely without leaving any residual tumor (to decrease the risk of recurrence) [4,5]. Meanwhile, it is also important to preserve the function and innervations of the surrounding pelvic viscera, especially when excising a tumor that is known to be mostly benign [1,6]. Being a rare tumor, many

pediatric surgical centers treat no more than 2-3 cases of SCT per year [1]. Hence, a proper preoperative imaging is crucial in identifying expected surgical challenges that might be encountered during such unfamiliar operations.

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 [7]. Usui et al. have shown early delivery and 'predominantly' solid tumors to be associated with higher risk of mortality [8]. The tumor size is another important factor that has been used in predicting the prognosis of antenatally diagnosed cases [9,10].

Through studying the preoperative cross-sectional imaging of cases of SCT, we tried to identify relevant imaging features that would have an impact on the surgical plan and prognosis; and collect these features in a grading system to provide guidance for pediatric surgeons during excision of such rare tumors.

MATERIALS AND METHODS

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. Presacral 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 (hamartomas) rather than true neoplasms [11].

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. Owing to the retrospective nature of the study, the scientific committee approved reporting our observations without an IRB number.

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: The sacral (perineal) approach was used to excise the sacrococcygeal masses either via the classic 'chevron' incision [4], or the vertical posterior sagittal approach [12]. In one case, a combined abdomino-perineal approach was needed to excise a tumor with deep intra-abdominal extension.

RESULTS

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. In one case, the ante-natal MRI was also available (Fig.1). 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 (remaining cases with unavailable pathology reports may be assumed to be benign, as immature or malignant cases should have been referred to the oncology unit for adjuvant chemotherapy).
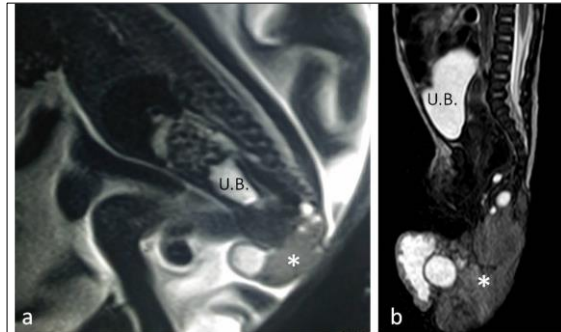

Figure 1: Ante-natal diagnosis of SCT. a) ante-natal MRI (22 weeks gestation) showing a mass (*) arising from the caudal end of the fetal vertebral column (SCT). b) Repeating the MRI postnataly at day 2 before surgical excision. U.B.: urinary bladder

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).

Tumor size: We categorized patients according to the maximum tumor dimensions in two perpendicular planes into three grades: T1, T2, and T3 (Table 1, Fig.2). Larger tumors (T2, and T3) with significant solid components were associated with higher vascularity. This was reflected on the size of its feeding vessels seen at operation (median sacral artery).

Table 1: Distribution of cases of SCT according to tumor size

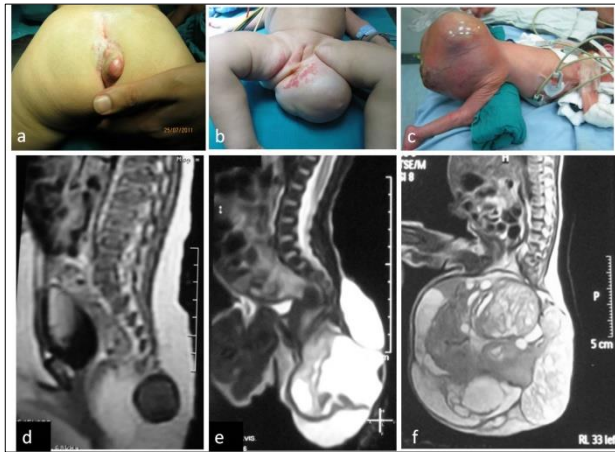| Grade	Dimensions	Number of cases
T1	Less than 5x5cm	8
T2	5 - 10cm	12
T3	More than 10x10cm	4

Figure 2: 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.

Figure 3: 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.

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 (the number of the corresponding vertebral body) (Fig.3). Four main categories could be identified: 1) coccygeal; 2) lower sacral (S 3, 4, 5); 3) upper sacral (S 1, 2); and 4) lumbar (abdominal). As expected, the last 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] Also, the level at which the median sacral artery can be secured by ligation (to guard against bleeding complications during excision of the mass) was found to shift upwards with deeper tumor extension. Exposure of the artery is usually performed from below after separation of the coccyx. However, for tumors associated with intra-abdominal extension, the median sacral artery was

approached from above (opposite the sacral promontory). Moreover, SCTs with deep extension (upper sacral/lumbar) were found to be associated with marked compression and distortion of the pelvic organs (sometimes causing hydrocolpos and hydronephrosis (Fig.4).
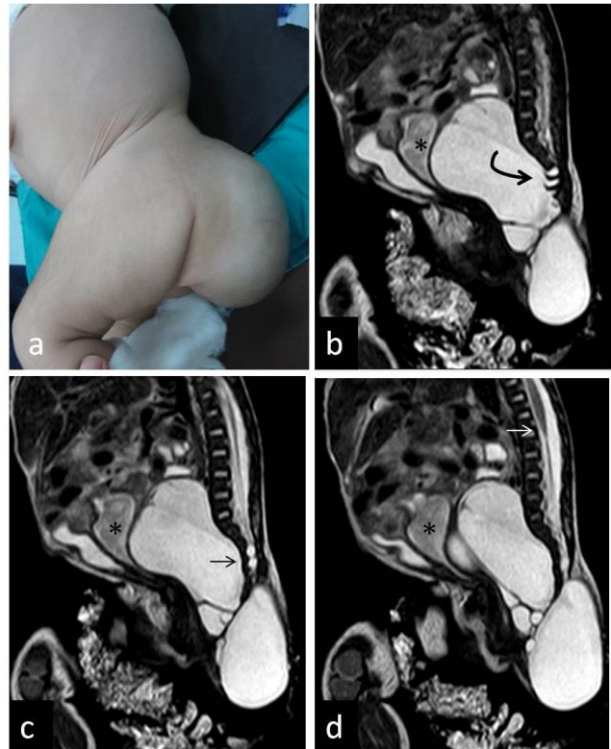

Figure 4: Pure cystic type of SCT (C1). a) Three-day-old female presenting with a cystic mass at the lower back, initially misdiagnosed as meningeocele. (b,c,and d) Sequential sagittal T2WIs, showing the deep extension and the multi-cystic nature of the mass. b) The pre-sacral cystic lesion is sending out-pouching through the neural foraminae (curved black arrow) into the spinal canal. c) Demonstration of the intra-spinal extension of the mass (straight black arrow). d) Mid-sagittal plane demonstrating the normal termination of the spinal cord (white arrow), with intact vertebral bodies down to the coccyx. Note the vaginal distension (*) and bladder compression by the mass effect of the cyst.

Tumor consistency: We could identify two main categories: predominantly cystic, and predominantly solid tumors. These were further sub-classified as shown in Table 2. The purely cystic subtype (C1) was associated with low vascularity, easier dissection, and benign pathology; even large cysts extending into the abdomen could be excised completely from below through the sacral approach (Fig.4; Fig.5a,b). However, the purely cystic appearance of the tumor and the close proximity to the vertebral column provoked some confusion about the diagnosis, especially those showing out-pouching into the neural foraminae (Fig.4). 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.4). Another subtype of the predominantly cystic SCT (C3) could be identified with its characteristic irregular internal cyst wall thickening caused by the in-growth of solid tumor component (Fig.6). We had three cases of subtype C3, all were associated with immature pathology. On the other hand, the predominantly solid tumors per se were not associated with poorer prognosis unless there were signs of tumor invasion (Fig.7) and/or distant metastasis (Fig.8).
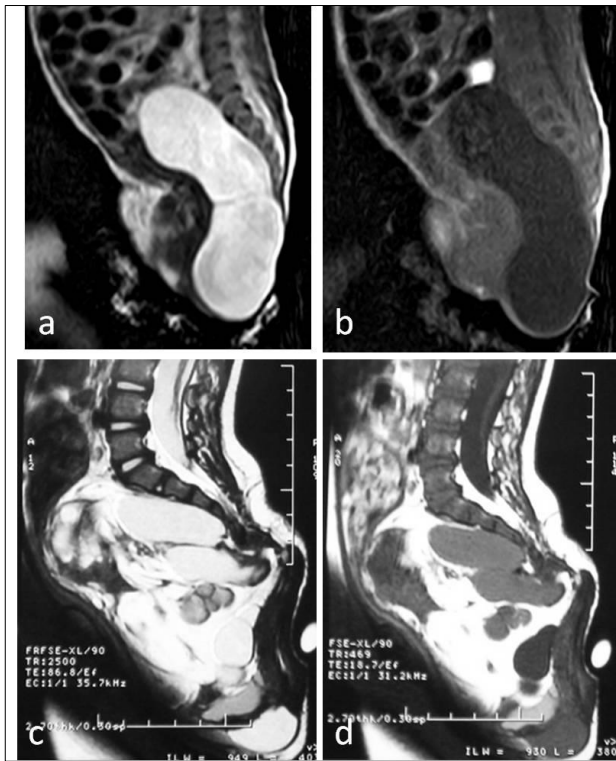

Figure 5: Differentiation between two different subtypes of predominantly cystic SCT by MRI. (a and b) Sagittal T2 and T1 WI of a one-day-old female with purely cystic subtype of SCT (C1); note the homogenous hyperintense signal in T2, hypointense in T1, and thin capsule. (c and d) Sagittal T2 and T1 WI of a 12-month-old female with complex multi-compartmental cyst (C2) displaying variable signal intensities and presence of fatty components.
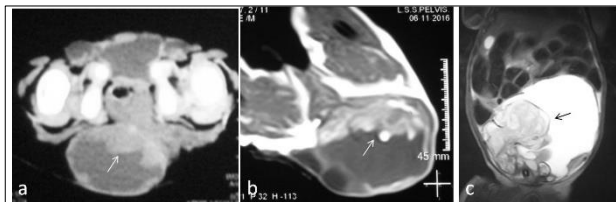

Figure 6: Subtype C3 of the predominantly cystic SCT; the cyst cavity is marked by an arrow that is pointing to its characteristic irregular internal wall thickening . a) CT scan of a 10-month-old boy (coccygeal mass). b) MRI axial T1WI of a 2-month-old girl (coccygeal mass). c) MRI coronal T2WI of a 25-day-old female; note the marked intra-abdominal extension of the tumor, with 'fungating' internal wall thickening (black arrow).

In this study group, the pathology of the tumor whether mature or immature teratoma was not related to certain age group at presentation (both pathologies were detected in the neonatal and older age groups). However, we had two cases with malignant pathology; both presented after the neonatal period (7 months and 3 years). Overall, we had three mortalities: the two cases with malignant pathology, and another mortality in the neonatal period was related to early postoperative complications (sepsis and metabolic derangements).

Figure 7: 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 breeching the capsule (white arrows).
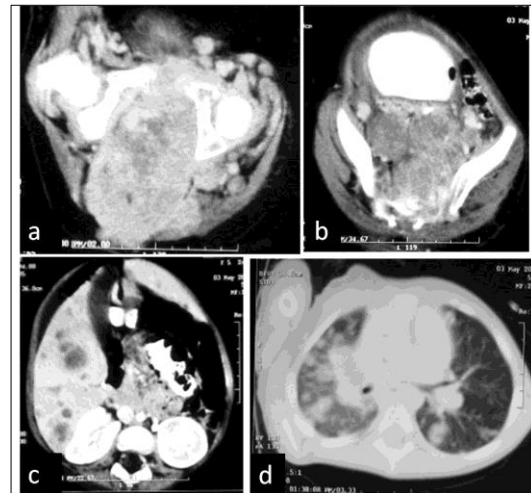

Figure 8: CT scan of a 3-year-old girl presenting with metastasizing malignant sacrococcygeal tumor. a) Predominantly solid sacrococcygeal mass. b) Tumor invasion of bony sacrum. c) Abdominal cuts showing liver metastasis. d) CT chest showing pulmonary metastasis.

DISCUSSION

SCT is considered the most common tumor in the neonatal period, but may also present later during infancy or childhood [2]. Excision of the tumor is usually feasible, and the prognosis is generally good. [6] Most tumors are benign [1]; however, the risk of recurrence and the occasional presence of malignant foci are major concerns. [2, 15] Unsatisfactory cosmesis of the buttock region remains a common complaint after SCT excision. [16] Other delayed sequels include neurogenic bladder and fecal incontinence. The cause of bowel and bladder dysfunction has been suggested to be either iatrogenic (during tumor resection), due to mass effect of the tumor, or both. [6, 17]

Table 2: Grading of cases of sacrococcygeal teratoma according to tumor consistency

Tumor consistency	Grade	Characteristic imaging features	Number of cases	Age	Pathological correlation
Predominantly cystic	C1 (Fig.5a,b)	Purely cystic with homogenous fluid content (hypointense in T1WI and hyperintense in T2WI). Thin cyst walls and septae.	4 cases	1 day-10 months; Mean=2.6 months Median=3 days	Mature cystic teratoma (3 available pathology reports)
	C2 (Fig.5d,c)	Complex multi-compartmental with heterogenous fluid content (variable MR intensity) and adipose tissue (fat). Thicker cyst walls and septae.	10 cases	3 days-25 months Mean=7.4 months Median=4 months	Mature cystic teratoma (4 available pathology reports)
	C3 (Fig.6)	The cyst wall shows irregular internal thickening caused by in-growth of solid tumor component into the cyst cavity.	3 cases	25 days-10 months Mean=4.3 months Median=2 months	All three cases proved to be Immature cystic teratoma
Predominantly solid	S1 (Fig.1), (Fig.2f)	Tumor is displacing and distorting surroundings without invasion.	5 cases	1-30 days Mean=7 days Median=1 day	Mature teratoma (1 available pathology report)
	S2 (Fig.7), (Fig.8)	Signs of tumor invasion through surrounding tissues including breeching of tumor capsule + distant metastasis	2 cases	7-36 months Mean=21.5 months Median=21.5 months	Both were malignant

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. More recently, there has been increasing interest in the ante-natal diagnosis of SCT, and reports on the feasibility of fetal intervention in selected cases [7-9, 18, 19].

Many pediatric surgical centers treat fewer than 2-3 new cases of SCT per year [1], which makes it a relatively unfamiliar operation. 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 with SCT who were managed at our hospital, while correlating their preoperative imaging to the operative and pathological findings.

SCTs have a tendency to expand in two opposite directions either in the cranial, or more commonly in the caudal direction. The latter represents the external (easy accessible) component of the tumor. On the other hand, tumor extension in the cranial direction forms the deep (hidden) part which requires more surgical experience during dissection. The near-by vertebral column is an important anatomical landmark that can be used as a scale to measure the exact degree of tumor extension in relation to the bodies of the corresponding vertebrae. SCTs with deep extension (above the body of S3) may require a combined abdomino-perineal approach for excision [13, 14]; moreover, they have been found to cause

marked compression and distortion to surrounding pelvic structures. In such situations, careful dissection and proper reconstruction of the pelvic musculature after tumor excision is crucial for preservation (as much as possible) of the functional potential in these patients. This would be better performed by an experienced pediatric colorectal surgeon [1].

Solid SCT are thought to carry poorer prognosis [8,20]. This may 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 hyper-dynamic circulation, besides the risk of bleeding complications. [7] Controlling the median sacral artery prior to surgical excision has been recommended in some reports, which can be performed via laparoscopy. [21] The other poor prognostic aspect with solid tumors is related to the risk of malignancy. [20] In our series, eight cases were predominantly solid; two of them (25%) proved to be malignant and eventually died. Both cases showed signs of tumor invasion plus or minus distant metastasis in their preoperative imaging. Distant metastases may be a sign of inoperability and an indication for chemotherapy.

On the other hand, predominantly cystic SCT were not always benign as it might be expected. [20] 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. The pure cystic type C1 was always associated with low vascularity, easy dissection, and mature pathology;

however, it may cause diagnostic confusion with meningeoceles. [22, 23]

MRI had the advantage of superior soft tissue resolution (tumor consistency), better demonstration of the vertebral and spinal anatomy, depiction of the length of the 'non-ossified' coccyx to be excised with the tumor, and of course the lack of ionizing radiation. However, more time and sometimes general anesthesia were needed to complete the examination. On the other hand, CT scan was superior in detection of intra-lesional calcifications (fig. 9) and vertebral bony destruction by tumor invasion (fig. 8b).
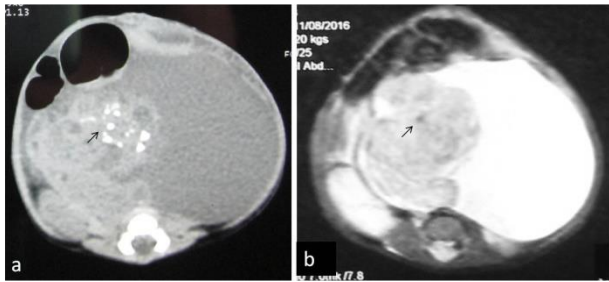
Figure 9: Imaging of predominantly cystic Sacrococcygeal teratoma (C3) with abdominal extension in a 25-day-old female: a) CT scan; b) MRI axial T2WI. Note the superior demonstration of tumor calcification (black arrow) in (a) than (b).

We believe our proposed preoperative grading system for SCT can help in clarifying some prognostic factors and anatomical landmarks important in planning surgery for such rare tumors. However, this represents a preliminary work that still has to be validated by its application to other cohorts with SCT. Also, the study did not address the evolving issue of ante-natal diagnosis except in one case. However, the detailed imaging characteristics that we have discussed are mainly based on MRI studies. This may prove to be useful for ante-natal diagnosis as well, which is also based on MRI studies.

CONCLUSION

MRI is a useful modality in the preoperative assessment of cases of SCT by providing a grading system that can indicate for the prognosis and degree of expected surgical challenge.

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