Sacrococcygeal teratoma: management and outcomes

Hussam S. Hassan and Akram M. Elbatarny

Background/aim **Sacrococcygeal teratomas are the most common tumor during the neonatal period. They are either benign (mature) or malignant (immature, composed of embryonic elements). This retrospective study aims at reviewing our experience in management and outcome of this pathology during the period from January 2008 to November 2013.**

Patients and methods **Charts were designed to collect the following data: the age at presentation, sex, clinical presentation, associated anomalies, Altman's classification, investigations, management modality, tumor histology with respect to type and resection margins, outcome of treatment, and bowel or urinary complications.**

Results **A total of 20 patients were included in this study. There were six male patients and 14 female patients. Ten patients were Altman type I, seven were type II, one was type III, and two were type IV. Surgical intervention was carried out using a posterior sacral approach in 17 patients (all type I and II patients) or combined abdominal and posterior sacral approaches in the remaining three patients (types III and IV). Recurrence occurred in two patients; these were types III and IV. We had one mortality. Two patients reported involuntary bowel movements, two reported frequent soiling, and five reported constipation.**

Conclusion **Awareness about this pathology among practitioners is essential and would have avoided complications in this series. Early diagnosis and complete excision with removal of the coccyx is associated with good prognosis. Recurrence is related to tumor spillage during excision. Long-term lower gastrointestinal problems (constipation, fecal soiling) correlate with Altman's classification of the tumor.** ***Ann Pediatr Surg*** **10:72–77 © 2014 Annals of Pediatric Surgery.**

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Introduction

Sacrococcygeal teratomas (SCTs) are congenital neoplasms that arise from the coccyx and comprise tissues derived from all three germ layers. They are believed to arise from embryologically multipotent cells within Hensen node, a remnant of the primitive streak in the coccygeal region [1].

Although it is the most common tumor during the neonatal period, SCTs are rare with an incidence of approximately one in 35 000–40 000 live births. Female individuals are affected more frequently with a female-to-male ratio of almost 4 : 1 [2,3]; 18% of these infants have additional congenital anomalies [4].

SCTs are either benign (mature) or malignant (immature, composed of embryonic elements). Mature teratomas are more common in both neonates (68%) and older children (73%) [5].

Altman *et al*. [6] have classified SCTs into four groups. Type I tumors are almost exclusively exterior with minimal pelvic component; type II tumors have a significant pelvic component (hour-glass pattern); type III tumors have a larger proportion of intra-abdominal and intrapelvic component than the external component; and type IV tumors are exclusively presacral with no external component. Altman and colleagues also reported that type I tumors were rarely malignant.

SCT can be diagnosed during antenatal ultrasonography. Polyhydramnios is found in up to 20% of cases [7]. They have a wide variety of clinical presentations depending on their size, vascularity, and degree of mass effect upon adjacent structures [8].

Before 2 months of age, most of these lesions are benign, and surgical extirpation with coccygectomy can be accomplished with relatively low morbidity and mortality [9,10]. Excision of the coccyx is mandatory in all cases. Failure to remove the coccyx results in 30–40% recurrence rate, with a higher probability of malignancy [11].

The use of α-fetoprotein (AFP) as a tumor marker is well established, and persistent elevated level may indicate a residual tumor, recurrence, or malignant degeneration [12]. However, it should be performed with caution in infants because its levels are normally elevated in the first 8 months of life [13]. The mean time required for AFP to be normalized after SCT resection is about 9 months [14].

Recurrence after resection varies from 2 to 35%. This may result from incomplete surgical excision with the presence of microscopic residues, nonresection of the entire coccyx, and/or tumor spillage [15,16].

Although the postnatal mortality of SCTs is low [17], prenatally diagnosed SCTs can be associated with higher mortality [18]. Outcomes after surgical resection during

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the neonatal period are generally favorable, although tumor recurrence with malignant transformation can occur in 10–20% of patients [19].

Although severe distal motor deficits are unusual after resection, other neurologic sequelae can be commonly observed and include subtle abnormalities in gait as well as fecal and/or urinary dysfunction in up to 50% of patients [20].

This study was a retrospective file review for cases of SCTs at our institution regarding presentation, complications, management, and outcome.

Patients and methods

Records of all cases of SCTs seen at the Pediatric Surgical Unit in Tanta University Hospitals and affiliated Hospitals during the period from January 2008 to November 2013 were reviewed.

Special charts were designed to retrieve the following data from records: the age at presentation, sex, clinical presentation, associated anomalies, Altman's classification, investigations, management modality, tumor histology with respect to type and resection margins, and outcome of treatment.

The data retrieved from the follow-up visits included: wound complications as disruption or necrosis, any masses at the resection site, careful PR, stabilization of cardiac condition after excision of the huge highly vascular cases, and serum AFP evaluation and pelvic ultrasound every 3 months.

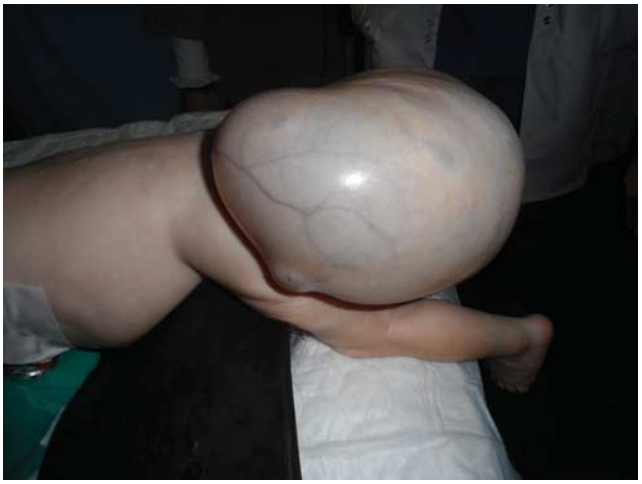
All patients operated upon from 2008 to 2010 (3–5 years of age) were contacted to fill in a special chart evaluating urinary incontinence, soiling, and constipation. Questions were categorized into parameters – (i) involuntary bowel control (inability to feel the urge to use the toilet to have a bowel movement, inability to verbalize it, and to hold the bowel movement until the patient reaches the bathroom); (ii) soiling; (iii) constipation; and (iv) urinary incontinence – according to the parameters used for evaluation of bowel function in patients with anorectal malformations (ARM) introduced by Peña [21].

The study was approved by the ethical committee.

Results

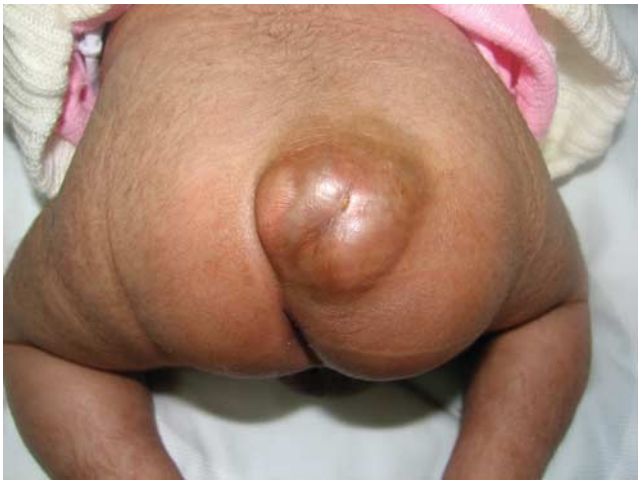
A total of 20 patients were included in this study. There were six (30%) male patients and 14 (70%) female. The age at presentation ranged from 1 day to 70 days. The diagnosis was made antenatally in nine patients, at birth in nine, and after the neonatal period in two. The diagnosis was made by clinical examination, which showed the visible external component of tumor in 18 (90%) patients. Plain radiograph showed anterior displacement of the rectum by the tumor in 10 (50%) patients. Double contrast computed tomography showed that the rectum was compressed and the urinary bladder was displaced in 10 (50%) patients. No patients showed intraspinal extension. Ten (50%) patients were Altman type I (Fig. 1), seven (35%) were type II (Fig. 2), one (5%) was type III (Fig. 3), and two (10%) were type IV (Fig. 4).

Fig. 1



Type I sacrococcygeal teratoma.

Fig. 2



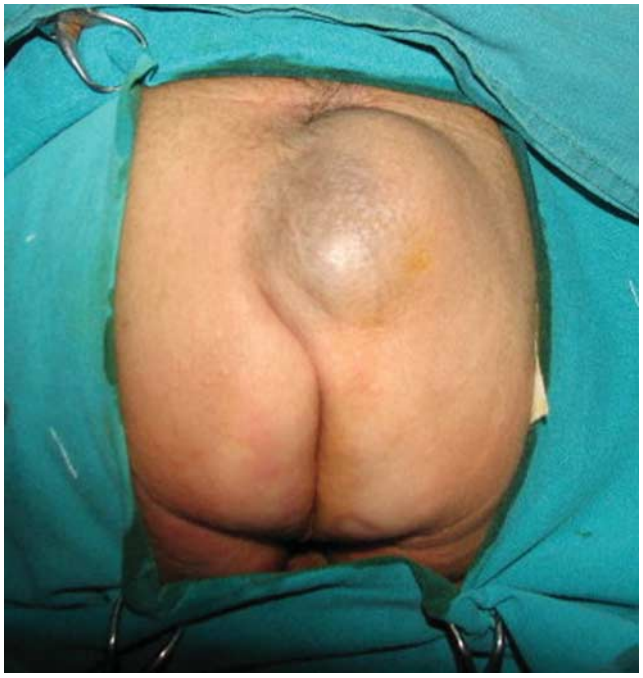
Type II sacrococcygeal teratoma.

Associated congenital anomalies were found in three (15%) patients. These included imperforate anus (two cases) and left primary megaureter (one case).

Eighteen (90%) patients were treated on selective basis; two (10%) patients required urgent intervention, one due to tumor rupture (Fig. 5) and another due to hemorrhage inside the tumor (Fig. 6).

Surgical intervention was carried out using a posterior sacral approach in 17 (75%) patients (all type I and II patients) (Fig. 7) or combined abdominal and posterior sacral approaches in the remaining three (types III and IV) patients (Fig. 8). The tumor was resected *en block* with the coccyx in all patients. Spillage due to rupture of the tumor during dissection occurred in two (10%) patients.

Fig. 3



Type III sacrococcygeal teratoma.

Fig. 4

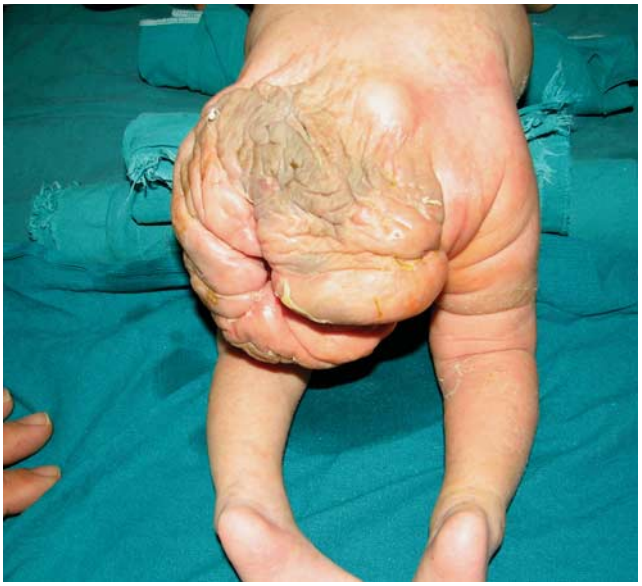
Type IV sacrococcygeal teratoma. UB, Urinary Bladder; UT, uterus.

Macroscopically, four (20%) cases of our series were solid, six (30%) were cystic, and 10 (50%) were mixed.

Histopathological examination of the resected tumor revealed that 15 (75%) patients had grade 0 pathology (only mature tissue), three (15%) had grade I (rare foci of immature tissues), one (5%) had grade II (moderate quantities of immature tissues), and one (5%) had grade III (large quantities of immature tissue with malignant yolk sac elements).

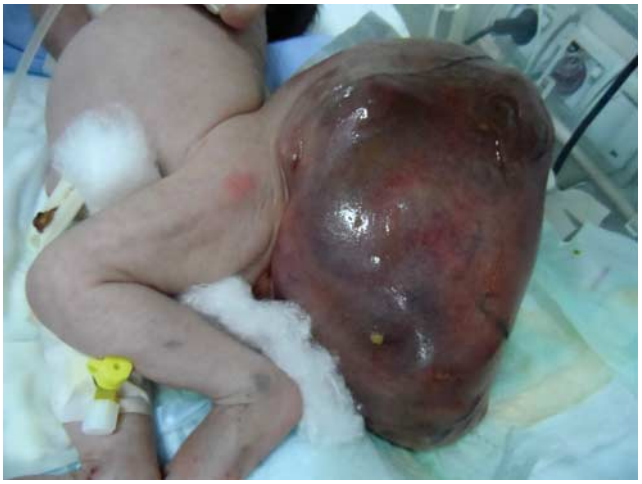
Adjuvant chemotherapy was used in one patient proved to be malignant by histopathological examination.

Fig. 5



Ruptured sacrococcygeal teratoma.

Fig. 6



Hemorrhage in a sacrococcygeal teratoma.

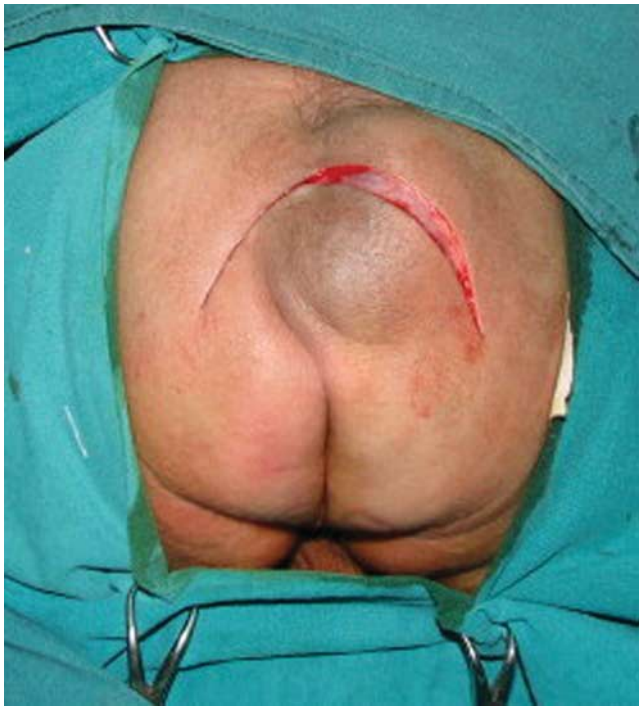
The follow-up period ranged from 6 months to 5 years. AFP level measures decreased within 3–9 months. It became elevated again in recurrent cases. Recurrence occurred in two patients; these were types III and IV.

Only one (5%) mortality occurred in this series; this case was an Altman type IV, which was malignant on histopathological examination (grade III) and received adjuvant chemotherapy after surgical extirpation.

Ten patients were operated early in this study (during the period from January 2008 to December 2010, now 3–5 years of age). We lost two of them from follow-up.

Among the remaining eight patients, two (25%) reported involuntary bowel movements (loss of the ability to feel the urge to use the toilet and hold the bowel motion until

Fig. 7



Sacrococcygeal teratoma excision: posterior sacral approach.

reaching the bathroom). Two (25%) patients had frequent soiling in their underwear during the day and sometimes during the night. Five (63%) patients reported constipation; of them, two had soiling and one had involuntary bowel movements. No patient reported urinary problems.

The operative scar was satisfactory in all (Fig. 9) but five (25%) patients who showed hypertrophic ugly scars. All five patients were having large tumor size.

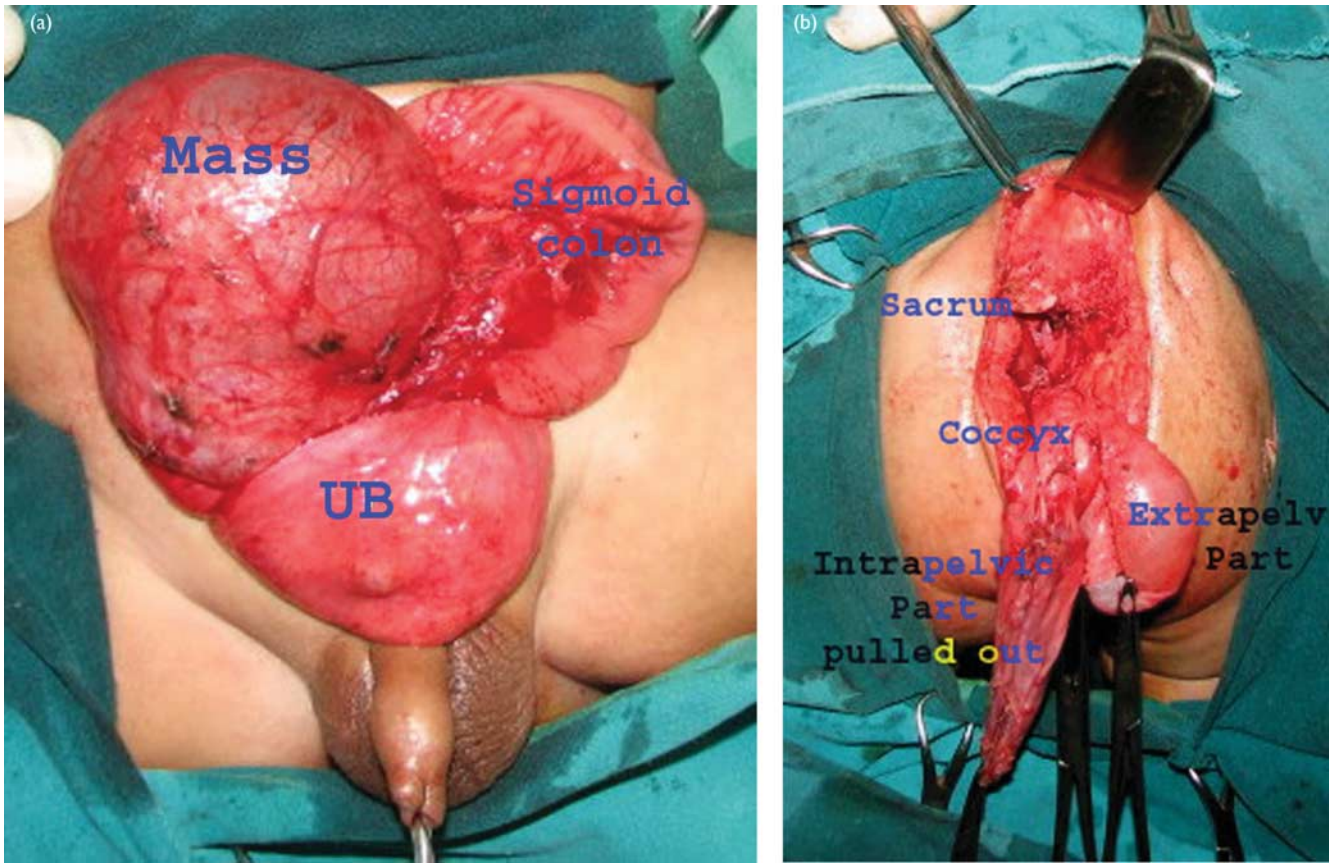
Discussion

SCT is the most common tumor diagnosed in the neonate. Approximately, 80% of affected infants are girls, a 4:1 female-to-male preponderance [3]. In our series, female infants were 70%. This slight difference may be explained by the relatively small number of patients in our series.

In this study, associated congenital anomalies were found in three (15%) patients, two cases of imperforate anus and one case of left primary megaureter. The reported incidence in the literature varies widely from 5 to 26% [22–24].

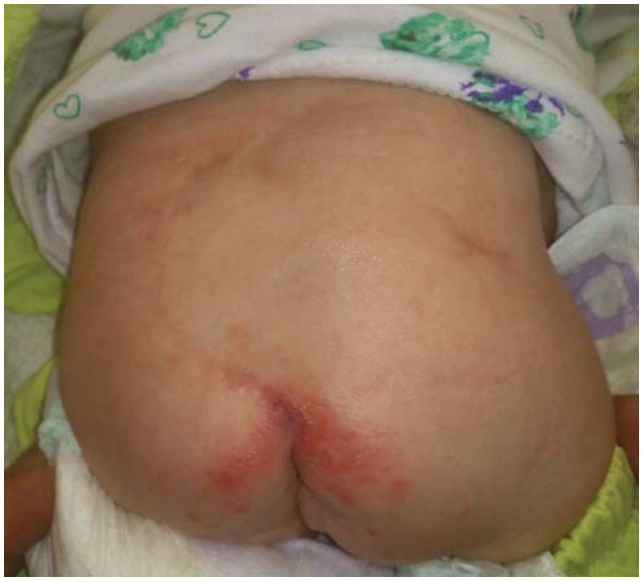
Although described as early as the 19th century, it was not until the advent of antenatal ultrasonography that SCTs were diagnosed *in utero* [25]. The majority of SCTs

Fig. 8



Resection of SCT type III; combined abdominal and posterior sacral approach, (a) abdominal part. (b) the mass is pulled out of the posterior sacral incision. UB, Urinary bladder.

Fig. 9



Satisfactory scar of excision.

present between the 22nd and the 34th week of gestation. The diagnosis of SCT on routine sonograms is associated with a greater than expected incidence of prenatal and perinatal complications [26].

In their published report on their survey among the members of the American Academy of Pediatrics, Surgical Section, Altman *et al.* [6] classified SCTs according to their extent into four groups: type I (47%), type II (35%), type III (8%), and type IV (10%). This was comparable with the incidences in this series.

Antenatal diagnosis is important to avoid complications during delivery. In the current study, only nine (45%) patients were diagnosed antenatally. The awareness among obstetrics–gynecologists regarding the importance of antenatal diagnosis of these patients is increasing in our locality. In a previous series, only 20% of cases were diagnosed antenatally [27].

Roybal *et al.* [28] recommended preemptive early delivery by cesarean section when the tumor exceeds the diameter of 5 cm, to avoid complications such as rupture and hemorrhage. In the current study, among the nine patients diagnosed antenatally, five patients had elective cesarean section because of a large tumor size exceeding 10 cm. One of these patients had hemorrhage and the tumor size was huge (the baby's weight was 4 kg, the tumor weight was 1400 g). This patient was managed on an urgent basis.

However, one patient with huge SCT (primary management was at a peripheral hospital) was not diagnosed antenatally and underwent vaginal delivery. This patient had a tumor rupture and underwent urgent operative intervention.

Macroscopically, four (20%) cases of our series were solid, six (30%) were cystic, and 10 (50%) were mixed. This result is in agreement with other results in the literature [29].

SCTs are graded histologically as follows [30]: grade 0, tumor contains only mature tissue; grade I, tumor contains rare foci of immature tissues; grade II, tumor contains moderate quantities of immature tissues; and grade III, tumor contains large quantities of immature tissue with or without malignant yolk sac elements. Grading of SCTs, unlike that of ovarian teratomas, does not seem to correlate directly with prognosis [5]. In this series, the only case proven to be malignant had a grade III histopathology. However, it is difficult to draw a conclusion from this association.

The incidence of malignancy in SCTs increases with age. By the age of 9 months, the incidence of malignancy is around 70% [31]. In this series, the only case with malignancy was diagnosed at the age of 3 months.

The primary treatment of SCTs is early surgical resection with complete excision of the coccyx [9,10]. Adjuvant chemotherapy is used in malignant cases in the form of combination of vincristine, actinomycin D, and cyclophosphamide, with or without adriamycin [32]. Early surgical intervention is associated with better prognosis [9,10]. In this series, the only patient with malignant disease and mortality was the one diagnosed and managed lately (at 90 days).

The surgical approach depends on the degree of pelvic extension [6]. Surgical intervention was carried out using a posterior sacral approach in 15 (75%) patients (all type I and II cases) or combined abdominal and posterior sacral approaches in the remaining three patients (types III and IV). Intradural invasion of a SCT is a rare variant that has generally been associated with a favorable neurologic outcome. However, extension into the spinal canal may cause paraplegia [8]. We did not encounter any intradural or intraspinal extension in any of our patients. This is expected, as intraspinal extension of SCTs is exceedingly uncommon, being clearly documented in only a few cases [8].

One case (type IV), primarily managed at a peripheral hospital, was misdiagnosed as a pelvic abscess and was evacuated. Recurrence occurred after 3 months, and she was explored abdominally and the tumor was excised (the coccyx was not removed). Another recurrence occurred 4 months later (as a retrorectal mass with an external gluteal part resembling Altman type III). The case was referred to our institution and had the tumor and coccyx removed through a combined abdominal and posterior sacral approach. The patient did well after that, with no recurrence.

The other case that recurred after excision was a type III SCT in which spillage of tumor tissue occurred during the primary operation.

Of the eight patients in this series who are still followed up as an outpatient and who are at least 3 years of age, two (25%) patients reported involuntary bowel movements (loss of the ability to feel the urge to use the toilet and hold the bowel motion until reaching the bathroom).

These two patients were Altman types II and III. Two (25%) patients had frequent soiling in their underwear during the day and sometimes during the night. These two patients were Altman types II and III. Five (63%) patients reported constipation; of them, two patients had soiling and one had involuntary bowel movements. These patients were Altman types II and III. No patient reported urinary problems. Bowel complications correlated with the Altman's classification. All of them occurred in types II and III. No such complications occurred in type I SCT. This result contradicts the one reported by Derikx *et al.* [33] who found no correlation between these complications and Altman's classification. The incidence is a bit high; however, it should be noted that this is not the overall incidence but rather the incidence among the eight patients who were regular in long-term follow-up.

Conclusion

SCT is a common neonatal neoplasm. Awareness about this pathology among practitioners is essential, especially antenatal diagnosis, and would have avoided complications in this series. In addition, delivery in a tertiary hospital by cesarean section, when needed, should be emphasized. Early diagnosis and complete excision with removal of the coccyx is associated with good prognosis. Recurrence is related to tumor spillage during excision, incomplete resection, and leaving the coccyx behind. Long-term lower gastrointestinal problems (constipation, fecal soiling) correlate with Altman's classification of the tumor.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

References

1 Moazam F, Talbert JL. Congenital anorectal malformations: harbingers of sacrococcygeal teratomas. *Arch Surg* 1985; **120**:856–859.
2 Moore SW, Satgé D, Sasco AJ, Zimmermann A, Plaschkes J. The epidemiology of neonatal tumours. Report of an international working group. *Pediatr Surg Int* 2003; **19**:509–519.
3 Laberge JM, Puligandla PS, Shaw K. Teratomas, dermoids, and other soft tissue tumors. In: Holocomb GWIII, Murphy JP, editors. *Ashcraft's pediatric surgery*. 5th ed. Philadelphia, PA: Saunders Elsevier; 2010. pp. 915–935.
4 Reinberg Y, Long R, Manivel JC, Resnick J, Simonton S, Gonzalez R. Urological aspects of sacrococcygeal teratoma in children. *J Urol* 1993; **150**:948–949.
5 Keslar PJ, Buck JL, Suarez ES. Germ cell tumors of the sacrococcygeal region: radiologic–pathologic correlation. *Radiographics* 1994; **14**: 607–620.
6 Altman RP, Randolph JG, Lilly JR. Sacrococcygeal teratoma: American Academy of Pediatrics Surgical Section Survey-1973. *J Pediatr Surg* 1974; **9**:389–398.
7 Rothschild MA, Catalano P, Urken M, Brandwein M, Som P, Norton K, Biller HF. Evaluation and management of congenital cervical teratoma. Case report and review. *Arch Otolaryngol Head Neck Surg* 1994; **120**:444–448.
8 Kunisaki SM, Maher CO, Powelson I, Gemmete JJ, Hirschl RB, Mychaliska GB. Benign sacrococcygeal teratoma with spinal canal invasion and paraplegia. *J Pediatr Surg* 2011; **46**:e1–e4.
9 Rescorla FJ, Sawin RS, Coran AG, Dillon PW, Azizkhan RG. Long-term outcome for infants and children with sacrococcygeal teratoma: a report from the Childrens Cancer Group. *J Pediatr Surg* 1998; **33**: 171–176.
10 Makin EC, Hyett J, Ade-Ajayi N, Patel S, Nicolaides K, Davenport M. Outcome of antenatally diagnosed sacrococcygeal teratomas: single-center experience (1993–2004). *J Pediatr Surg* 2006; **41**: 388–393.
11 Berman MC, Cohen HL. *Diagnostic medical sonography: a guide to clinical practice; obstetrics & gynecology*. 2nd ed. Philadelphia: Lippincott-Raven; 1997. pp. 313–314.
12 Kuhajda FP, Taxy JB. Oncofetal antigens in sacrococcygeal teratoma. *Arch Pathol Lab Med* 1983; **107**:239–242.
13 Brewer JA, Tank ES. Yolk sac tumor and alphafetoprotein in first year of life. *Urology* 1993; **42**:79–80.
14 Barreto MW, Silva LV, Barini R, Oliveira-Filho AG, Sbragia L. Alpha-fetoprotein following neonatal resection of sacrococcygeal teratoma. *Pediatr Hematol Oncol* 2006; **23**:287–291.
15 Huddart SN, Mann JR, Robinson K, Raafat F, Imeson J, Gornall P, *et al.* Sacrococcygeal teratomas: the UK children's cancer study group experience. I. Neonatal. *Pediatr Surg Int* 2003; **19**:47–51.
16 Göbel U, Calaminus G, Engert J, Kaatsch P, Gadner H, Bökkerink JP, *et al.* Teratomas in infancy and childhood. *Med Pediatr Oncol* 1998; **32**: 8–15.
17 Swamy R, Embleton N, Hale J. Sacrococcygeal teratoma over two decades: birth prevalence, prenatal diagnosis and clinical outcomes. *Prenat Diagn* 2008; **28**:1048–1051.
18 Makin EC, Hyett J, Ade-Ajayi N, Patel S, Nicolaides K, Davenport M. Outcome of antenatally diagnosed sacrococcygeal teratomas: single-center experience (1993–2004). *J Pediatr Surg* 2006; **41**:388–393.
19 Derikx JP, De Backer A, van de Schoot L, Aronson DC, de Langen ZJ, van den Hoonaard TL, *et al.* Factors associated with recurrence and metastasis in sacrococcygeal teratoma. *Br J Surg* 2006; **93**:1543–1548.
20 Draper H, Chitayat D, Ein SH, Langer JC. Long-term functional results following resection of neonatal sacrococcygeal teratoma. *Pediatr Surg Int* 2009; **25**:243–246.
21 Peña A. Anorectal malformations. *Semin Pediatr Surg* 1995; **4**:35 –47.
22 Gross RW, Clatworthy HW Jr, Meeker IA Jr. Sacrococcygeal teratoma in infants and children: report of 40 cases. *Surg Gynecol Obstet* 1951; **92**:341–352.
23 Ein SH, Adeyemi SD, Mancer K. Benign sacrococcygeal teratoma in infants and children: a 25 year review. *Ann Surg* 1980; **191**:382–384.
24 Carney JA, Thompson DP, Johnson CL, Lynn HB. Teratomas in children: clinical and pathologic aspects. *J Pediatr Surg* 1972; **7**: 271–282.
25 Kum CK, Wong YC, Prabhakaran K. Management of fetal sacrococcygeal teratoma. *Ann Acad Med Singapore* 1993; **22**:377–380.
26 Holterman AX, Filiatrault D, Lallier M, Youssef S. The natural history of sacrococcygeal teratomas diagnosed through routine obstetric sonogram: a single institution experience. *J Pediatr Surg* 1998; **33**:899–903.
27 Hashish AA, Fayad H, Al-Attar A, Radwan MM, Ismael K, Ashour MM, Elhalaby E. Sacrococcygeal teratoma: management and outcomes. *Ann Pediatr Surg* 2009; **5**:119–125.
28 Roybal JL, Moldenhauer JS, Khalek N, Bebbington MW, Johnson MP, Hedrick HL, *et al.* Early delivery as an alternative management strategy for selected high-risk fetal sacrococcygeal teratomas. *J Pediatr Surg* 2011; **46**:1325–1332.
29 Danzer E, Hubbard AM, Hedrick HL, Johnson MP, Wilson RD, Howell LJ, *et al.* Diagnosis and characterization of fetal sacrococcygeal teratoma with prenatal MRI. *Am J Roentgenol* 2006; **187**:W350–W356.
30 Graf JL, Housely HT, Albanese CT, Adzick NS, Harrison MR. A surprising histological evolution of preterm sacrococcygeal teratoma. *J Pediatr Surg* 1998; **33**:177–179.
31 Hager T, Sergi C, Hager J. Sacrococcygeal teratoma – a single center study of 43 years (1968–2011) including follow-up data and histopathological reevaluation of specimens. *Eur Surg* 2012; **44**:255–266.
32 Raney RB Jr, Chatten J, Littman P, Jarrett P, Schnaufer L, Bishop H, D'Angio GJ. Treatment strategies for infants with malignant sacrococcygeal teratoma. *J Pediatr Surg* 1981; **16 (Suppl 1)**:573–577.
33 Derikx JP, De Backer A, van de Schoot L, Aronson DC, de Langen ZJ, van den Hoonaard TL, *et al.* Long-term functional sequelae of sacrococcygeal teratoma: a national study in the Netherlands. *J Pediatr Surg* 2007; **42**:1122–1126.