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Sacrococcygeal teratoma with intraspinal extension: A case series and review of literature

Jesse Seilern und Aspang, Katherine Mary Burnand, HanLim Ong, Kate Cross, Dominic Thompson, Stefano Giuliani *

Great Ormond Street Hospital for Children, Department of Paediatric Surgery–Specialist Neonatal and Paediatric Surgery, NHS Foundation Trust, Great Ormond Street, London WC1N 3JH

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ABSTRACT

Background: Sacrococcygeal teratoma (SCT) is the most common teratoma in neonates and arises from the coccyx. SCT with intraspinal invasion is extremely rare and only reported in a few cases.
Methods: 37 patients with SCT were identified at our institution between 2000 and 2018. Three of these patients had SCT with intraspinal extension. A literature review for intraspinal extension associated with SCT, including mode of diagnosis, presentation, surgical approach and neurological sequelae, between 1993 and 2018 was also conducted.
Results: The authors report three cases of infants who were antenatally and/or postnatally diagnosed with a sacrococcygeal teratoma extending into the spinal canal. We illustrate the challenges of accurate diagnosis and therapeutic management. Postnatal magnet resonance imaging (MRI) was the best method to define spinal anatomy and extension of the tumors prior to surgery. Management with a multidisciplinary team approach including neuroradiology, neurosurgery and general surgery was used in our two most recent patients. The literature review yielded 6 cases of SCT with intraspinal extension.
Conclusion: Intraspinal extension in SCT is rare but should be excluded at birth before attempting any resection. In case of positive spinal invasion on Ultrasonography (US), MRI is essential to plan for surgery and possible laminectomy to be able to perform a radical resection of this congenital tumor. We recommend this multidisciplinary approach.
Level of evidence: Level IV.

Sacrococcygeal teratoma (SCT) is the most common neonatal tumor with an incidence of 1 in 35,000–40,000 live births [1]. Consisting of all three embryonic layers, SCTs can be benign or malignant. In 1974 Altman et al. classified the different location-types (type I–IV) as ranging from primarily exophytic (I) to completely intrapelvic (IV) [2]. The vast majority present in utero or at birth and are benign. The most common cause of mortality in neonates with SCT is internal tumor hemorrhage, which correlates with size and type, followed by high output cardiac failure, and perioperative bleeding [3]. Nevertheless, the survival rate of benign SCT ranges from 60% to 99% [4].

Despite the tumor developing at the base of the spine from pluripotent stem cells, intraspinal invasion is a particularly uncommon variant, presenting as a sacrococcygeal mass extending into the spinal canal diagnosed by US or MRI. With only a few cases reported in the literature, this tumor presentation has its unique operative challenges and undetermined long-term outcomes.

The aim of this paper is to present our experience of SCT with intraspinal extension and to review the literature in order to assist surgeons with the correct surgical planning and decision making for achieving radical resection and cure for this rare variant of a benign tumor.

1. Methods

With institutional approval, we reviewed all children who underwent SCT surgery between 2000 and 2018 resulting in 37 cases. Cases without intraspinal extension ($n = 31$) or cases where the teratoma was associated with myelomeningocele or as part of the Currarino spectrum ($n = 3$) were excluded.

The literature search was performed using the electronic National Institute of Health database PubMed.gov and Google Scholar. Broad search terms used ("sacrococcygeal" and "teratoma"), generating 990 hits. Only articles written in the English language from 1988 to 2018 were reviewed. Only documented cases of neonates or infants with SCTs with spinal extension were included, while other spinal defects such as meningocele, myelomeningocele and spina bifida in conjunct with SCT or Currarino triad were excluded.

* Correspondence to: S. Giuliani, Great Ormond Street Hospital for Children NHS Foundation Trust, Great Ormond Street, London WC1N 3JH. Tel.: +44 0207 405 9200x0669.
E-mail address: stefano.giuliani@gosh.nhs.uk (S. Giuliani).

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1.1. Case 1

A term baby girl delivered by spontaneous vaginal delivery was noted at birth to have a mass arising over the lumbosacral spine. Antenatal scans had been normal. She passed urine and meconium soon after birth. There was a palpable soft tissue mass (15 × 10 × 10 mm) covered by intact skin on clinical evaluation and symmetrical muscle bulk in the buttocks and lower limbs. A subsequent ultrasound (US) suggested occult spinal dysraphism with a lipomyelocele. However, magnetic resonance imaging (MRI) revealed a heterogeneous mass, causing compression of the sacral nerve roots with enhancement (Fig. 1). No other abnormality was demonstrated in the spinal canal above the level of the conus or intracranially. It was most likely an SCT (23 × 60 × 32 mm) with cranial tumor extension into the spinal canal. Alpha-fetoprotein (αFP) and bHCG were normal at the time of resection.

During surgery at 26 days of age, the tumor was dissected circumferentially towards the lumbar vertebrae. A laminectomy was performed, providing a good view of the tumor from above and below. The tumor was visualized intimately binding to the dura on the right at the S1 level at the origin of the nerve root. The mass was removed en bloc along with the coccyx, but the dura was opened and the S1 nerve root sacrificed. Histology demonstrated a mature teratoma without yolk sac elements. She developed paraparesis with a neuropathic bladder and bowel, which became obvious soon after surgery. Currently, she is 16 years old, independently mobile and active in competitive swimming. She regularly self-catheterizes during the day and uses bowel washouts. Thus far she has remained recurrence-free.

1.2. Case 2

A late-preterm female infant was delivered via emergency cesarean section owing to prolonged rupture of membranes. A large cystic sacral mass suggestive of SCT had been identified via US at 25 weeks' gestation without the suggestion of an intraspinal component. At birth she was in good condition and physical examination revealed a skin covered soft tissue sacral mass measuring 15 × 10 × 10 mm. She passed urine and opened her bowels spontaneously soon after birth and lower extremity motor function appeared normal. Her initial serum αFP was elevated with a normal bHCG. A US suggested sacrococcygeal teratoma extension into the spinal canal, as an echogenic lesion consistent with fatty tissue extending below the conus at L3–L5 was described to be attached to the filum. A preoperative MRI mapped a 50 × 20 × 60 mm sized tumor with cranial extension into the sacral canal (Figs. 2 & 3).

On day 18, the girl underwent surgical excision of the SCT as a joint procedure with general surgery and neurosurgery. Intraoperative neurophysiologic monitoring was discussed but dismissed owing to projected uninterpretable measurements, as well as concern about prolonged propofol infusion in neonates. Incision was made around the tumor-bulk with a cephalad extension for sacral laminectomy. Following isolation of the SCT from the rectum and pelvic floor, laminectomy was performed at the level of S4 and extension of the SCT to S1 was visualized. The intraspinal component was mobilized while preserving the lower sacral nerve roots and the tumor stalk was dissected from the dura without suggestion of intradural extension. After final excision and coccygectomy, pelvic floor, gluteal muscles and soft tissue were reconstructed and closed in separate layers. Microscopic pathologic examination of the specimen revealed a mature teratoma. The neonate made a good recovery and follow-up has been unremarkable at 10 months. Beta human chorionic gonadotropin (βHCG) and αFP levels fell to normal.

1.3. Case 3

A term male infant was delivered via spontaneous vaginal delivery in good condition. Physical examination revealed a skin covered soft tissue mass in the right buttock measuring 15 × 10 × 30 mm. The baby passed urine and opened his bowels spontaneously soon after birth and lower extremity motor function appeared normal. His initial αFP and βHCG level were normal. The pelvic ultrasound on day 4 of life showed an echogenic lesion consistent with fatty tissue, without evidence of an intraspinal component. However, on day 8 of life a feed and wrap MRI of the lumbosacral region with contrast revealed a sacrococcygeal mass (82 × 33 × 72 mm) with intraspinal extension up to the level of L5.

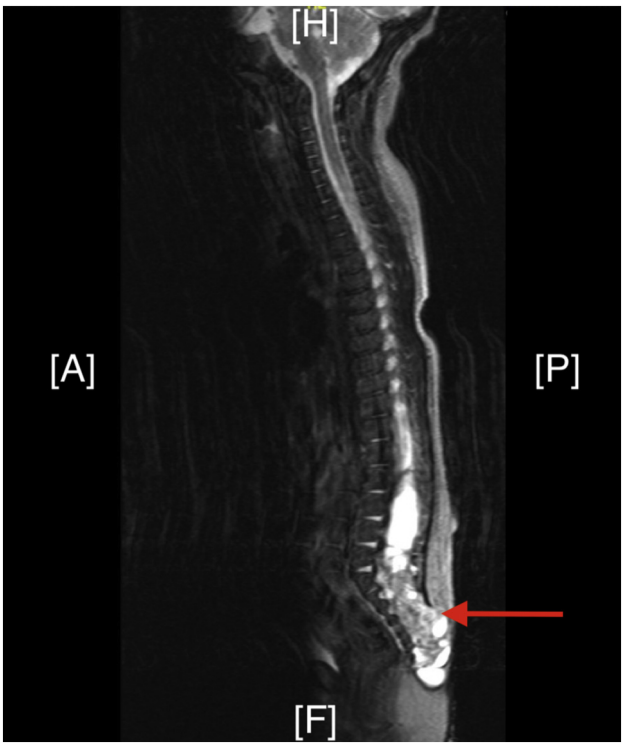

Fig. 1. Sagittal T2-weighted preoperative MRI showing a heterogenous mass, causing compression of the sacral nerve roots (red arrow). A = anterior, P = posterior, H = head, F = foot.

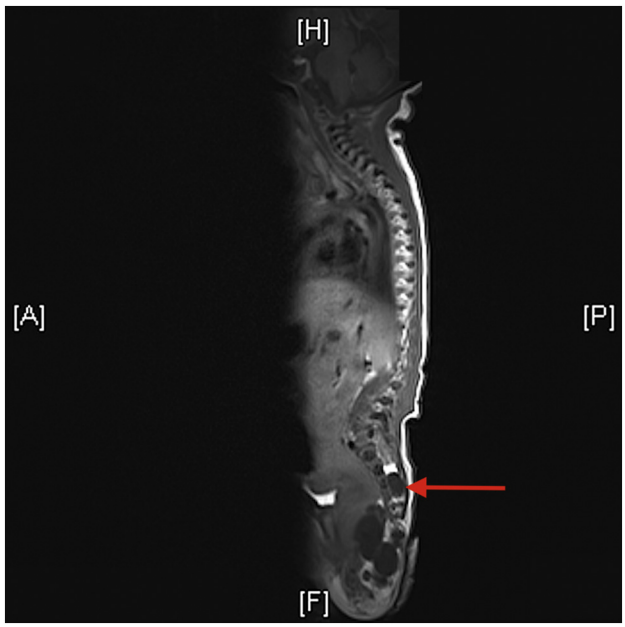

Fig. 2. Preoperative sagittal T1-weighted MRI with intraspinal extension marked (red arrow). A = anterior, P = posterior, H = head, F = foot.

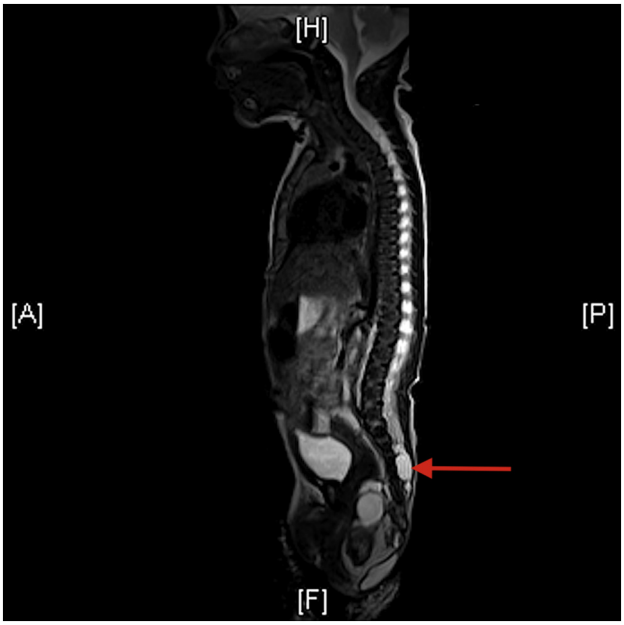

Fig. 3. Preoperative sagittal T2-weighted MRI with intraspinal extension marked (red arrow). A = anterior, P = posterior, H = head, F = foot.

On day 30 of life, the baby underwent surgical excision of the SCT as a joint procedure with general surgery and neurosurgery. The dissection was performed using a midline posterior incision across the bulk of the tumor extending 5 cm above the coccyx. Following isolation of the large cystic tumor expanding into the right buttock the SCT was visualized extending anterior to the sacrum with spinal infiltration. After circumferential isolation of the tumor, the coccyx was dissected and divided for an adequate approach to controlling median sacral arteries for dissection of the anterior extension. The intraspinal, extradural component was resected via posterior laminectomy and resected intact and en bloc. Microscopic pathologic examination revealed a mature teratoma. The neonate made a good recovery and was discharged home on postoperative day 5. Follow up has been unremarkable so far at 20 months. αFP level fell to normal.

2. Literature review

The literature search found 6 published cases of SCT with an intraspinal component. The characteristics of the cases are summarized in Table 1.

3. Discussion

Intradural teratomas and teratomas associated with spina bifida [5] have been described; however isolated SCT with intraspinal extension has only been reported in six neonatal cases between 1993 and 2017 [6–9]. The incidence of SCT with intraspinal extension was 3/37 (8%) in our case series. This is a higher incidence than reported in the literature (6 cases reported to date). We think this is important to consider during the preoperative assessment of a child with SCT. In particular, this could be related to the recurrence rate of such benign tumors even after complete resection of the coccyx. Roughly 11% of children with benign SCT show tumor recurrence 3 years postoperatively [10]. Recurrence is associated with tumor spillage at time of resection, failure to perform a coccygectomy and incomplete resection of the SCT [11,12]. Therefore, preoperative MRI may be helpful in decreasing the risk of incomplete resection not only evaluating intrapelvic extent of tumor but also assessing for intraspinal extension. Owing to the small number of recorded cases, the nature of the intraspinal component development (primary vs. secondary) is still unclear. Whether faster growth rate could correlate with the development of intraspinal component is also still to be determined. However, Coleman et al. found that faster SCT growth rate is associated with adverse outcomes including fetal and neonatal death [13].

The most common cause of mortality in neonates with SCT is internal tumor hemorrhage, which correlates with size and type, followed by high output cardiac failure, and perioperative bleeding [3]. Nevertheless, the survival rate of benign SCT ranges from 60% to 99% [4]. Optimal treatment of benign SCTs is complete excision of the tumor and coccygectomy within the first weeks of life. Yet, tumor recurrence with malignant transformation can occur in 10%–20% of patients who undergo surgical resection [10]. Furthermore, one-third of patients will have ongoing bladder and bowel dysfunction beyond childhood [14].

Intraspinal extension was prenatally detected in only one case by fetal MRI [15], while remaining cases were detected postnatally by US and/or MRI [6–9] or at operation [16]. The intraspinal component in our patients was visible on postnatal imaging; none had a fetal MRI.

In prenatal diagnosis of SCT, fetal MRI is mainly used to map out the size, consistency, and extension of the tumor in relation to the pelvic brim (important for surgical planning). Schey et al. [17] recommend US to accurately define anatomical extent of SCT in order to exclude invasion of adjacent structures. However, Danzer et al. [18] found that prenatal MRI was able to more accurately describe the extent of SCT compared to US. In the one case, where fetal MRI detected the intraspinal extension, fetal US had failed to demonstrate it. All authors of previous cases recommend timely diagnostic imaging of the SCT to provide suitable perioperative planning

Table 1
Overview of literature review of SCT with intraspinal extension.

Author & year	Diagnosis of SCT	Diagnosis of intraspinal component	Imaging modality	Intraspinal component +/− attachment	Level of intraspinal extension	Surgical intervention	Histology	Follow-up	Neurologic sequalae
Perrone EE, et al., 2017	Prenatal	Prenatal	US + MRI	Intradural + filum terminale + medulla	T-9	Sacral approach + laminectomy + intraoperative electrophysiologic monitoring	Mature teratoma	20 months	Neurogenic bladder
Shahjouei S, et al., 2015	Postnatal	Postnatal	MRI	Intradural + filum terminale	S-2	Sacral approach + laminectomy	Mature teratoma	12 months	no
Kunisaki SM, et al., 2011	Postnatal	Postnatal	X-ray + US + MRI	Intradural + filum terminale	T-12	Preoperative liquid embolization + anterior and sacral approach + laminectomy	Mature teratoma	8 months	Paralysis below T-12 + neurogenic bladder
Jelin E, et al., 2009	Prenatal	Postnatal	MRI	Intradural	L-4	laparoscopic approach + subsequent sacral approach (laminectomy NR)	immature teratoma	30 months	no
Riberio PRJ, et al., 1999	Postnatal	Postnatal	US + CT + MRI	Intradural	T-4	sacral approach + laminotomy	Mature teratoma	36 months	no
Powell RW, et al., 1993	Postnatal	Intraoperatively	X-ray	Intradural + filum terminale	NR	sacral approach + laminectomy	Mature teratoma	24 months	no

[6–8,15,16], which enables timely neurosurgical consultation and joint surgical planning [15].

Given the complications of intraspinal tumor resection [8,10,19–21], a majority of the reports stated that neurosurgical involvement aided in the outcome of their treatments. The most recent case of Perrone et al., 2017, reported intraoperative involvement of electrophysiologic monitoring during spinal dissection of the tumor. In one of our cases, electrophysiologic monitoring was also discussed but dismissed owing to projected uninterpretable measurements and potential anesthetic complications associated with prolonged propofol infusions.

Jelin et al. (2009) described initial laparoscopic approach to devascularize the intrapelvic mass before addressing the intraspinal extension in prone position. Although briefly discussed, this was not deemed appropriate given the size, location and vascular involvement of the SCTs in our case series.

Postoperative bladder and bowel dysfunction is well documented in patients with SCTs [22] with approximately 15% to 38% of children experiencing symptoms, presumably owing to either sacral plexus or pudendal injury [14,23,24]; the etiology of neurogenic bladder in the previously recorded cases remains unclear. So far, the level of intraspinal extension does not appear to correlate with neurological outcome (Table 1). Similar to our first case, Kunisaki et al., 2011, reported a case of a neurogenic bladder with paralysis from T12. Here, suspicion of spinal involvement was first raised on clinical evaluation of the newborn displaying obvious neurologic deficits (Table 1). Although a postnatal MRI was noted to be helpful for preoperative planning, the authors concede that better prenatal imaging and early detection of the intraspinal extension may have prevented paraplegia from mass effect via early delivery and urgent decompressive laminectomy [15].

Longer follow-up of all these patients will be necessary to determine if the neuropathic symptoms regress or develop over time, in order to draw any meaningful conclusions regarding bladder and bowel dysfunction, as well as tumor recurrence rates.

4. Conclusion

Preoperative imaging prior to SCT resection is essential not only to assess the pelvic extension of the tumor in relation to the sacral promontory and pelvic brim, but also to look for intraspinal extension. Postnatal MRI is observed to be more sensitive than US at detecting an intraspinal extension of the SCT and should be included in preoperative planning. Furthermore, detection of intraspinal components on fetal MRI may change both surgical approach and teams involved, as evidence of spinal cord compression can lead to subsequent changes in antenatal care, such as early delivery.

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Declarations of interest

None.

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