Sacroccygeal teratoma with an accessory limb an unusual presentation

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A R T I C L E   I N F O
Article history:
Received 14 February 2013
Received in revised form 20 February 2013
Accepted 21 February 2013

Key words:
Congenital
Sacroccygeal teratoma
Accessory limb

A B S T R A C T

A rare case of sacroccygeal teratoma (SCT) with a fully formed accessory limb. SCT is a neoplasm arising from the lumbosacral spine, usually a benign mature teratoma. This rare presentation of SCT, to our knowledge is the 7th case reported in the literature and is the first in the Pacific. Surgical treatment requires early vascular control, complete removal of the SCT and coccygectomy.

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1. Case report

A full-term female newborn baby who had a single, presumed normal antenatal scan at 28 weeks of gestation. She passed urine and meconium soon after a normal vaginal delivery in a small rural hospital. A week later, she was referred to the main tertiary hospital and was found to be non-dysmorphic with normal respiratory and cardiovascular systems and a large ulcerating sacrococcygeal teratoma (SCT) as in Fig. 1, which had displaced the anus anteriorly. The pelvic X-rays showed an extra-skeletal lower limb with fully formed digits as shown in Fig. 2. Having used the standard incision (Figs. 3 and 4) in the prone position as for other SCT tumors, the dissection was done with caution and with early vascular control, the SCT was excised-in-toto with coccygectomy and complete excision of the extra or accessory limb included in the specimen (Fig. 5). The accessory limb was hyper flexed within a separate layer of skin and was part of the main SCT, having shared the same blood supply (Fig. 6). This limb was not attached to the rectum or any other surrounding structures, which allowed a safe and complete excision. Lack of other preoperative imaging like a computed tomography (CT) scan or magnetic resonance imaging (MRI) as in this case is a major concern. The histology of the mass however confirmed a mature SCT with a fully formed limb.

2. Discussion

SCT is the most common neoplastic tumor in the newborn period [1]. It is a benign tumor with an incidence of 1 in 30,000–40,000 live births [2,3] and is more common in females with a 4:1 ratio. As in this case, sacrum is the most common site for germ cell tumors arising from the lumbosacral spine, usually a benign mature teratoma. This rare presentation of SCT, to our knowledge is the 7th case reported in the literature and is the first in the Pacific. Surgical treatment requires early vascular control, complete removal of the SCT and coccygectomy.

Despite the advanced fetal medicine care in developed nations, diagnosis is only made after delivery in most other developing countries. Smaller or intra-pelvic SCT tumors may not be quite obvious at birth and would often present later with lower limb weakness, constipation, urinary retention, abdominal distension and a pre-sacral mass on peri-rectal examination. Rarely, few children could present later with SCT malignancy as a result of late diagnosis or incomplete excision.

In most developed countries with pediatric surgical and fetal medicine centers, SCT’s are discovered on routine antenatal ultrasound scan [4]. Antenatal diagnosis often determines the potential outcome of the pregnancy, the mode and the site of delivery in these centers. Other determining factors include age and size of the fetus, the size of the tumor and its vascularity [5]. More vascular and large tumors seen on the fetal scan or MRI would often require a caesarean section to avoid significant bleeding into the tumor resulting in death [1]. These antenatal investigations are very limited or not available in our institution.

SCT can also be associated with anomalies such as anorectal malformations (ARM), bifid scrotum, hypospadias, spinal dysraphism, sacral agenesis, meningocele, hip dislocation and ventricular septal...
Fig. 1. A 10 × 8 cm sacrococcygeal mass with a central area of ulceration. Incision line as marked in black.

Fig. 2. X-ray of pelvis showing an accessory fully formed limb with the femur (single arrow) and tibia with the fibula (double arrows) being hyper flexed within the sacrococcygeal teratoma (SCT). The thick arrow head is demonstrating the normal limb.

Fig. 3. In the prone position demonstrating the incision to allow early access and ligation of the medial sacral vessels.

Fig. 4. Demonstrating the inferior part of the incision with a protruding part of the accessory limb correlating with the knee joint (arrow).

Fig. 5. Demonstrating the rectum (single arrow) and the left accessory blood supply (double arrows) after the right accessory blood supply had been divided.

Fig. 6. Excised sacrococcygeal teratoma (SCT) specimen with an accessory limb with a knee joint (double arrows) and five fully formed toes (single arrow) that was hyper flexed within the SCT tumor.
defect (VSD) [2]. Currarino triad has also been described as part of anomalies with SCT [2] but only a few cases of SCT with an extra or accessory limb have been previously documented.

To our knowledge, only six other SCT cases have been reported in the literature with an extra limb within the tumor [5–10] and we are reporting the 7th case report. This is also the first case report of such rare presentation from Fiji and the Pacific regions. Pre-operative pelvic X-ray would be quite useful in developing countries where CT scan or MRI is not readily available, as was found in this case. MRI would be more beneficial to assess any accessory blood supply apart from the medial sacral vessels [9] and it can also further define the SCT’s spatial relationship to surrounding organs [11]. The accessory vascular supply (artery and vein) in this case was located between the rectum and the peri-rectal muscles. One will need to be vigilant with SCT with an extra limb and be cautious despite our findings at surgery that they could share the same blood supply. The extra limb in this case was not attached to the rectum or any other important structures but was part of the SCT mass, hence it allowed for complete resection of the tumor once the main vascular and the accessory blood supply was controlled. This was combined with coccycectomy to complete the resection and avoid any recurrence.

References