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Sacrococcygeal teratoma: Atypical presentations in two neonates

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ABSTRACT

The etiology of sacral masses in neonates is highly variable. The differential diagnosis includes sacrococcygeal teratoma (SCT), meningocele, myelomeningocele, rectal abscess, lymphangioma, hemangioma, lipoma, perineal cyst, bladder neck obstruction, imperforate anus, and rectal prolapse. Not only are there numerous pathologies in the differential diagnosis but also each entity can present in a classic or atypical manner. As the atypical presentations of these pathologies can create a confusing clinical picture, one must have a high index of suspicion and include the entire differential diagnosis when evaluating patients with perineal masses. A combination of history and physical examination, laboratory analysis, as well as pre- and postnatal imaging can help provide clarity about the patient's diagnosis. In this report, we present the work-up, diagnosis, and treatment of two neonates with perineal masses and atypical presentations of SCTs.

1. Introduction

Sacrococcygeal teratomas (SCTs) are the most common extragonadal tumor of the newborn [1] with an incidence of 1 in 35,000 live births [2]. It typically affects females three to four times more often than males [3]. Embryologically, these masses are derived from the pluripotent cells in Hensen's node of the primitive streak. These lesions contain components arising from all three layers, endoderm, mesoderm, and ectoderm [4–7], and they typically have both solid and cystic components [7].

Classification of SCTs is determined by its anatomic location and degree of intra-abdominal extension. Type I lesions (46.7%) are predominantly external with a minimal presacral component; Type II (34.7%) are external with a significant intra-pelvic component; Type III (8.8%) have minimal external component with significant intra-pelvic and abdominal extension; and Type IV (9.8%) are located entirely within the abdomen and pelvis without an external component [8,9]. SCTs often present in the neonatal period with a large exophytic tumor that protrudes from the sacral region. They can also present in infancy or childhood with a mass located primarily in the pelvis [8].

Given recent advances in ultrasound technology, SCTs are now frequently diagnosed prenatally [10,11]. Once diagnosed, serial evaluation via ultrasound, echocardiography, and often Magnetic Resonance Imaging (MRI) is performed to monitor fetal growth, tumor size, and development of cardiac failure or hydrops [12,13]. On imaging, SCTs usually appear as a mass of mixed echogenicity extending from the sacrum or growing from the sacrum into the pelvis [6]. The average size of SCTs is 8 cm with a range of 1–30 cm [14]. The large size of some masses may result in the distortion of pelvic and sacral anatomy, rectal stenosis, urinary tract obstruction, dystocia, hip dislocation, or club foot [7,12]. Additionally, given the fragility of the tumor tissue, SCTs can undergo spontaneous hemorrhage resulting in arteriovenous shunting that may lead to high-output cardiac failure [12]. Associated findings may include lateral displacement of the anus, or infrequently the opening of the anus in the tumor [14]. Here, we discuss two patients with atypical presentations of SCTs.

2. Case number 1

Male infant was born at 38 weeks via uncomplicated vaginal delivery with a perineal mass (Fig. 1). The mass was first identified on antenatal ultrasound at 20 weeks and measured 1.1 × 1.3 × 0.8 cm in size. It was described as a solid mass without vascular flow, and did not appear to extend internally into the fetal pelvis.

On physical examination of the infant in the prone position, the base of the mass extended to the left of the patient's anus, causing the anus to be displaced to the right. The mass had a pedunculated appearance and did not appear to have skin overlying its surface (Fig. 2). The baby passed meconium and had no signs of gastrointestinal obstruction. Magnetic Resonance Imaging (MRI) on day of life (DOL) 1 showed a 3.4 × 2.3 × 2.3 cm heterogeneous, predominantly extracorporeal mass with soft tissue, cystic, fatty, and possibly mineralized components. The mass extended toward the distal portion of the coccyx, and therefore most likely represented a sacrococcygeal teratoma (SCT) (Fig. 3). The

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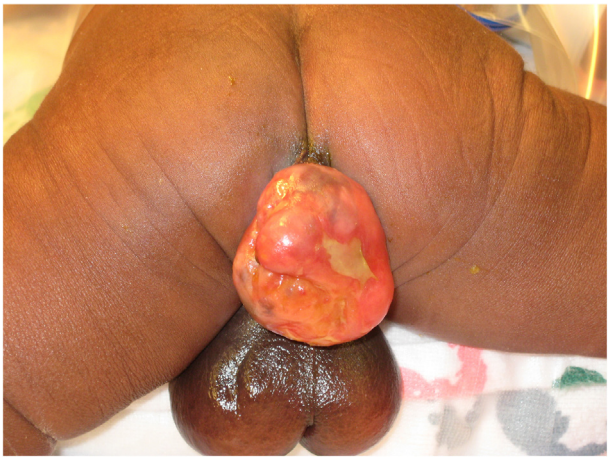

Fig. 1. Exophytic perineal mass.

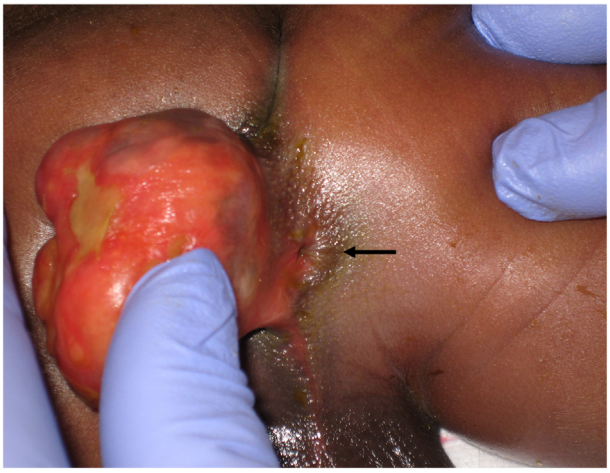

Fig. 2. Lateral displacement of the anus towards the right (black arrow) while the patient is in the prone position.

baby was also noted to have a 2–3 mm perimembranous ventral septal defect on echocardiogram and hypospadias. Renal ultrasound performed on DOL 3 was normal. Beta human chorionic gonadotropin (β-hCG) was within normal limits and Alpha-fetoprotein (AFP) was elevated at 38,760 ng/mL.

The patient was taken to the operating room for resection of the mass on DOL 3. Prior to incision, the Pena stimulator was used to identify the limits of the anal sphincter such that the circumferential dissection of the mass did not violate the sphincter. An elliptical incision was made around the base of the mass. The mass was then circumferentially dissected free from the surrounding tissue to the level of the rectum (Fig. 4a and b). A stalk was isolated and found to extend from the mass cranially toward the coccyx. A posterior sagittal midline incision was then made overlying the coccyx towards the posterior portion of the mass. The stalk was dissected proximally to the level of the coccyx, and a coccygectomy was performed. The entire specimen was sent for pathological examination (Fig. 5). Given the need to dissect a portion of the anus off of the mass, an anorectoplasty was performed. The anus was placed in the appropriate anatomic location within the sphincter, and the incision was closed without tension. There were no immediate intraoperative complications. Rectal dilation was performed on postoperative day (POD) 4. Pathology confirmed a teratoma with focal immature neural elements, and the longest segment measured 3.8 cm.

The child's postoperative course was unremarkable, and he was discharged on POD 8. Given the pathologic findings, no further

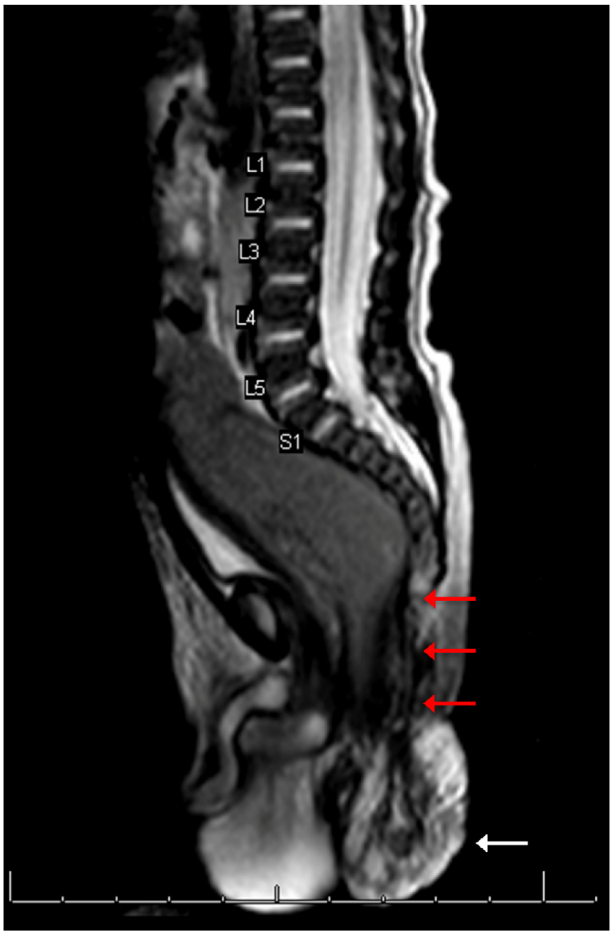

Fig. 3. Saggital view on MRI showing a heterogenous predominantly extracorporeal mass (white arrow) which extends towards the distal portion of the coccyx. Red arrows indicate the extension of the mass.

chemotherapy was necessary. At his most recent follow-up, which was 12 months postoperatively, he was doing well, tolerating feeds, and having normal stools. AFP level at this time was 5 ng/mL.

3. Case number 2

Female infant was born at 36 2/7 weeks via scheduled cesarean section for prenatally diagnosed perineal mass. Prenatal imaging consisted of the 21 week prenatal ultrasound, which revealed a complex mass at the level of the sacral spine, as well as fetal MRI, which showed a cystic and solid mass arising from the midline coccygeal and perineal region without evidence of significant intra-pelvic or intra-abdominal extension.

On physical examination, the mass appeared predominantly external with an overlying 6 cm circular skin defect (Fig. 6a and b). β-hCG was within normal limits and AFP was elevated at 80,123 ng/mL. MRI demonstrated a 10.8 × 5.2 × 8.4 cm mass with cystic and solid components (Fig. 7). The mass was shown to contact and truncate the coccyx. The patient was taken to the operating room on DOL 1 for resection. Using a scalpel, the skin defect was extended cephalad and the surrounding edges were dissected to provide adequate exposure of the mass. The mass included innumerable cysts, some of which showed evidence of intralesional hemorrhage. The mass was dissected along its superior edge in order to follow the bony midline structures to the coccyx. Next, the lateral aspects of the mass were dissected taking caution to preserve the gluteal musculature (Fig. 8). Prior to dissection of the inferior aspect of the mass, a Hegar dilator was placed in the rectum. The mass was then carefully dissected off of the rectum. At this