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Case Report

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Large neonatal sacrococcygeal teratoma in rural Palestine: A case report

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Abstract

The majority of solid tumors in newborns are sacrococcygeal teratomas (SCTs). These tumors originate in the sacrococcygeal region and contain tissue from all three germ layers. Nearly one in every 35,000 to 40,000 live births may be affected. Although the tumor is typically benign, the likelihood of malignant progression increases with the increasing age. The development of a mature teratoma can be attributed to either a pathological transformation of primordial germ cells or the development of a single germ cell tumor following the completion of the first phase of meiosis and the failure of meiosis type 2. Immature teratoma is associated with the malignant transformation of primitive germ cell layers and the renaissance of the primitive node. The diagnosis is not difficult and imaging provides the information to adequately care for affected infants. Histology, morphological categorization, problems such as hemorrhage or rupture, and mass effects are of utmost importance. Imaging features help to distinguish malignant tumors from benign tumors. However, imaging cannot predict the tumor's histological subtypes. Surgical resection is the primary method of treatment. In this article, we report a case of large neonatal SCT from rural Palestine and treated with resection and flap reconstruction.

Keywords: Neonate, sacrococcygeal teratoma, surgery.

Sacrococcygeal teratoma (SCT) is an uncommon tumor which develops on the base of the tailbone or coccyx.[1] The incidence in newborns is 1 in 35,000 to 40,000 live births, with a female-to-male ratio of 3:1 to 4:1.[1] This tumor may consist of various types of tissue and lacks a capsule or pseudo-capsule. Most SCTs are benign, and only 1 to 2% are malignant.[2,3] The primary treatment is surgical resection.[3] In this article, we present a patient with SCT in Palestine which was treated with entire resection and flap reconstruction.

CASE REPORT

A male infant weighing 2.8 kg with a large bulk at the base of the coccyx was born to a 23-year-old mother at 36 weeks of gestational age via cesarean section delivery due to the mass. There was no known medical history of genetic disorders, substance misuse, teratoma, or congenital defects. During the second trimester of pregnancy, ultrasound revealed a highly vascular sacrococcygeal area measuring 10×11 cm (Figure 1a). The male infant was delivered by the cesarean section. The Appearance, Pulse, Grimace, Activity and Respiration (APGAR) scores were 9 and 10.

He was immediately admitted to the neonatal intensive care unit (NICU) for further evaluation and surgical evaluation. Feeding was initiated and well tolerated. Preoperative examination included a pediatric cardiac assessment and echocardiography which revealed a normal foramen ovale and no signs of pulmonary artery hypertension. The alpha-fetoprotein (AFP) level was 180,000 (range: 87 ng/mL). Other laboratory test results

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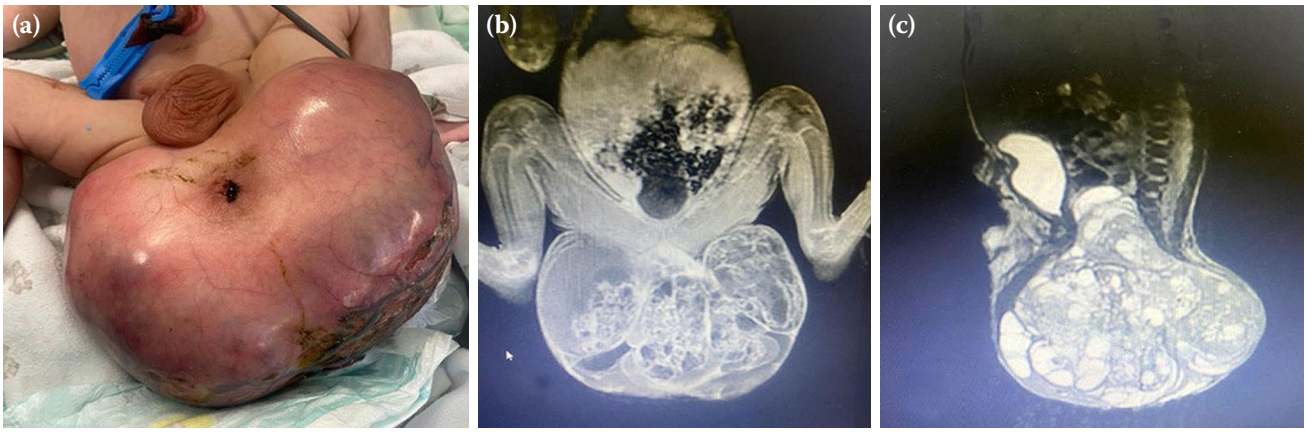

Figure 1. **(a)** Fetus with a huge sacrococcygeal teratoma. **(b, c)** Anomaly scan, massive multi-lobulated heterogeneously enhancing mass in the sacrococcygeal region measuring 15×9.5×11 cm.

including liver functions, kidney functions, complete blood cell count, and clotting status were normal.

Magnetic resonance imaging (MRI) was performed on the third day of delivery and revealed a massive multi-lobulated heterogeneously enhancing mass in the sacrococcygeal region measuring 15×9.5×11 cm in size. The lesion extended into the pelvic cavity, although there was no invasion in the surrounding regions (Figures. 1b, c).

The resection and flap reconstruction operation were completed successfully on the fourth day of life (Figure 2a). The excision of the mass was done under general anesthesia, with the infant in a prone position through a chevron incision. Intraoperatively, a Hegar dilator was placed inside the rectum as a guide, there was no rectal invasion or intraoperative rupture of the teratoma, and no drain was used. Reconstruction of the anal sphincter and perianal muscle was done, and the operation took three hours with continuous measurements of temperature fluid monitoring. The amount of blood loss was 90 mL and additional 50 mL of blood was transfused intraoperatively. Fentanyl and paracetamol were used for pain management.

The histopathological report revealed an immature teratoma, with a smooth, white, tan-cut surface composed of many cysts with serous and mucinous material. The inside of the tumor showed abnormal cells with components of hepatic tissue cells, cartilage, and neuroepithelial cells (Figures 3a, b).

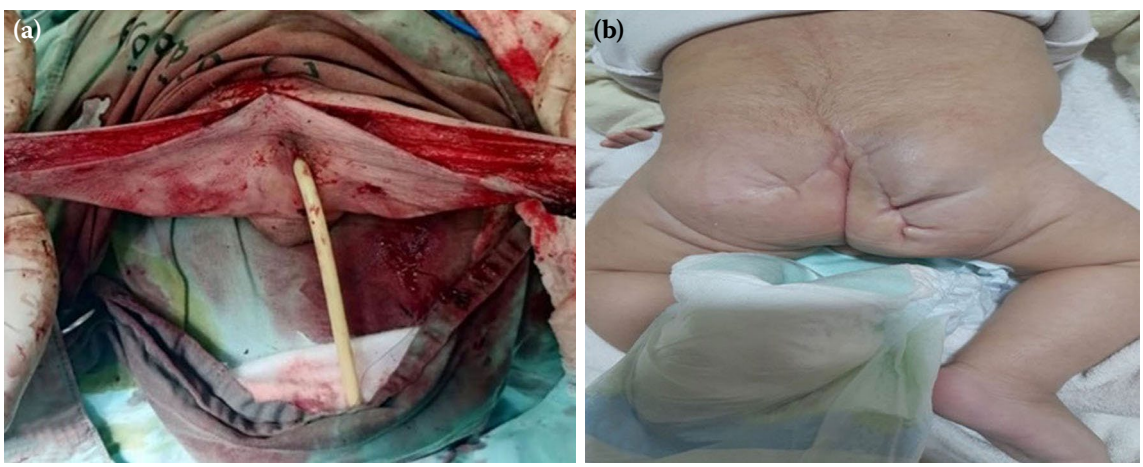

Figure 2. **(a)** Intraoperative view of sacrococcygeal teratoma. **(b)** Postoperative image of the patient.

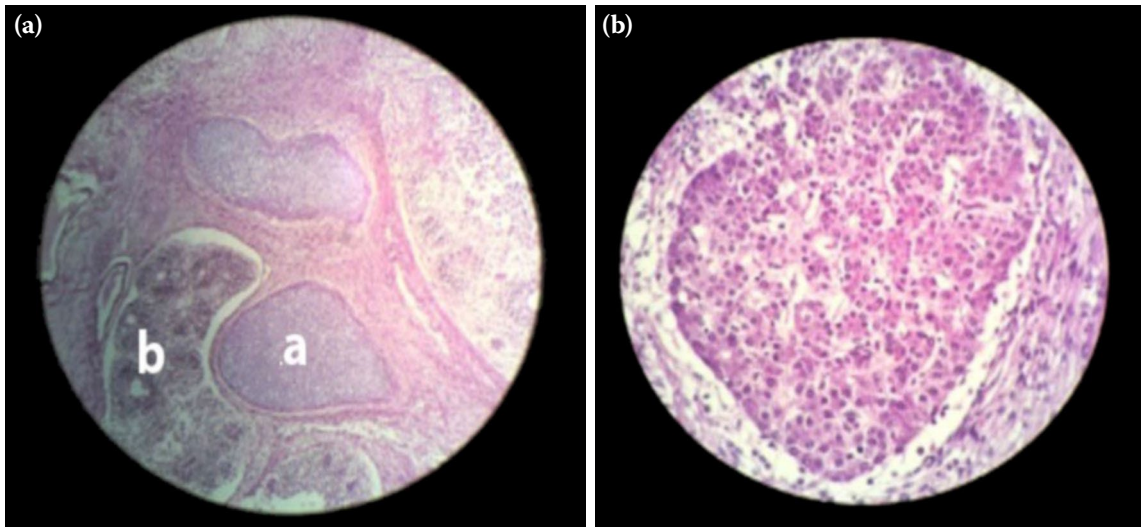

Figure 3. **(a)** Histopathological examination showing circular island of cartilage [a] and neuroepithelial cells [b], **(b)** hepatic tissue cells.

There was no evidence of malignancy or a yolk sac tumor. The AFP readings decreased over time, with 180,000 before surgery, 75,300 after surgery, and 1,110 at six months of age. Other blood tests, such as liver function, kidney function test, complete blood cell count, and clotting status continued to be normal.

Following the operation, the infant remained intubated and admitted to the NICU. He was extubated approximately 16 h after the operation. He received respiratory support through continuous positive airway pressure (CPAP). Regular wound dressing and assessment of the vital signs were stable, and the infant was transferred to the nursery on the postoperative Day 4. He ate well and remained active.

One week later, he developed severe respiratory distress which was diagnosed as sepsis. He was treated, stabilized, and in good general health after four days. Regular dressing changes were recommended and the parents were advised to place the infant in either lateral or prone positions. At one month of follow-up, there was no bowel or bladder incontinence or neurological deficits.

Scars are still require cosmetic procedures at the site of operation in the future (Figure 2b).

A written informed consent was obtained from the parents and/or legal guardians of the patient.

DISCUSSION

The SCT tumor typically appears on the fetus's tailbone or coccyx. The growth can be solid, cystic, or a combination of both.[4] In majority of cases, it does not cause symptoms or blockage in the rectum or bladder.

The case we present involved a male neonate with a mass located in the midline near his coccyx. Of note, SCT is the most frequently recognized fetal neoplasm, more common in females with a male-to-female ratio of 1:4. The tumor may arise from all three germ layers.[3] Most of SCTs (60 to 70%) are benign. The incidence of malignancy increases with age and with the immaturity of the tumor.[5,6] In general, SCTs are categorized as either immature teratomas, which include embryonic components or structures that have not been fully differentiated yet; or mature teratomas, which include fully differentiated bone, teeth, and hair tissue; or malignant teratomas, which include tumors of the yolk sac, choriocarcinoma, and embryonal carcinoma.[7] In our case, there was an immature teratoma.

The Section on Surgery of the American Academy of Pediatrics (AAP) classifies SCT as Grade 1 (47%), developing only outside the fetus, but can have a small pre-sacral component; Grade 2 (35%), present externally, but with significant intrapelvic extension; Grade 3 (10%), mostly a pelvic mass projecting

into the belly, but also visible externally; and Grade 4 (8%), tumor developing entirely inside the fetal pelvis. Our patient had an immature teratoma Grade 2, which is associated with a low risk of developing malignancy. According to Altman types of SCT, it is divided into three types: type 1, external SCT with small internal parts; type 2, dumbbell-shaped tumor with similar internal and external components; type 3, internal SCT and smaller external components; type 4, exclusively internal part.[9] The tumor in our case was considered type 2.

The MRI assists in diagnosis and in differentiating the mass from myelocystocele, myelomeningocele, and all other posterior neural tube masses.[10] In addition, serial ultrasound evaluation is used for early diagnosis and monitoring of the mass during the pregnancy and for detecting complications that may occur, such as cardiomegaly, hydrops fetalis, and fetal demise.[11]

Birth complications may arise when sacrococcygeal tumors are large, as they are more likely to rupture, cause bleeding, and lead to dystocia during birth.[12-14] Since the mass in our case was identified during the second trimester, the obstetrician decided to perform a cesarean delivery to prevent potential complications.

The main treatment for SCTs is complete surgical resection of the tumor and the coccyx.[15] Full surgical excision and regular monitoring of AFP are enough in non-malignant cases, followed by beta human chorionic gonadotropin level measurement for three years.[16] In malignant cases, postoperative adjuvant chemotherapy with platinum is considered the ideal way to avoid the 4% SCT recurrence rate.[17] In our patient's case, the ultrasound, MRI, lab test, tumor markers, AFP follow-up, and multidisciplinary approach between family medicine, obstetrics, pediatrics, and pediatric surgery allowed for an excellent neonatal outcome despite the poor socioeconomic state, ongoing conflict, and the compromised health facilities in Palestine.

In conclusion, SCT is a prevalent neoplasm in pediatric patients. As described in our case, detailed prenatal ultrasound imaging, MRI, and AFP levels are crucial in the diagnosis and monitoring of the tumor to choose the mode of delivery and treatment. In a rural area of occupied Palestine, a team approach to comprehensive treatment with regular follow-up was an important component in achieving a satisfactory outcome in the face of many challenges, such as the lack of medical transportation. Medications, poverty and unemployment, a limited healthcare workforce, and a deficiency in specialized care all play a significant role.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

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