Case Report

A rare case of sacroccocygeal teratoma type II with malignant yolk sac component

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ABSTRACT

Sacrococcygeal teratoma (SCT) is one of the most common neoplasms diagnosed prenatally. Obstetric ultrasound plays a role in the diagnosis and management of these tumors during pregnancy. In this case report, we discuss the postnatal outcome and a multidisciplinary approach in a neonate with massive SCT and preterm delivery. A 36-year-old mother with a pregnancy complicated by gestational diabetes and polyhydramnios and a large mass in the sacrococcygeal region delivered a girl child at 35+4 weeks gestation by elective cesarean section following an *in utero* transport to our center. The teratoma, measuring 15×12 cm, was completely excised along with the coccyx and perineum postnatally on day 3 of life. As histopathology revealed a small cluster of pre-malignant yolk sac tissue, the infant is under the care of a hemato-oncologist. Multidisciplinary care including good obstetric practices with serial monitoring with ultrasonography, *in utero* transfer to a center with Level III Neonatal Intensive Care, and pediatric surgery services with good nursing care result in improved outcomes in these complexes prenatally diagnosed tumors.

Key words: *In utero* transfer, Malignant yolk sac tissue, Pediatric surgery, Sacrococcygeal teratoma

Sacrococcygeal teratoma (SCT) is believed to arise early in gestation from the totipotential cells of Hensen's node, a remnant of the primitive streak in the coccygeal region, although the exact embryonic origin is still uncertain. Mature (benign) teratomas are common in neonates and older children. Immature teratomas are usually cystic, while malignant tumours are solid. It is usually diagnosed in the second trimester by routine sonography, which also aids in the evaluation and monitoring of the tumor throughout pregnancy to identify fetuses at increased risk of complications and to plan multidisciplinary treatment or intervention when appropriate [1]. Necessary ethical clearances and informed consent were received and obtained, respectively, before initiating the study from all participants.

CASE REPORT

A 36-year-old G4P2A1L2 mother was transferred to our birth center for advanced neonatal management (*in utero* transfer). Mrs. Y had regular antenatal check-ups, with the pregnancy complicated by gestational diabetes and polyhydramnios detected in the second-trimester scan. Blood sugars were regulated with Metformin therapy. A prenatal scan at 32 weeks revealed a mass lesion, teratoma measuring 11×10 cm, and a repeat scan at 35 weeks showed that it had increased to 15×12 cm. There was no evidence of hydrops in antenatal scans or other congenital anomalies. There was no history of medication other than iron, folic acid, calcium, and metformin during her pregnancy. There was no radiation or toxin exposure. A course of antenatal steroids was administered 24 h before delivery in view of prematurity.

A single, late preterm female baby weighing 4.2 kg was delivered by cesarean section (Fig. 1). At birth, the baby cried spontaneously, activity was good, and hemodynamics were stable. Baby had respiratory distress with a Silverman-Anderson score of 5, for which non-invasive respiratory support was started. Chest X-ray was done and shows features of transient tachypnea of newborn. Gradually, respiratory distress decreased, and support was weaned. The baby had a patent anus with an absent anal wink reflex. The movements and tone of the lower limbs were normal. A large SCT of 15×12×12 cm was seen in the sacrococcygeal region with a lobulated surface, variable consistency, and good vasculature (Fig. 2). There were no other external congenital malformations.

The 2D echo showed a small ductus arteriosus. The cranial ultrasound scan was normal. Abdominal sonography revealed bilateral hydronephrosis with an anterioposterior diameter of 7 mm (mild) and 12 mm (moderate) in the right and left kidneys,

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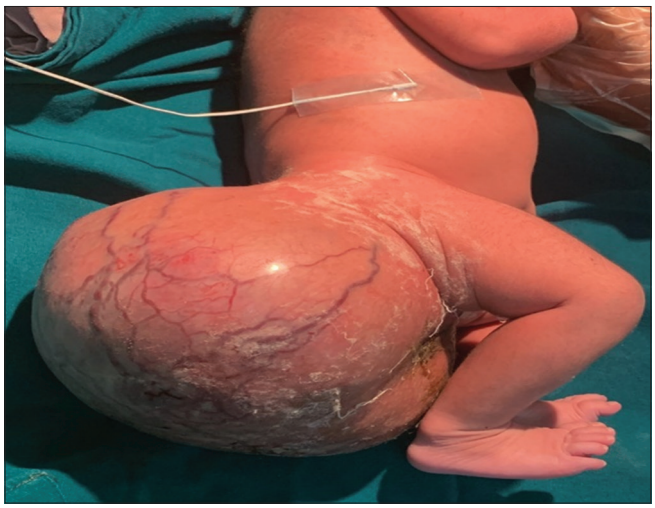

Figure 1: Baby delivered by cesarean section with SCT

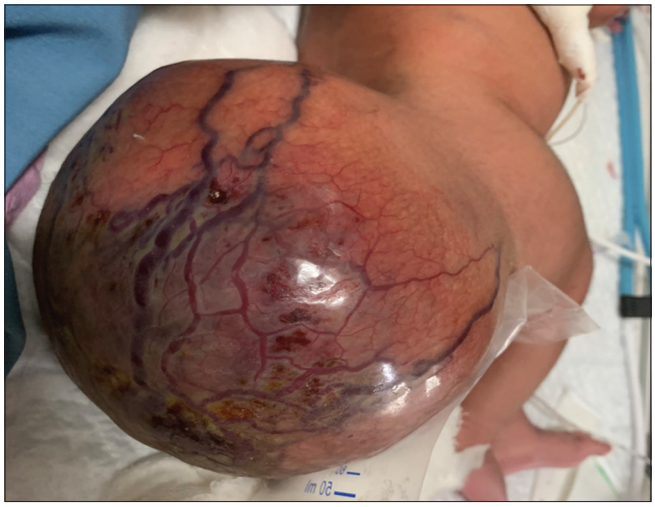

Figure 2: SCT with skin changes on day 2 of life

respectively. Baby received 2 weeks of intravenous antibiotics for culture (*Staphylococcus aureus*) positive bacterial sepsis. Cerebrospinal fluid analysis was normal. Preoperatively, alpha-fetoprotein was grossly elevated to 2000 ng/mL and serum ferritin was 4.8 ng/mL. An ultrasound scan revealed a large and solid cystic mass lesion in the posterior aspect of the SCT region with an exophytic component. Tumour was classified as SCT type II according to the American Academy of Pediatrics Surgical Section (AAPSS) classification.

A complete excision (Chevron incision) of the teratoma with coccyx and perineal reconstruction was performed under general anesthesia at 48 h of life (Figs. 3 and 4). Tumor of size 15 x 13 cms and weight of 960 g was excised (Fig 5). Histopathology of the tumor specimen was suggestive of grade III immature teratoma (25% neuroectodermal elements, a few suspicious foci of malignant yolk sac tumor) (Fig. 6). Wound dressing and nursing were provided regularly in the post-operative period. The baby was discharged on day 15 of life in stable condition. Serial monitoring of alpha-fetoprotein levels was advised. Combination chemotherapy was to be initiated if the titers were rising.

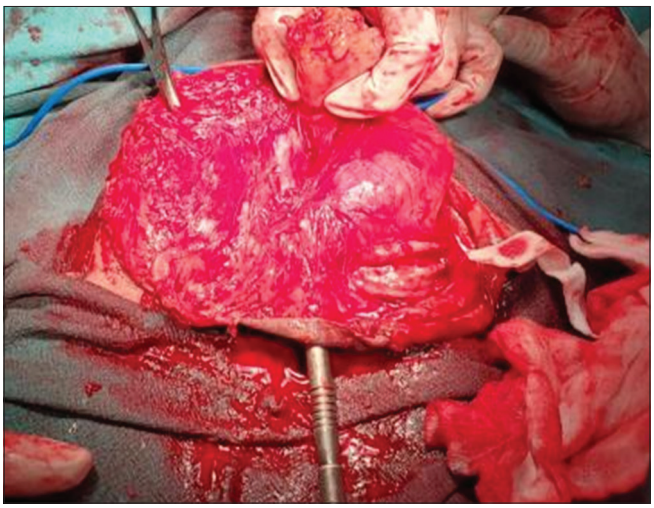

Figure 3: Intraoperative tumour with extension into the pelvis, type II

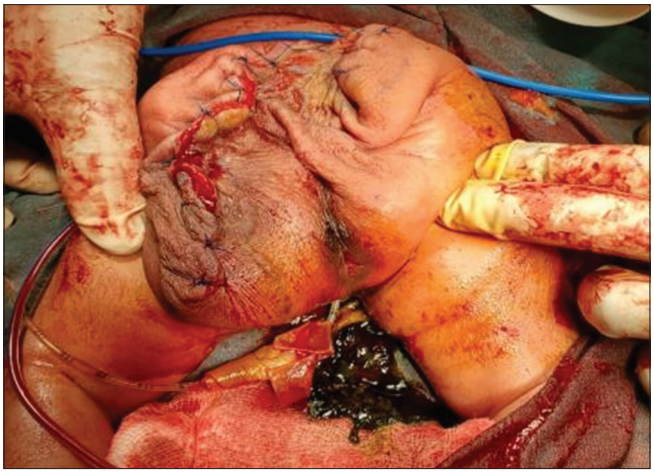

Figure 4: Tumor excised and closing of chevron incision

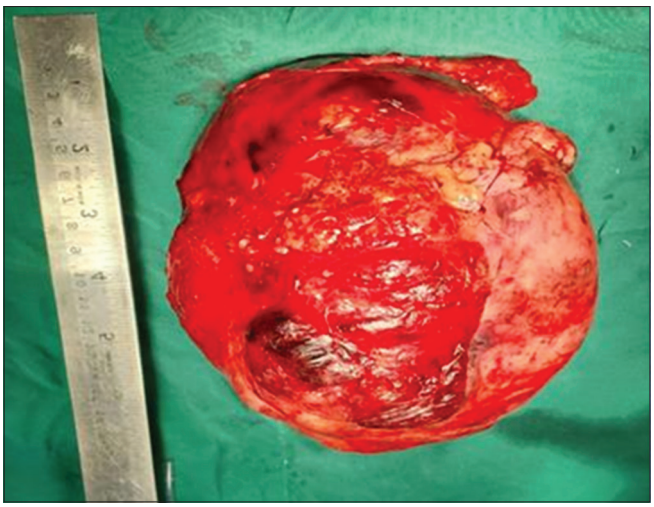

Figure 5: Gross view of a tumor weighing 960 g and measuring 15×13 cm

DISCUSSION

SCT is the most common congenital tumor of the neonatal period, with a reported incidence of 1 in 35,000–1 in 40,000 live

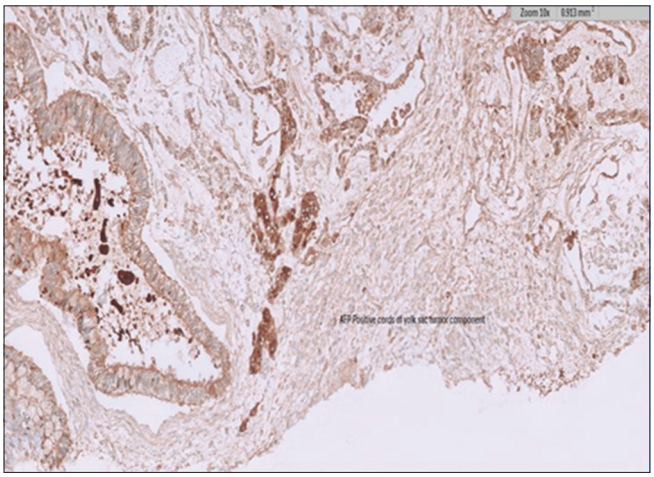

Figure 6: Histopathological examination shows AFP positive cords of yolk sac tumor

births, and approximately, 80% of affected infants are female. This makes SCTs one of the most frequently diagnosed prenatal neoplasms [2,3]. We present a case of SCT diagnosed at 32 weeks of gestational age and associated with maternal gestational diabetes mellitus and polyhydramnios. Serial ultrasound scans showed rapid growth of the tumor with premature termination of pregnancy at 36 weeks for the fetal indication. A successful multidisciplinary approach resulted in good maternal and neonatal outcomes.

SCT is a rare extragonadal, and one of the most common neonatal tumors. It can also occur in infancy or childhood. They can cause rectal or bladder obstruction due to tumor growth or be asymptomatic. Although more commonly reported in female infants, tumors in males result in malignant degeneration [4,5]. Various studies report a female-to-male ratio of 3–4:1, but the exact reason for female predominance is not fully understood [6,7]. The case we have presented here is that of a female baby.

Based on the American Academy of Pediatrics Surgical Section survey (1962–1972), Altman *et al*., in 1973, classified SCT into four types based on external components and intrapelvic/intra-abdominal extent of the tumor [8]. SCTs seen at birth are usually Altman types I and II and rarely type III, but type IV is typically seen later in life. Prenatal diagnosis of SCT Altman types I and II, and even large types III and IV, is usually made by prenatal ultrasonography at 24–34 weeks of gestation [9]. The presence of a heterogeneous and well-circumscribed exophytic mass at the caudal end of the fetus is pathognomonic. The presence of placentomegaly, cardiomegaly, or non-immune fetal hydrops indicates a poor outcome. SCTs can grow rapidly in the fetus and require very high blood flow, leading to fetal heart failure, a condition known as hydrops (15%). If neglected, hydrops can also be dangerous for the mother, leading to similar symptoms of swelling, hypertension, and pulmonary edema with shortness of breath, also known as "mirror syndrome." In our case, there were maternal polyhydramnios and bilateral fetal hydronephrosis, but no evidence of hydrops.

The tumor was classified as SCT type II. Alpha-fetoprotein levels and close prenatal monitoring are paramount to looking for complications. In our case, pre-operative alpha-fetoprotein was over 2000 ng/mL and serum ferritin was 4.8 ng/mL. Recently, the prenatal diagnosis of SCT has increased, and there have been reports of the feasibility of the fetal intervention in selected cases [10].

Cesarean delivery is recommended for a mother whose fetuses harbor large SCTs (>10 cm in diameter) with high vascularity. Altman *et al*. defined the sizes as small (2–5 cm), medium (5–10 cm), and large (>10 cm) in diameter and also classified them based on the extent of the tumor. The recurrence rate without removal of the coccyx is up to 37%. Therefore, complete surgical removal of the tumor with a coccyxectomy is paramount to prevent a recurrence. Complications include wound infections, neuropathic bladder, bowel incontinence, constipation, and diarrhea. Sepsis due to wound infection is the leading cause of death in the early post-operative period. Poor cosmetic results in the buttocks are the most common long-term complication [11].

Malignancy is more common in large SCTs and is associated with Grade II or IV tumors. Although our case was classified as a type II SCT according to the AAPSS classification, histopathological examination revealed an immature grade III teratoma with a few suspected foci of malignant yolk sac tumor.

An excellent survival rate has been reported in the literature; however, the mortality rate for a mass >10 cm is 18% [8,9]. A close follow-up every 3–6 months, with physical examination including rectal examination, serum alpha-fetoprotein, and diagnostic imaging, is recommended for at least 3 years, because the probability of recurrence varies from 11% to 22%, even in completely excised mature neonatal SCTs [12,13].

CONCLUSION

Multidisciplinary care including good obstetric practices with prenatal serial ultrasonography to monitor tumor size and associated perinatal complications, *in utero* transfer to a center with a Level III neonatal intensive care, and pediatric surgery services with good nursing care led to improved neonatal outcomes. A close follow-up with a rectal examination, serum alpha-fetoprotein, and diagnostic imaging is recommended for 3 years to prevent recurrences. There is a need to evaluate the feasibility of fetal intervention for tumor resection or radiofrequency ablation.

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