Prenatal diagnosis of type I sacrococcygeal teratoma and its management

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Summary

Purpose: To report a case of type I sacrococcygeal teratoma (SCT) diagnosed prenatally and managed surgically successfully in the neonatal period. *Case report:* A gravida 2, para 1, woman at 32 week's gestation was referred for suspected fetal anomaly. On US a 14 x 12 cm mass with solid and cystic components was detected in the sacral region of the fetus. On MRI the tumor had no apparent intrapelvic or intraabdominal extent, indicating type I SCT. Cesarean section was performed at 34 weeks' gestation due to signs of deteriorating high output cardiac compromise in the fetus. In the neonatal period stabilization of the infant was achieved. At age ten days the mass was successfully excised surgically. *Conclusion:* Prenatal determination of SCT, follow-up with sonography, time, and mode of delivery are indicative factors for prognosis in SCT.

Key words: Doppler studies; Magnetic resonance imaging; Prenatal diagnosis; Sacrococcygeal teratoma.

Introduction

Sacrococcygeal teratomas (SCT) have an incidence of 35,000 to 40,000 live births and are therefore the most common tumors in newborns [1]. The male to female ratio is one to four [1]. The course of SCT's are ultimately associated with prenatal and perinatal complications. The majority of SCTs have low morbidity and mortality when diagnosed prenatally and managed during the neonatal period [1]. However, a certain subgroup with a large, predominantly solid and highly vascular formation has a poor prognosis due to high output cardiac failure, fetal hydrops and intrauterine fetal demise [1].

Successful management of SCT cases depends on an accurate prenatal diagnosis and classification of the tumor, time of delivery, and mode of delivery. According to the literature, when SCT is diagnosed earlier than 30 week's gestation its prognosis is generally poor [2].

We report a case of type I SCT diagnosed prenatally at 32 week's gestation by (2D) two dimensional-ultrasound (US) and fetal magnetic resonance imaging (MRI) evaluations. The patient was successfully managed surgically and during the neonatal period.

Case Report

A 25-year-old woman, gravida 2, para 1, was referred to our clinic at 32 weeks of gestation for suspected fetal anomaly, polyhydramnios and preterm labor. On initial transabdominal 2D-US examination we detected a mass with solid and cystic components measuring 14 x 12 cm in the sacral region of the fetus (Figures 1 and 2). There was no evidence of fetal hydrops, placentomegaly, or polyhydramnios. We confirmed the gestational age with breech presentation consistent with 32 weeks of gestation and observed normal female genitalia. Thus we made a presumptive diagnosis of SCT.

Further detailed investigations were performed to establish a definite diagnosis. Color Doppler sonography showed that the mass was well vascularized, with normal fetal vascular flow. Fetal MRI revealed a clear presentation of the tumor (17 x 12 cm in diameter) and its relationship to the sacrum which was without apparent intrapelvic or intraabdominal extent indicating type I SCT (Figure 3).

Serial sonograms done by her previous doctor up to 30 weeks' gestation demonstrated no anomalies. Furthermore, α-fetoprotein and F-βhCG testing at $17^{4/7}$ weeks' gestation revealed a low risk of Down's syndrome, open neural tube defects and Trisomy 18 (1.30 multiples of median (MOM) and 1.45 MOM, respectively). The patient's history was uneventful except for the Rh incompatibility.

The patient was hospitalized and monitored with serial US, fetal Doppler studies and fetal echocardiograms, and external fetal monitoring. To prevent preterm labor and enable lung maturity, sedation and hydration were done and a course of betamethasone was administered to the patient. On serial US at $33^{4/7}$ weeks' gestation signs of placentomegaly and polyhydramnios were detected. Furthermore, fetal echocardiography and Doppler studies revealed dilated cardiac ventricles and a dilated inferior vena cava. Tricuspid valve regurgitation was detected on fetal echocardiography (Figure 4). To reverse polyhydramnios one liter of amniotic fluid was aspirated and the fluid was checked for fetal lung maturity. After the amniodrainage Rh prophylaxis was carried out for the protection of the mother against Rh isoimmunization.

Since the fetus was showing signs of deteriorating high output cardiac compromise and the L/S ratio indicated a mature lung profile, we decided to perform a cesarean section. Cesarean section was done with midline abdominal and vertical uterine incisions to minimize trauma to the SCT during delivery. A female infant with a 17 x 10 x 14 cm SCT was delivered

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Fig. 1

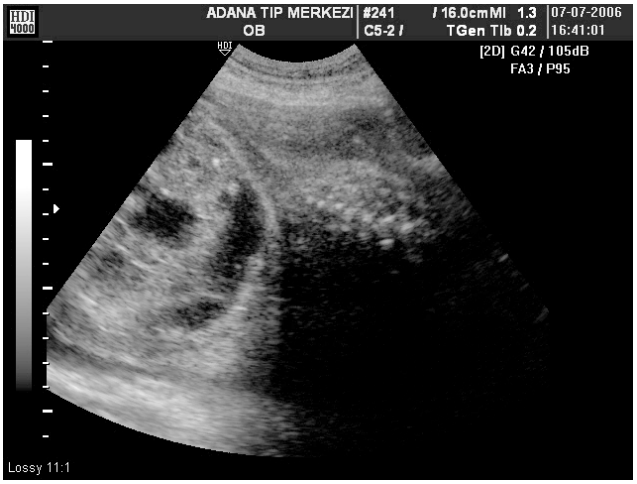

Fig. 2

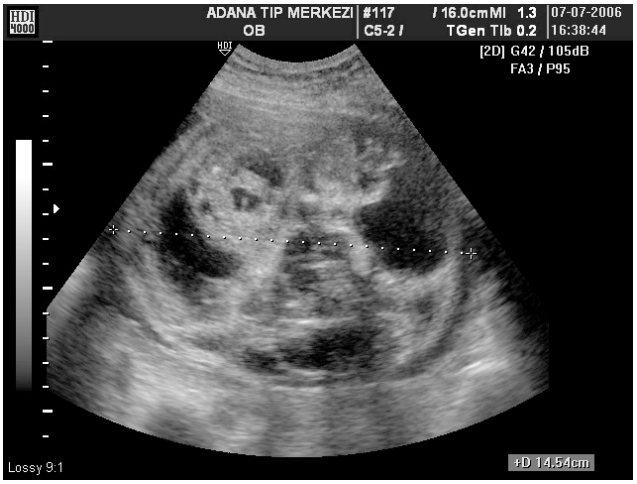

Fig. 3

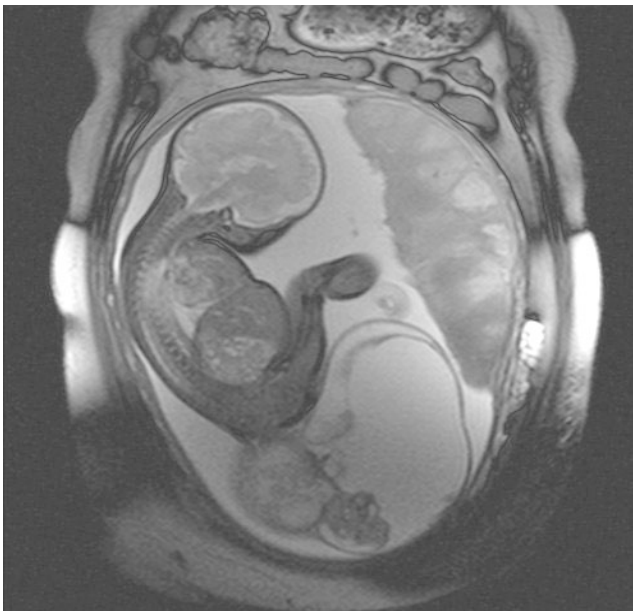

Fig. 4

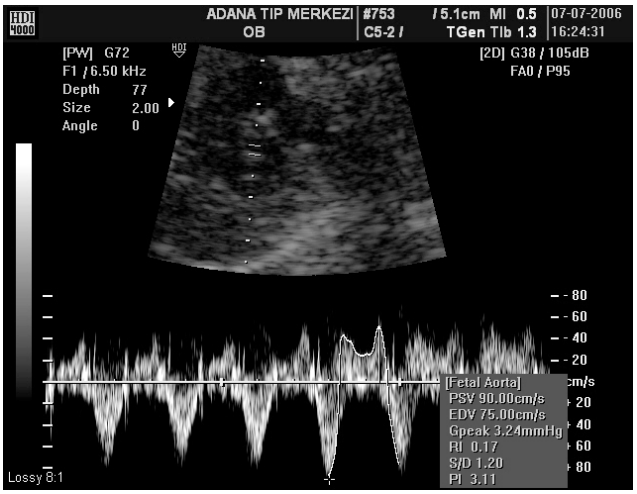

Fig. 5

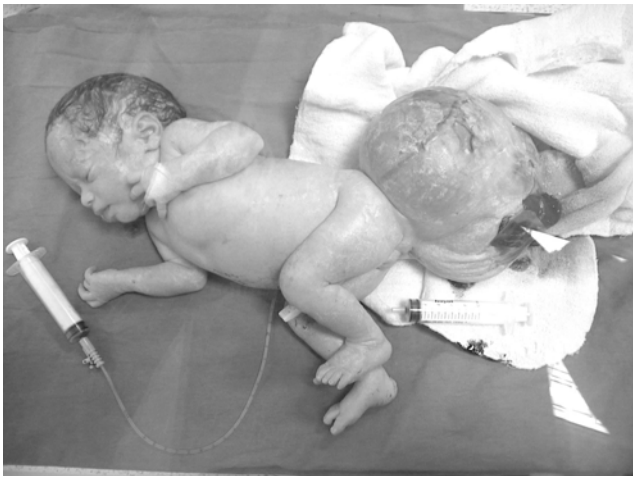

Figure 1. — Midline sagittal view of the fetus showing a sacrococcygeal teratoma.

Figure 2. — Axial view of the fetus demonstrating a sacrococcygeal teratoma.

Figure 3. — Prenatal MRI of the SCT.

Figure 4. — Severe tricuspid regurgitation in the 34-week fetus with SCT.

Figure 5. — Postnatal view of the female fetus with the SCT.

with Apgar scores of 4 and 6, at 1 and 5 min, respectively (Figure 5). The combined weight of the infant and the tumor was 3100 g. Maternal postoperative recovery was uncomplicated. At age ten days, the female infant underwent excision of SCT.

The pathologic examination confirmed this mass as a mature cystic teratoma. The postoperative course was uneventful.

Discussion

Sacrococcygeal teratoma is a germ cell tumor, comprising elements from all three germinal layers; endoderm, mesoderm, and ectoderm [3]. The majority of these tumors are usually fully differentiated, mature and benign, as in our case [3]. However, 4%-20% have malignant tissue at delivery [3].

The American Academy of Pediatrics Surgical Section has classified SCT according to the amount of presacral and external tumor present, stage and timing of diagnosis, ease of resection and malignant potential based on MRI findings: Type I (47%) is predominantly external, with a minimal presacral component as in the case presented; Type II (35%) is external with significant intrapelvic component; Type III (8%) is apparently external but predominantly internal; Type IV (10%) is completely internal [4].

US has proven to be an important tool in the diagnosis and surveillance of fetuses with SCT. Morover, US can also aid in determing the timing and mode of delivery.

MRI is a valuable adjunct to US in providing crucial information for the definitive diagnosis. It provides better visualization of the skeleton and pelvic bones and better contrast between the cystic and solid components of teratomas, and delineates the intrapelvic extent [4]. In the presented case, MRI was instrumental in counseling the parents and in describing the lesion to them.

Color Doppler can identify areas of increased blood flow through the tumor mass [5]. Serial evaluation of fetal Doppler velocimetry studies, especially of the descending aorta and inferior vena cava is useful to search for signs of high output cardiac failure. In the present case the signs of deteriorating high output cardiac compromise detected on serial Doppler evaluation were one of the reasons for an urgent delivery.

In SCT cases alfa fetoprotein (AFP) levels may be elevated during the second trimester. However, in the literature maternal serum screening and amniotic fluid markers result in inconsistent findings in pregnancies complicated by SCT [6]. The use of AFP as a tumor marker in the follow-up is well established, and persistently elevated levels may indicate a residual tumor or recurrence as well as malignant degeneration [7, 8]. Triple test results were in normal range in our case.

Cases diagnosed after 30 weeks of gestation generally have a good prognosis. However, earlier detection of tumor prior to 30 week's points out early occurrence and a rapid growing pattern of the tumor which causes arterio-venous communications that subsequently develop high output congestive heart failure, placentomegaly, polyhydramnios, fetal hydrops, and fetal demise [9]. The prognosis of SCT ultimately depends on the gestational age at diagnosis.

Planned delivery and postnatal surgery is the management of SCT especially after 30 weeks of gestation. Timing and mode of delivery is essential to avoid complications during delivery such as prematurity, tumor rupture and dystocia. Prognosis is best in the cases diagnosed after 30 weeks' gestation without complications like congestive heart failure, placentomegaly, and polyhydramnios. If any of these complications are present on the following serial US examinations urgent delivery is necessary. In our case we detected these signs except for the fetal hydrops on serial US examinations.

Midline transabdominal and vertical uterine incisions should be performed since they minimize the trauma to SCT. After delivery, the tumor should be resected as soon as possible to avoid malignant degeneration, which we did in our case [10].

In conclusion, the indicative factors for improving the prognosis in SCT cases are accurate prenatal determination of SCT with US and MRI, close follow-up with sonographic and Doppler evaluations of the possible complications, and the mode and time of delivery.

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