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Case Report

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Prenatal diagnosis and follow-up of giant sacrococcygeal teratoma: a case report

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Abstract

Objective: To emphasize the favorable effect of elective and planned delivery of newborns with giant sacrococcygeal teratomas on neonatal outcomes.

Case: A 26-year-old, primigravid woman was admitted to hospital at 39 weeks of gestation because of severe polyhydramnios and a giant solid-cystic mass in the sacrococcygeal area of the fetus. A healthy newborn with a giant sacrococcygeal teratoma (25×25×20 cm) was delivered by cesarean section in elective conditions. On the second postnatal day, total excision of the mass was performed by pediatric surgeons. Histopathologic examination revealed mature cystic sacrococcygeal teratoma. The newborn was discharged with a well-developed scar tissue on 15th postnatal day.

Conclusion: Sacrococcygeal teratomas diagnosed in the prenatal period can reach very large sizes. Especially those without heart failure can reach up to the term. The planning of elective delivery in these cases is of vital importance in terms of reducing perinatal mortality and morbidity.

Keywords: Fetus, sacrococcygeal teratoma, ultrasonography.

Özet: Büyük sakrokoksigeal teratomun prenatal tanısı ve takibi: olgu sunumu

Amaç: Büyük sakrokoksigeal teratomlu yenidoğanların elektif ve planlı doğumlarının neonatal sonuçlar üzerindeki olumlu etkisini vurgulamak.

Olgu: Yirmi altı yaşında primigravid kadın olgumuz, şiddetli polihidramniyos ve fetüsteki büyük katı kistik kitle nedeniyle gebeliğin 39. haftasında hastaneye başvurdu. Büyük sakrokoksigeal teratomu (25×25×20 cm) olan sağlıklı yenidoğan, elektif koşullarda sezaryen doğum ile dünyaya getirildi. İkinci postnatal günde, pediyatrik cerrahlar tarafından kitlenin total eksizyonu gerçekleştirildi. Histopatolojik muayenede, matür kistik sakrokoksigeal teratom saptandı. Yenidoğan, 15. postnatal günde iyileşmiş yara dokusuyla taburcu edildi.

Sonuç: Prenatal dönemde tanı alan sakrokoksigeal teratomlar oldukça büyük boyutlara ulaşabilir. Özellikle kalp yetmezliği olmayanlar miada ulaşabilir. Bu olgularda elektif doğum planlaması, perinatal mortalite ve morbiditeyi azaltmak bakımından hayati öneme sahiptir.

Anahtar sözcükler: Fetüs, sakrokoksigeal teratom, ultrasonografi.

Introduction

Although sacrococcygeal teratoma (SCT) is a rare tumor, it is an important cause of perinatal/postnatal mortality and morbidity. A multidisciplinary approach to determine the optimal time for surgical resection, to plan the mode of delivery, and to provide postnatal care and follow-up would minimize all these possible risks.[1]

We aimed to present a case in which successful postnatal management of the fetus with a prenatally-diagnosed giant SCT was achieved by immediate postnatal excision of the mass.

Case Report

A 26-year-old primigravid woman was referred to our hospital from another healthcare center due to severe

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polyhydramnios and a giant mass of the fetus which would possibly need pediatric surgery. The solid-cystic exogenous mass measured 25×25×20 cm by ultrasound in the sacrococcygeal region (**Fig. 1**). The columna vertebralis was intact. Amniotic fluid index was measured as 35 cm and placentomegaly was noted. Arteriovenous shunting in the mass, fetal hydrops and fetal anemia were not detected as median cerebral artery (MCA) Doppler flow indices were within normal ranges (0.96 Mom). Fetal echocardiographic examination was normal and no other associated malformation was present. Fetal MRI was not planned because of the advanced gestational week. On the second day of admission, as the gestation was 39 weeks, cesarean section was performed because of the tumor size. 3900 g girl baby was delivered with an Apgar score of 8 at 1 minute and 9 at 5 minutes. A skin-covered giant mass with a diameter of 28 cm located at the sacrococcygeal region was observed at postpartum examination of the newborn (**Fig. 2**). Alpha-feto protein (AFP), beta-human choriogonadotropic hormone and carsinoembryogenic antigen levels were measured as 37,786 IU/ mL, 30 mIU/mL and 3.2 ng/mL, respectively. Postnatal hemoglobin levels were 15.9 g/dl. Resection of the mass was planned as soon as possible before heart failure developed. After neonatal umbilical vein and radial artery catheterization, total excision of the mass, along with coccyx, was achieved without rupture, although it was strictly adherent to the rectum and had presacral involvement. The mass weighted 1900 g. During the 4-hour-operation, 20 ml/kg/h IV fluid and 15 ml/kg/h erythrocyte suspension were given to the neonate who, after the operation, was transferred to the neonatal intensive care unit with mechanic ventilation. Also with postoperative albumin, fresh frozen plasma and erythrocyte transfusions, the neonate went well hemodynamically at the early postoperative period. The neonate was extubated at 12th hour and discharged on the 15th day with a well-developed scar tissue overlying the operated sacral area.

Microscopically, the solid components were composed of a mixture of mature tissues including pancreatic tissue (**Fig. 3a**), peripheral nervous tissue (**Fig. 3b**), and stratified squamous epithelium (**Fig. 3c**). Based on these histopathological features the diagnosis was mature cystic SCT. Levels of highly elevated tumor markers in the preoperative period were decreased to normal levels in the 3rd month postoperatively. The follow-up visits were problem-free for 9 months as this paper was written.

Discussion

In the era of routine prenatal screening and improved fetal imaging using ultrasonography (USG) and magnetic resonance imaging (MRI), most SCTs are now diagnosed in utero.[2] Close surveillance with serial ultrasonographic imaging and echocardiography is recommended

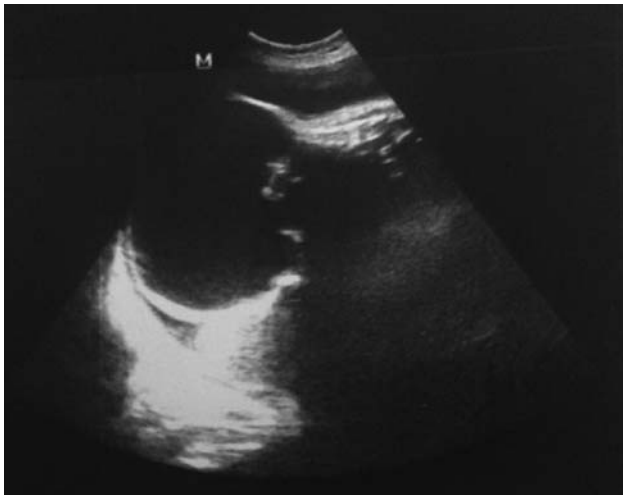

Fig. 1. Mass on ultrasound. The solid-cystic exogenous mass was measured 25×25×20 cm.

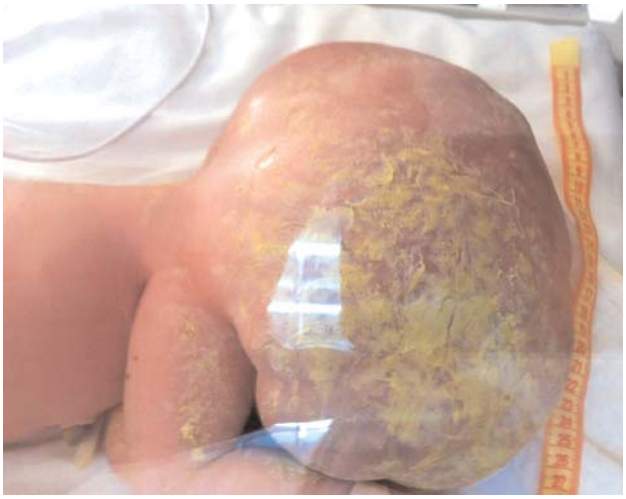

Fig. 2. A giant mass with a diameter of 28 cm located at the sacrococcygeal region.

so that fetal intervention or early delivery can be performed when necessary.[2] There are some prognostic factors in the SCTs. One of them is tumor size. The tumor size being greater than 10 cm is especially high in vascular tumors, and these tumors, along with heart failure, increase the risk of fetal hydrops and intrauterine fetal demise.[2] Although the diameter of the tumor was 28 cm in our case, we did not detect any sign of heart failure. Gestational age is other prognostic factor. While the prognosis is poor among infants diagnosed before the 30th gestational week, the survival rate is very high among infants born just before term.[3] The reason why such a large mass reaches to term without any complications is due to the absence of heart failure and additional organ anomalies, not ideal prenatal management. Anatomical location is important, too. Altman et al. described a four–stage- classification system of SCTs according to their anatomical location which appears to be associated with overall prognosis, with best survival being in Type 1,[4] as in our case. The incidence of various congenital malformations associated with SCTs range from 5% to 26%. Of these, anorectal and genital malformations are of prime concern.[5] Other associated anomalies include spinal dysraphism, sacral agenesis, dislocation of the hips caused by a large tumor, and meningocele.[6] Sivrikoz et al. presented an atypical presentation of a tethered spinal cord, associated with a SCT.[7] Gothwal et al. reported a SCT associated with Prune Belly syndrome.[8] Perrone et al. presented a SCT case with a large intraspinal component that was causing compression of the lower spinal cord.[9] Due to the large tumor size, detailed examination and anatomical scanning with USG may not be possible in every case. Krekora et al. reported agenesis of the right forearm of a newborn, which had not been detected prenatally, despite many examinations.[10]

In our case we performed prenatal and postnatal USG and postnatal MRI for detecting associated congenital malformations. No other malformations were detected.

The tumor size and location are essential for planning the mode of delivery in these cases. In the absence of obstetrical indications, vaginal delivery is acceptable for small tumors. Elective cesarean delivery is recommended for SCTs measuring more than 5 cm in diameter due to the risk for traumatic injury, rupture and subsequent hemorrhage from the tumor.[11] Hemorrhage is

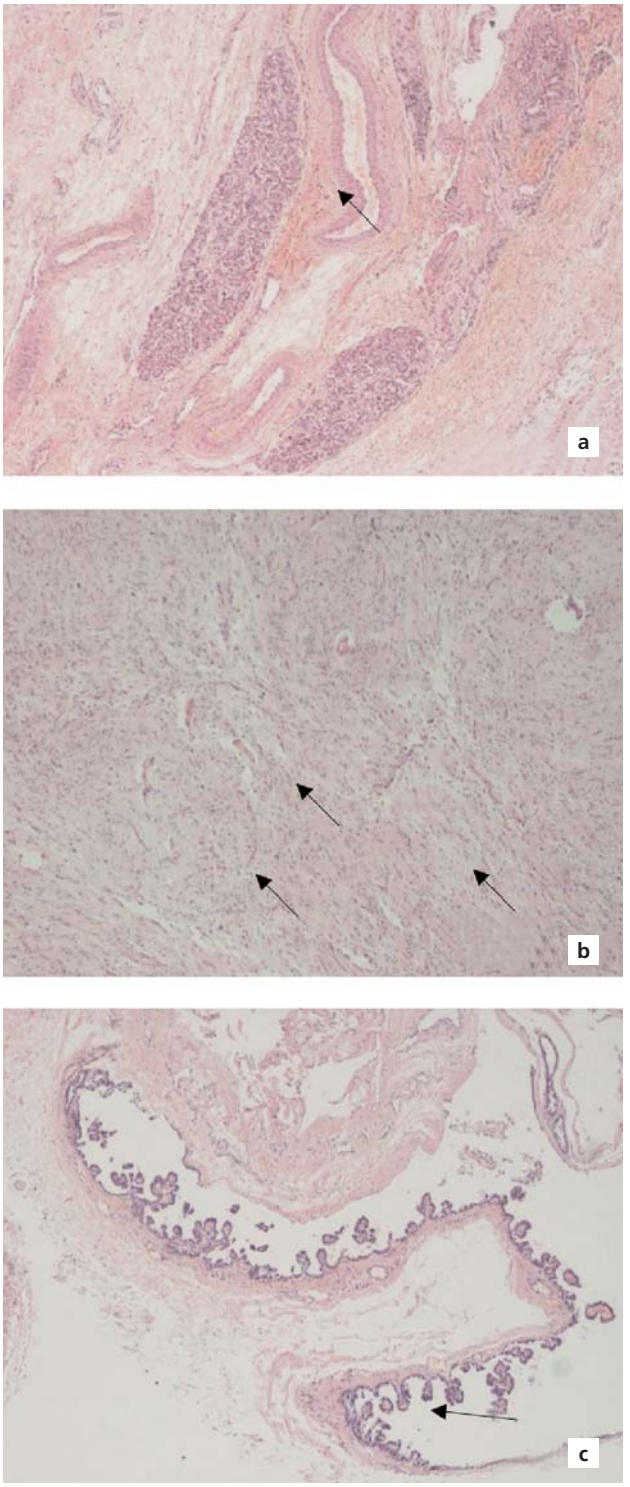

Fig. 3. Histopathological features. Microscopic view of sacrococcygeal teratoma at 400 magnifications with H&E stain. Pancreatic tissue (**a**), peripheral nerve tissue (**b**), and stratum squamous epithelium (**c**) are indicated with **arrows**, respectively.

the most common cause of neonatal mortality in patients with SCT. High-output cardiac failure, intratumoral hemorrhage and perioperative bleeding are the most common causes of early death and are all strongly associated with larger tumor sizes.[12] Tumor rupture may be caused by uncontrolled labor or complications during delivery.[13] Alani reported a ruptured giant SCT during cesarean section and its successful postnatal management.[14] Advanced gestational age may be related the resistance of the rupture in a SCT. According to literature, upper vertical incision is usually preferred during caesarian section,[15] as in our case. However, lower segment incision was performed in some patients because of the increased risk of bleeding and uterine scar rupture at the following pregnancies.[1] Maturation of a SCT is related to fetal/neonatal prognosis. Mature teratomas are the most common[16] and usually have excellent prognosis if completely excised. The surgical approach involves complete excision of the tumor including the coccyx in order to reduce recurrence,[17] as in our case. Timing for tumoral resection in the postpartum period is important. Elective excision of the tumor is usually performed within the first few days of life.[15] As recommended, resection was performed on the second postpartum day in our newborn. SCTs must be followed up with physical examination, USG, and MRI at the postpartum period.

Shortly, prenatal diagnosis has improved the perinatal management of these lesions that might benefit from fetal intervention. A comprehensive prenatal evaluation including conventional USG, Doppler USG, echocardiography and fetal MRI, is essential for adequate counseling and optimal perinatal management. Antenatal counseling helps the parents to better understand the natural history, fetal intervention, and perinatal management of these tumors. Fetal surgical debulking improves survival in cases of SCTs with cardiac decompensation. Additionally, the use of an EXIT procedure reduces the morbidity and mortality in a complicated delivery in cases with cervical and mediastinal-teratomas.[18] Conditions amenable to intrauterine surgical treatment are rare; the mother may consider termination of pregnancy as an option. Fetal treatment can be lifesaving but it carries risks to both the infant (preterm premature rupture of the membranes, preterm delivery) and the mother.[19] One of the prenatal therapeutic options is to occlude the feeding vessels by radiofrequency ablation,[20] but it has not been accepted as a routine approach because of high mortality at term.

Conclusion

The aim of this paper was to present a giant mature SCT managed successfully without any pregnancy complications. A multidisciplinary approach is crucial for SCT cases in both prenatal and postnatal periods.

Conflicts of Interest: No conflicts declared.

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