Multidisciplinary Management of Giant Fetal Sacrococcygeal Teratom

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Sacrococcygeal Teratoma (SCT) is one of the most frequent tumors of the newborn period. SCT is seen in varying sizes, and there is a relationship between the size and the clinical course. Postoperative prognosis of SCT with a low malign potential is good in the newborn period. Prenatal follow-up and postnatal management are rather important.

We described our patient who was diagnosed in the early weeks and followed up till week 37, and then was given birth by caesarean section. The hemodynamic findings were stabilized during the newborn period. the patient was successfully operated by pediatric surgeons to restore her health.

Key Words: Sacrococcygeal teratoma, Multidisciplinary management

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Introduction

Although sacrococcygeal teratoma (SCT) is a rare tumor, it is the most frequent tumor in the neonatal period.[1] This extragonodal germ-cell tumor locates in the sacral region most frequently.[2] Perinatal mortality exceeds 50% when diagnosis is made in the antenatal period.[3] Seventy-five percent of the affected infants are girls. Perinatal morbitidy and mortality rates are high because of fetal cardiac insufficiency, tumor rupture and anemia.[4] There are other abnormalities in patients with SCT with a rate to 15% spina bifida, meningomyelocele, imperforated anus and vagina and uterus duplication are among these.[5] In general, SCT is confused in the intra-uterine period with neurogenic tumors, dermoid cyst, lipoma and angiomas.[6] Particularly MR can be helpful in cases where ultrasonograhy is not sufficient for the diagnosis. Differentiation of SCT and meningomyelocele with the ultrasonogram during of the initial diagnosis, was difficult in our case. The intrauterine fetal MR, which was taken with this reason, was helpful for the diagnosis. In this article, we describe the follow-up of a giant SCT that grew to a size of 28x30 cm together with radiology, pediatric surgery and newborn units throughout the prenatal and postnatal management.

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Case Report

The twenty-six year-old primigravida was referred to our clinic with the pre-diagnosis of fetal anomaly in the twentieth week of her pregnancy. In the obstetric ultrasonogram of the patient who was in the twentieth week of her pregnancy according to the date of her last menstruation, it was seen that the fetal biometric measures were consistent with the week of the pregnancy. In the fetal sacrococcygeal region, a multilocular formation was observed with a size of 3x4 cm and cystic components predominantly. No other structural fetal anomalies were observed. Normal fetal caryotype was confirmed with amniocentesis. Although SCT was thought primarily, it was not possible to differentiate this diagnosis from meningomyelocele. Fetal MR was performed. MR findings confirmed our diagnosis that was favoring SCT. (Figure 1)

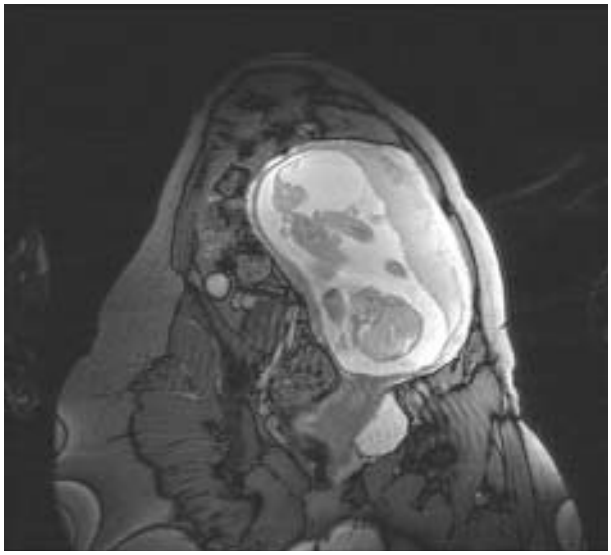

Figure 1: Intrauterine fetal MR

The required information was given to the patient. She was reported in the weekly meeting of the ethical committee that prenatal diagnoses and abnormalities were discussed. It was decided that the anomaly was SCT and pregnancy would be let to continue and followed up by the relevant departments in common after obtaining the opinions of the pediatric surgeons and newborn specialists in the multidisciplinary meeting. She was followed ultrasonographically with regular intervals. Ascites or hydrothorax, which are among fetal hydropsis and congestive heart failure were not seen. Thickness of the placenta did not change. However, rapid enlargement of the mass was observed, especially after the week 32.Upon starting of labor pain in the patient who was followed till the Week 37, decision for labor was made after informing the pediatric and pediatric surgery departments. For the patient that SCT had a size of about 28x30 cm in the latest obstetric ultrasonogram, caesarean section was planned to prevent rupture of the tumor.

A girl infant was given birth (Figure 2) with an Apgar score 7 in the first minute and 8 in the eighth minute. The tests were performed in the newborn department and she was operated by the pediatric surgeons (Figure 3) in postnatal day 2 after respiratory and hemodynamic stability were ensured. It was seen that 90% of the intraoperative mass lesion consisted of the cystic component. Approximately 1500 cc fluid was aspirated. It was seen that the infant weighed 2450g right after the surgery. Mother was discharged in postoperative day 3, and the infant in postoperative day 6, without any problems.

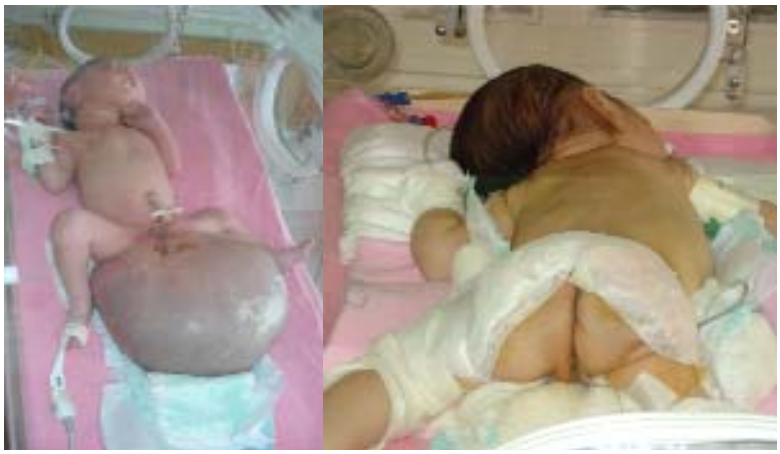

Figure 2: Infant before operation Figure 3: Postoperative view

Discussion

SCT is seen in the newborns with a frequency of 1:35,000-40,000.[7] It is thought that it takes origin from multi-potential cells in the Hensen lymph node.[8] Most of these tumors are histologically benign, and are classified as mature and immature.[9]

Prematurity, intra-uterine heart failure and hydropsis are the main causes for fetal mortality.[10] Fetal hydropsis in SCT develops secondary to a high-output heart failure which is the result of a vascular leakage because of the tumor growing. Again, solid component of the tumor being greater than 10 cm, increased vascularization, fetal cardiomegaly and placentomegaly are responsible for the poor prognosis.[11] Our case was one that was followed up till term with a big cystic component and a good prognosis. Hydropsis or placentomegaly was not observed in our case.

Gestational age is also among the factors that determine the prognosis in babies born with SCT. While the survival rate is 7% before thirtieth gestational week, the same rate is 75% in the following weeks.[12] We therefore waited till the Week 37, when pain started in our patient.

Although making the diagnosis is mostly easy with ultrasonography, it sometimes can be confused with cordoma, fibromas, rectal dublication, meyelomeningocele and presakral neurogenic tumors in the definitive diagnosis. Additional radiological diagnostic methods can be used when such difficulties are encountered.[13] Likewise, intra-uterine fetal MR helped us in this case, which the location was challenging for the diagnosis.

Malign transformation is possible if the surgery is delayed or if inadequate excision is performed. This rate, which is 7 to 10% within the first 2 months, increases to 37% in around the first year of life.[6]

In general, cesarean delivery is recommended for dystocia in tumors larger than 5 cm and the trauma of labor. Teratomas can rupture and cause severe bleeding during a traumatic labor.[14] Although classical vertical incision is recommended to prevent this, lower segment Phannenstiel incision is more beneficial, since the risks of hemorrhage, infection, and scar rupture in the following pregnancies increase in the former technique.[15] We therefore also performed lower segment transverse incision on our patient. Cases that vaginal delivery was achieved with no complications in cases with cystic SCT by prenatal percutaneous needle drainage are present in the literature. [16]

Sudden growth of the tumor was in the last trimester in our case that was diagnosed in an early gestational age. Absence of fetal hydropsis indicated a good prognosis. Delivery was delayed as much as possible to ensure fetal development. Caesarean delivery was performed to prevent massive bleeding related to the rupture of the tumor and dystocia. Pediatric and pediatric surgery teams were ready during the delivery against the possible risk of rupture. Multidisciplinary approach is essential in such risky cases, like in our case.

Dev Fetal Sakrokoksigeal Teratomun Multidisipliner Yönetimi

Sakrokoksigeal Teratom (SCT) yenidoğan döneminin en sık rastlanılan tümörlerinden bir tanesidir. SCT değişik boyutlarda olup büyüklük ile klinik seyir arasında ilişki vardır.Malign potansiyeli düşük olan SCT da yenidoğan döneminde cerrahi sonrası prognoz iyidir.Prenatal takip ve postnatal yönetim oldukça önemlidir.

Biz bu yazıda erken haftalarda tanısı konularak 37. haftaya kadar takip edilip sezaryenle doğumu yaptırılan hastamızı anlattık. Yazımızda yenidoğan bölümünde hemodinamik bulguları stabillenen, pediatrik cerrahlar tarafından başarıyla opere edilip sağlığına kavuşturulan bu olguyu sunduk.

Anahtar Kelimeler: Sakrokoksigeal teratom, Multidisipliner yönetim

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