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Huge fetal sacrococcygeal teratoma: Antenatal and postnatal management

Dev fetal sakrokoksigeal teratom: antenatal ve postnatal yönetim

Kazandı M Akman L Şahin Ç

Ege Üniversitesi Kadın Hastalıkları ve Doğum Anabilim Dalı, izmir, Türkiye

Summary

Sacrococcygeal teratoma (SCT) is one of the most common fetal tumors (1/30000). Ultrasonography is an important method for diagnosis and following the SCT. Pregnancy can be managed by diagnosing the teratoma, monitoring its growth rate and fetal complications with the USG. Some complications such as fetal hydrops, dystocia, hemorrhage and rupture can be seen. In this report, we present a 25 year old woman with 25 weeks gestation according to her last menstrual date. We determined a 5x4 cm of sacrococcygeal teratoma through ultrasonographic examination. Because of the acute enlargement of the sacrococcygeal teratoma, a cesarian section was performed without complication at 36 gestational weeks with no fetal compromise developing such as fetal hydrops or placentomegaly. The fetus underwent surgery in the pediatric surgery. The sacrococcygeal teratoma was removed and the histopathology revealed immature teratoma. Neonates with SCT after exicision require long-term follow up for functional impairment. Management of the sacrococcygeal tumors are required with a multidiciplinal approach by the obstetrician, pediatrist and pediatric surgeon.

Key Words: Fetal sacrococcygeal teratoma, antenatal management.

Özet

Sakrokoksigeal teratom, fetal tümörler arasında yaygın izlenen (1/30000) tümörlerden biridir. Tanı ve takipte ultrasonografi en önemli yöntemdir. Fetal sakrokosigeal teratom tanısı konan gebelikler takip edilebilir. Bu gebeliklerde tümörün büyüme hızı ve fetal komplikasyonlar ultrasonografi ile izlenir. Fetal hidrops, distosi, kanama ve rüptür gibi komplikasyonlar görülebilir. Makalemizde 25 yaşında, 25. gebelik haftasında ultrasonografi ile değerlendirmede 5x4 cm büyüklüğünde fetal sakrokoksigeal teratom saptanan hastamızı sunduk. Tümörün ani hızlı büyümesi nedeniyle, fetal hidrops veya plasentomegali gibi komplikasyonlar gelişmemişken gebeliğin 36 haftasında sezeryan ile komplikasyonsuz olarak doğum gerçekleştirildi. Yenidoğan, pediatrik cerrahi kliniğinde opere edildi. Tümör çıkartıldı ve histopatolojik dğerlendirmesi immatür teratom olarak saptandı. Opere edilen yenidoğanlar uzun dönem fonksiyonel bozulmalar açısından takip edilmelidir. Sakrokoksigeal teratomlu hastaların yönetimi obstetrisyen, pediatrist ve pediatrik cerrahı içeren multidisipliner yaklaşımı gerektirmektedir.

Anahtar Sözcükler: *Fetal sakrokoksigeal teratom, antenatal takip.*

Introduction

Sacrococcygeal teratoma (SCT) is one of the most common fetal tumors (1/30000). Although enlargement may be huge, they are generally benign and can be removed surgically.

Yazışma Adresi: Mert KAZANDI

Dokuz Eylül Üniversitesi Tıp Fakültesi, Kadın Hastalıkları ve Doğum Anabilim Dalı, İzmir, Türkiye

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In addition to differentiated mature cells not belonging to the pelvic region, there can be immature cells. It contains three embryologic layers of endoderm, ectoderm and mesoderm (1). Ultrasonography (USG) is an important method for diagnosis and following the SCT. Pregnancy can be managed by diagnosing the teratoma, monitoring its growth rate and fetal complications with USG (2).

Some complications such as fetal hydrops, dystocia, hemorrhage and rupture can be seen. Fetal death or preterm labor is the result of polyhydramnios,

cardiomegaly and fetal hydrops are due to high output cardiac failure because of increased blood and metabolic necessity of the growing tumor. If this complication is seen before 30 gestational weeks survival rate is 7% (3).

Case

A 25 years old primigravid patient, at 25 weeks was referred to our clinic because of fetal sacral mass. Her medical and family history were insignificant. Laboratory results were normal.

Obstetric USG showed a 5X4 cm mass at the sacrococcygeal region of fetus with cystic and solid components, containing echogenic and acustic regions that can be due to bone. There was no sign of hydrops or polyhydramnios. Fetal echocardiography was normal. MRI showed a 6X4 cm mass having cystic signals at the sacral localization that was in accordance with sacrococcygeal teratoma (Figure-1). The patient received consultation in conjunction with the pediatric surgery clinic, and the diagnosis was type 1 sacrococcygeal teratoma (Table-1). Since there was no evidence of polyhydramnios or/and placentomegaly, after receiving approval of the parents, the pregnancy was allowed to continued until fetal viability. The patient was monitored closely with ultrasonography to determine the size of the mass and fetal hydrops. During follow up, the fetal sacral mass size abruptly enlarged (16X13 cm) (Figure-2) and a cesarean section was performed without complication at 36 gestational weeks while no fetal compromise developed such as fetal hydrops or placentomegaly. Neonate weighted 4100 g. and APGAR score was 1 and 6 (Figure-3). He was referred to pediatric surgery and received surgery on the 3rd day. Histopathological diagnosis was immature teratoma. The neonate was followed up with AFP levels and phsiycal examination..

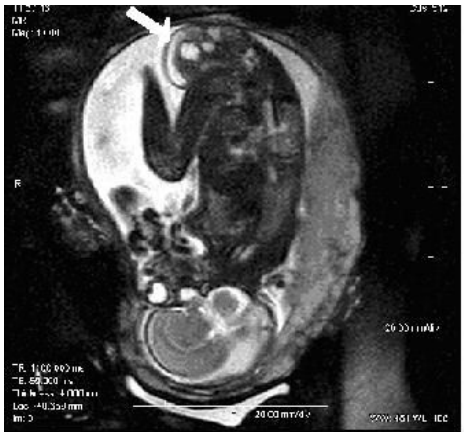

Figure 1. MRI showed a 6X4 cm mass having cystic signals at sacrococcygeal region of the fetus that was in accordance with SCT.

Table 1. American Academy of Pediatrics, Surgical Section classification.

Type I: Tumor is primarily external and has only a minimal presacral component
Type II: Tumor is primarily external but has a significant intrapelvic portion
Type III: Tumor is partially external but is predominantly intrapelvic with abdominal extension.
Type IV: Tumor is located entirely within the pelvis and abdomen

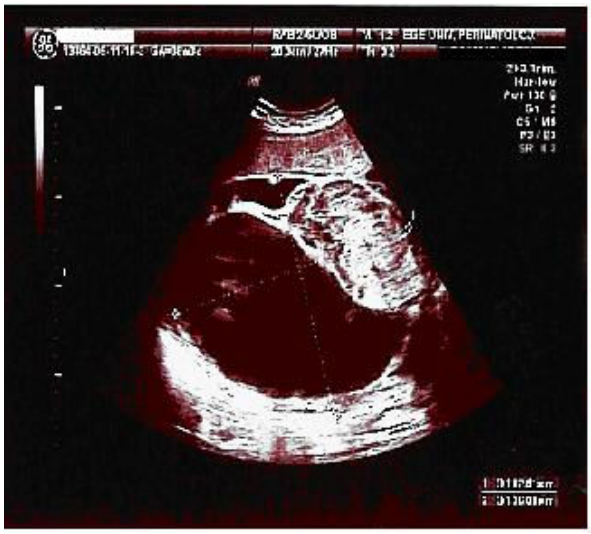

Figure 2. USG showed a enlarged mass (16X13 cm) with cystic and solid components during follow up pregnancy.

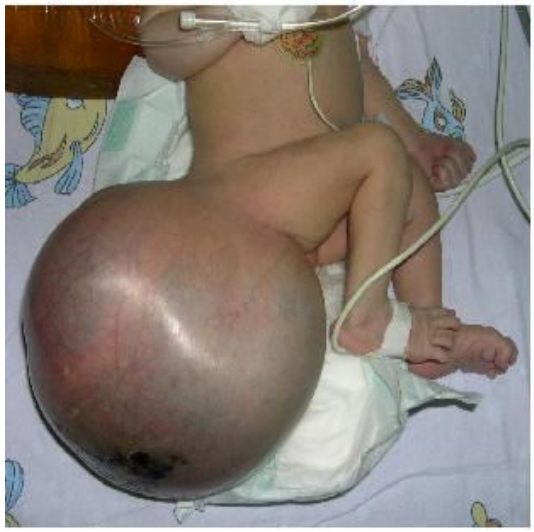

Figure 3. Gross image of the SCT after ceserian section of the neonates.

Discussion

Teratomas are the most common tumors of neonates. Embryologically they originate from pluripotent cells and contain three embryologic layers of endoderm, ectoderm and mesoderm. The distribution of these three layers is explained by cessation and disorder in the migration of primordial germ cells. Generally these mature cells do not belong in these places and frequently originate from neurons. Although teratomas are seen at different parts of the body, over 50% of them are at sacral region in neonates.

Generally diagnosis is made with USG. In a study with a large series and in multiple centers, the average determination time of SCT is 26 gestational weeks (4). Sonographic examination can show cystic and solid components, or either of them can be dominant. Frequently hydramnios is also present. Polyhydramnios is a sign of high output cardiac failure. The enlargement rate of SCT and fetal hydrops signs must be followed with USG. In the cases with dominant solid components an increased vascular structures risk for fetal hydrops development is higher (5). Fetal hydrops develops because of high output cardiac failure due to a shunt between placenta and systemic circulation and it is a poor prognostic factor. Fetal hydrops and its early signs such as polyhydramnios and placental thickening can trigger preterm labor (6). If fetal hydrops develops before gestational viability, fetal death is inevitable (4). In SCT cases, alpha-fetoprotein and acetylcholinesterase levels are increased in amnios fluid.

Rapid growing tumors, tumors larger than 10 cm., fetal hydrops, preterm labor and malign tumor pathology are signs of a poor survey (7,8). Since our patient did not have fetal hydrops or cardiac failure signs, the amnios fluid was within normal ranges, and tumor growing rate was slow, after receiving approval of the parents, we decided to follow her until fetal pulmonary maturity was adequate for survival.

Various prenatal interventions can be made for fetal SCT. Cystic SCTs can be drained percutaneously. Two cystic SCT cases from Kay et al. (2) were delivered vaginally without complication after prenatal percutaneous drainage and they reported the method can be an alternative to caesarian for SCTs. It was shown that radiofrequency ablation can cause intratumoral hemorrhage, intrauterine death, preterm labor and tissue necrosis (9). Adzick et al. (10) were the first who used open fetal surgery in 1997. However this method has premature labor and postnatal morbidity risks (7). For the success of prenatal interventions they must be conducted before high output cardiac failure development (11,12).

Prenatal definition of SCT is important for prenatal management and surgical planning. During vaginal labor, severe dystocia and extremely vascular tumors can cause fetal death due to hemorrhage. Gross et al. (13) recommend caesarian section for tumors larger than 5 cm. It is reported that tumor resections preventing traumatic labor have good results (7). In addition to labor dystocia, tumor rupture can cause massive hemorrhage. Large tumors can cause difficulties even with caesarian sections. Prophylactically, we used t-shut dissection to widen the space during the caesarian section.

After SCT definition, accompanying anomalies must be investigated. The rate for accompanying renal, muscular-skeletal and nervous system is 18%. Meningomyelocele and sacral hemangioma must be remembered in differential diagnosis. In particular, a cystic sacrococcygeal teratoma may be misdiagnosed as an anterior sacral meningomyelocele, especially when presenting as a posterior cystic mass (14).

Neonates with SCT after excision require long-term follow-up for functional impairment. Surgical trauma, tumor compression or invasion of nerves contribute to this condition. Garba et al (15) reported that approximately one-third of patients developed some degree of long-term functional impairment, including soiling, neuropathic bladder, vesicoureteric reflux, and recurrent urinary tract infections. Schmidt et al. (16) reported functional abnormalities (fecal soiling, nocturnal enuresis, perineal anesthesia and recurrent urinary tract infections) in 5 of 17 patients. In a questionnaire study, Halley et al. (17) reported the functional results after resection of neonatal SCT. In the study, only one patient (1/14) had problems with fecal or urinary continence, or lower extremity weakness. However, constipation (5/10) was seen relatively often.

Tumor recurrence occurs in 7.5% to 22% of cases (18). Derikx et al. (19) reported that the recurrence rate of SCT in 173 children was 11% within 3 years and was associated with immature and malignant histology and incomplete resection. Postoperative monitoring of serum AFP levels is also useful in detecting early tumor recurrence.

As a result of early diagnosis of fetal SCT, routine USG monitoring for fetal complications is important for prenatal management and surgical planning of SCT. While planning delivery of the fetus, the least traumatic method must be selected and fetus must be referred to pediatric surgeons in a stable state and as early as possible. During the postoperative period, long-term follow up for functional impairment including urinary or/and fecal disorders and recurrence is necessary. Fetal SCT needs coordination between the obstetrician, pediatrist and pediatric surgeon.

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