Prenatal diagnosis and follow-up of giant sacrococcygeal teratoma

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Sacroccygeal teratoma is a non-organ-specific soft tissue lesion composed of extragonadal tissues located in the pericoccygeal region. Sacrococcygeal tumor, a germ cell tumor, is quite rare in the general population; however, it is the most common congenital tumor in newborns. The incidence for this tumor is estimated to be 1:40,000 live births [1]. Although it has a benign histological character, it has a relatively high mortality rate (>50%) among patients diagnosed prenatally [2].

We hereby, present a patient who presented to our department with an estimated gestational age of 32 weeks, whose fetus was observed to have a sacrococcygeal teratoma measuring 20 cm by ultrasound (US) and fetal magnetic resonance imaging (MRI) evaluations.

A 23-year-old primigravid woman presented to our department for antenatal follow-up at the 32nd gestational week. She stated that she had been followed-up regularly in another medical center throughout previous gestational weeks. A solid lesion with solid cystic components with a size of 20 cm × 18 cm located in the sacrococcygeal region of the fetus was detected on ultrasonic examination of the patient. Her amniotic index was measured as 210 mm and placentomegaly was noted. We confirmed the gestational age consistent with 32 weeks of gestation. Furthermore, a detailed investigation was performed to affirm clear diagnosis.

Color Doppler sonography designated that the mass was not well vascularized, and there was no evidence of fetal hydrops. Fetal echocardiographic examination was normal. Fetal MRI investigation revealed a solid lesion measuring 20 cm × 17 cm with a hypointense peripheral margin extending through the pelvic and anal region, involving diffuse cystic areas. Its main component was located at the right side of the fetus. The lesion primarily suggested the diagnosis of Type I sacrococcygeal teratoma (Fig. 1).

The patient was hospitalized and monitored with serial US and external fetal monitoring. There was no designated aggravation on the fetus but regular contractions were detected on cardiotocography. The patient had no cervical dilatation. Corticosteroid therapy was initiated for pulmonary maturation as well as nifedipine for tocolysis. The patient underwent consultations by the departments of pediatric and pediatric surgery, and it was concluded that the patient should be followed-up until term and postpartum surgery should then be performed. However, cesarean section was performed on the 32nd week and 6th day of gestational age because of early membrane rupture and labor was began. A female infant was born weighing 3,340 g and Apgar score of 6 at 1 minute and 9 at 5 minutes. A lesion consistent with teratoma with a diameter of 20 cm located at the sacrococcygeal region was observed during postpartum examination of the newborn (Fig. 2). A necrotic hemorrhagic region with a diameter of 2–3 cm was seen to be located on the teratoma. Alpha-Feto Protein (AFP), beta-Human Chorionicotroductive Hormone and Cansinoembryogenic Antigen were measured as 308701 IU/mL (∗1), 483 mIU/mL (∗1) and 2.16 ng/mL (∗), respectively.

The newborn underwent a surgical procedure on the second postpartum day at the department of pediatric surgery. A solid lesion weighing 1,700 g was removed from the sacrococcygeal region. Histopathological examination revealed that the diagnosis of immature teratoma and histologic grade of the cells was Grade III. No early complications were developed and the newborn was discharged after 12 days of postoperative care. The newborn was consulted by pediatric oncologist and chemotherapy was advised, but the infant didn’t receive chemotherapy, because her parents did not consent for administration of chemotherapy. No recurrence occurred yet at follow-up of 11th month using physical examination, ultrasonography, and MRI.

Teratomas account for 37–52% of newborn tumors, whereas 40–70% of teratomas are located in the sacrococcygeal region [3]. It is the most commonly observed tumor among newborns. The female: male ratio is 4:1. Teratomas are classified as mature (fetal) and immature (embryonic) histopathologically. Although most teratomas are benign, they may lead to perinatal complications (83–90%) and increase the mortality rate [3]. Malignancy is the major poor prognostic
factor [4] and 4–20% have malignant tissue at delivery [5], as in our case. Gestational age and surgical operability are among other prognostic factors. The incidence of the tumor size being greater than 10 cm is especially high in vascular tumors, and these tumors, along with disorder of the cardiac output and heart failure, increase the risk of fetal hydrops and intrauterine fetal demise [6]. The prognosis is poor among infants diagnosed before the 30th gestational week because of the increased risk of premature delivery and fetal hydrops resulting from uterine distension [1]. The survival rate is very high among infants born just before term. Our patient had no fetal abnormality, except for polyhydramnios and placentomegaly at 32 weeks of gestation. Despite the tumoral width, no increased vascularization was detected on Doppler ultrasonographic examination. There was no ultrasonographic evidence of fetal hydrops, and fetal echocardiographic examination was normal.

Altman et al have classified sacrococcygeal teratomas according to the ratio of tumoral involvement of sacral wall components. Type I is located out of the wall, whereas Type IV is located totally in the presacral region. The tumor is referred to as Type II and Type III when more than half of the lesion is located in and out of the pelvic wall, respectively [7]. The more the tumor invades the sacral wall, the harder it is to remove the lesion surgically. Our case was assessed in conformance with Type I. US, MRI, Color Doppler are providing crucial information for definitive diagnosis and can also aid in determining the timing and mode of delivery.

Cesarean section is recommended for patients with lesions greater than 5 cm because of the risk of perinatal delivery trauma and dystocia [4]. It should be kept in mind that teratomas may lead to complications such as bleeding and rupture. Upper vertical incision is usually preferred during caesarian section [4]. However, lower segment incision was performed in our patient because of the increased risk of bleeding and uterine scar rupture following pregnancies because of lower segment incision.

Surgical therapy and especially histological features are the main factors determining the survival rate during the postnatal period. Accurate timing for tumoral resection in the postpartum period is important. Resection was performed on the second postpartum day after stabilization of the hemodynamic status in our newborn.

The recurrence rate following resection of teratomas has been reported to be about 11%. Although half of recurrences are malignant, and the long-term survival rate is relatively high because the tumor is chemo-sensitive. Therefore, at least 3 years of follow-up including serum AFP levels and physical examination is recommended in the postoperative period. The AFP level is high in the normal population during fetal life, whereas it decreases to normal levels at the ninth postpartum month [2]. A rapid increase in AFP level is diagnostic for recurrence [4]. Neither physical examination nor radiological images revealed evidence of recurrence in our patient until the 11th month of the follow-up period despite diagnosis of immature teratoma.

Although sacrococcygeal teratoma 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.

References


Fig. 1. Magnetic resonance image of the sacrococcygeal teratoma.

Fig. 2. Necrotic area on the surface of the about 20 cm diameter giant Type I teratoma located at the sacrococcygeal region of the newborn.

