Sacrococcygeal Teratoma a Rare Disease: Case

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Abstract: Sacrococcygeal teratomas are rare congenital tumors that develop early in fetal life. Fetuses with this malformation are at risk of significant perinatal morbidity and mortality. This case report demonstrates the benefits of the early diagnosis and intervention of sacrococcygeal teratoma. In this case study a fetus was identified with sacrococcygeal teratoma during 33 weeks antenatal scan of 11.48×10.97×9.02 cm size. The mother opted for elective caesarean section following counseling but due to pre-term labour at 36 weeks of gestation, the mother underwent emergency caesarean section without any complications. A newborn healthy female was delivered without any fetal complications. The baby was scheduled for enbloc surgical resection of the tumor on the 3rd day of life. Histopathological report did not reveal any malignancy. Neonate had an uneventful recovery. Neonate was followed up with regular follow-ups.

[Almushait M. **Sacrococcygeal Teratoma a Rare Disease: Case Report**. *J Am Sci* 2012;8(9):154-156]. (ISSN: 1545-1003). http://www.jofamericanscience.org. 22

Keywords: fetal sacrococcygeal teratoma; ultrasonography

1. Introduction

Sacrococcygeal teratomas (SCT) originate from Hensen's node (primitive knot), which is an area of the primitive streak. As the mesoderm proliferates, the primitive streaks may persist moves further caudally where the remnant of Hensen's node descends to the tip of the coccyx or its anterior surface gives rise to a SCT. >90% is benign (Donellan and Swenson, 1968).

Although generally a rare condition, it is said to be the most common tumor in the newborn period, with a reported incidence of approximately 1 in 35,000–40,000 live births (Backer et al., 2001, Flake, 1993, and Pantanowitz et al., 2001). It is a more common tumor in newborn females (Pantanowitz et al., 2001), with a male: female ratio of about 1: 3–4 (Altman et al., 1974 and Keslar et al., 1994).

The tumors are divided and classified according to their location, type and tumor components according to American Academy of Pediatrics, Surgical Section Classification presented in

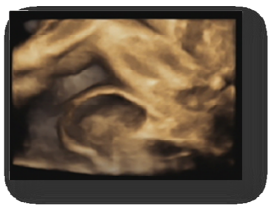
Table 1.

Type	Tumor components
I	Completely external
II	Mostly external with some intrapelvic tumor
III	Mostly intrapelvic with some external tumor
IV	Completely intrapelvic or presacrals

2. Case Report

A 31-year-old nulliparous woman Gravida 2 Para 0 Abortion 1, her last menstrual period (LMP) was on 20.02.2011 and the expected date of delivery (EDD) was on 27.11.2011. She presented herself to the antenatal outpatient clinic for regular antenatal checkup at 33 weeks gestation. A year ago, the patient had a missed abortion at 8 weeks of gestation which was followed by dilatation and curettage. Medical and family history was insignificant. There was no family history of birth defects. Relevant hematological and biochemical investigations were all within normal limits. Three dimensional ultrasonography (USG) revealed a single viable fetus with anterior fundal placenta with estimated gestational age of 33 weeks. The measurements were as follow at 33weeks –BPD 8.74cm, OFD 11.51, HC 32.52 cm, AC 30.46 cm, and FL 6.38 cm.

USG revealed a mass at the sacrococcygeal region of the fetus. The tumor contained considerable part of solid component and showed vascularity. The fetus was diagnosed to have type II SCT composed of cystic components as well as calcific foci with the 11.48×10.97×9.02 cm size and there was a pre-sacral extension of the cystic component (Figure 1, 2). The condition was not associated with hydrops or polyhydramnios. The spine, abdomen and pelvic appeared normal. There was no extension of the tumor to the bladder. The fetus did not present any other congenital anomaly. There was no evidence of intrapelvic extension of the mass.



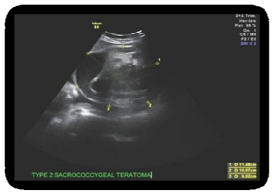


Figure 1 **Figure 2**

3D ultrasound showing the sacrococcygeal mass

Following the diagnosis, the patient was referred to the pediatric consultant and surgeon. The pregnancy was allowed to continue until fetal viability with regular antenatal checkups. The patient was closely monitored with USG to determine the size of the mass, amount of liquoramni, placentomegaly, fetal hydrops and fetal wellbeing (lung maturity). Serial scans at weeks 34 and 35 revealed no increase in the size of the mass. By week 36, the patient was advised about the risk of continuing the state of pregnancy following a family counseling for elective caesarean section, but the patient presented to the emergency with pre-term labor and an emergency caesarean was performed without any complications. It was a newborn female of 3.1kg, apgar score of 5 and 8 in 1st and 5th minutes respectively after birth. Placental size and weight were within normal limits (Figure 3).

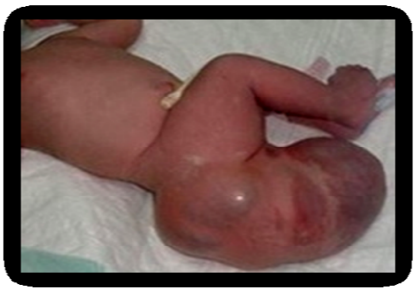


Figure 3. Gross image of the neonate with SCT post caesarean section

After that, the baby was transferred to the neonatal intensive care unit and referred to the pediatric surgery department. On observation, the baby was active and healthy looking with a mass in the sacrococcygeal region. The baby passed meconium. On clinical examination the mass showed dilated veins over its surface and a variable consistency. The systemic examinations were normal. There was no intrapelvic extension of the mass.

The newborn was scheduled for surgery on the 3rdday of life. The tumor was sub totally removed by enbloc surgical resection via sacral approach and sent for biopsy and was shown to be benign. The baby had an uneventful postoperative recovery. A follow-up of biochemical profile and physical examination was recommended to determine the developmental milestones of the baby.

3. Discussions

This is a typical case report of a newborn with SCT diagnosed at week 33 during a routine antenatal scan. Early prenatal USG detection of SCT allows optimal perinatal obstetric and surgical management. Since the presented patient did not have fetal hydrops or cardiac failure signs, the amniotic fluid was within normal ranges, and tumor growing rate was slow. The sacrococcygeal area is the most common extragonadal site for teratoma. They are the rare tumors that develop at the base of the spine by the tailbone (coccyx) although most of these tumors are benign, they grow large, and surgical intervention is essential. 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 (Kazandı et al., 2011).

In most centers, diagnosis of SCT is made during antenatal period (Chuileannain et al., 1999). Early diagnosis will enhance coordinated and well planned management of the patient. A study reported a large series in multiple centers that the average determination time of SCT is 26 gestational weeks (Tongsong et al., 1999). Usually diagnosis is made with USG (Bond et al., 1990). Three-dimensional ultrasound may better define the degree of involvement of the sacrum and pelvic structures for prognostic importance (Adzick et al., 1997).

Sonography can reveal mass at the sacrococcygeal region of the fetus. The tumor contains considerable part of solid component and vascularity. Diagnosis can determine the type of SCT composition and can show the cystic components as well as calcific foci with the size. USG must be followed to observe for further enlargement of SCT and any signs of fetal complications. Prenatal detection can be done by using ultrasound, computed tomography, or MRI in the postnatal period allowing better surgical planning.

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, caesarean section is recommended for tumors larger than 5 cm (Sherer et al., 1997). In addition to labor dystocia, tumor rupture can cause massive hemorrhage. Large tumors can cause difficulties even with caesarean sections (Gross et al., 1987).

Neonates with SCT after excision require long-term follow-up for functional impairment. Surgical trauma, tumor compression or invasions of nerves contribute to this condition. Tumor recurrence occurs in 7.5% to 22% of cases (Hedrick et al., 2004). A report suggested 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 essential to detect early tumor recurrence (Bilik et al., 1993).

Early diagnosis of fetal SCT, with routine USG monitoring, is important for prenatal

management and surgical planning of SCT (Derikx et al., 2006). While planning delivery of the fetus, the least traumatic method must be selected and the fetus must be referred to pediatric surgeon as early as possible. During the postoperative period, long-term follow-up is mandatory. Fetal SCT needs to be managed by cooperation and team efforts of the obstetrician, pediatrician and pediatric surgeon.

4. Conclusion

SCT are relatively uncommon tumors in the neonatal period. With advancement in the less invasive diagnosis and therapeutic techniques, many clinical obstetrician screen for SCT during antenatal period. Early surgical management is mandatory to prevent the spread in case of malignancy. A minimum of three post-operative follow-up is advised to detect the reoccurrence of the case.

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7/25/2012