

Sacrococcygeal Teratoma

Maryam Alahmdi, MD, Arab board OB/GYN* Rehab Farhan Al Ani, MD, BSc** Abida Qureshi, MBBS, MCPS, FRCOG*** Fayza Haider, MD, MRCSI, CABS****

Sacrococcygeal teratoma (SCT) is the most common tumor in newborns, which accounts for 1 in every 35,000 live births. We report a case of pregnant women with a fetus that had Sacrococcygeal teratoma. Ultrasound showed SCT 9 cm x 5.6 cm. She had delivered by cesarean section and the baby was operated on 15 July 2014 at age of one week of life. The baby had uneventful recovery and continued to be in good condition till now.

* Chief Resident
Feto-maternal Specialist
** Senior Resident
*** Consultant
Department of Obstetrics and Gynecology
Bahrain Defense Force
**** Consultant
Pediatric Surgeon
Salmaniya Medical Complex
Email: drm.alahmadi@gmail.com

Sacrococcygeal teratoma (SCT) is a rare tumor in the sacrococcygeal region. The coccyx is usually involved and has been reported to have genetic cause. The tumor is composed of the all three germ cells (ectoderm, mesoderm and endoderm).

SCT could be benign or malignant depending on the level of maturity but approximately 80% tend to be benign. Malignant change mostly occurs in males, those presenting in utero have poor prognosis due to complications compared to those presenting in older infants.

SCT is commonly found in babies and children but also have been reported in adults. Approximately 60% of fetuses have been diagnosed with SCT through routine use of prenatal ultrasound examination. The diagnosis of SCTs prenatally is important because large tumors can lead to congestive heart failure, hydrops, and high rates of perinatal mortality. In addition, the histological differentiation and the potential for malignancy is important for termination of pregnancy. However, the ultrasonography does not allow such differentiation¹⁻³. It is now possible to make early diagnosis of SCTs if prenatal screening applied in the first trimester, our case was a male, although there is recognized predilection with M:F ratio of 1:4^{4,5}.

If SCT is discovered, ultrasound is advised to reveal any associated abnormalities followed by a series of ultrasounds throughout the pregnancy to monitor the size and growth of the SCT and to check for the presence of any complications. A series of fetal echocardiograms may be recommended to closely monitor the fetal heart and check for early signs of heart failure.

Delivery is indicated only when fetal lung maturity is assured or there are fetal complications, e.g. fetal hydrops. Cesarean section should be performed for large SCT (more than 5cm). The treatment of SCT is postnatal surgical excision. It should be performed as early as possible to prevent further complications.

Most SCTs are benign and only require careful monitoring after removal. The long-term prognosis of babies born with an SCT is excellent and most would live normal lives. It is important to monitor the child for signs of recurrence, as it is fairly common in the first few years of life.

The aim of reporting this case is to highlight the importance of performing detailed scan for all pregnant women to detect abnormalities early.

THE CASE

A twenty-nine-year-old female (gravida 8 para 6 ab 1) at 33 weeks pregnancy attended the clinic for regular antenatal follow-up. The first scan was performed at 20 weeks which revealed to be normal; her expected date of delivery was 22 July 2014. She had previous six full terms normal vaginal deliveries. The last child was one year and half and she had one first trimester miscarriage. Her glucose tolerance test of pregnancy was normal, 5.4 mmol/L, serology was negative, HB was 10.4, blood group was A+. She was seen regularly in the clinic.

The scan showed single active, male fetus, the estimated weight was 2 kg, the placenta was anterior upper. The ultrasound features were the following: polyhydramnios, AFI 21 cm, biparietal diameter 84.6 mm, head circumference 306 mm, abdominal circumference 289 mm, femur length 62 mm. Normal Doppler studies were normal; there were no signs of hydrops and there was a good fetal lower limb movement. SCT of 9x5.6 cm (mixed echogenicity texture) was detected, no other anomalies were seen. The mother was informed about the scan findings and the condition of the fetus, see figure 1.



Figure 1: Coronal Sonogram Shows a Tumor External to the Fetus with Echogenic Tumor Content (The Arrows Point to the Mass)

At 34 weeks, ultrasound revealed a single viable fetus and no hydrops.

The patient presented with abdominal pain on 6 July 2014. Abdominal examination revealed soft with fundal height corresponding to 36 weeks. Vaginal examination revealed cervical os dilated at 1.5 cm with 60% effacement of the cervix; the vertex was at -3 station. The scan revealed single viable fetus, EFW 2.8 kg, and polyhydramnios with SCT.

She received dexamethasone, cross-matched for 4 units and had lower segment cesarean section on 6 July 2014. An alive male alive baby was delivered with huge SCT, see figure 2.



Figure 2: Sacrococcygeal Teratoma Extending from the Sacral Area and Coccyx

MRI revealed large pelvic and groin mass with multiple loculations and compartments. The mass was heterogeneous, predominantly of poor signal intensity on T1W1 and of high signal intensity on T2W1. The mass appears to displace the anus anteriorly; however, it did not appear to invade or encroach on the anus.

The baby was operated at the age of one week. The mass was completely removed with the coccyx, see figure 3.

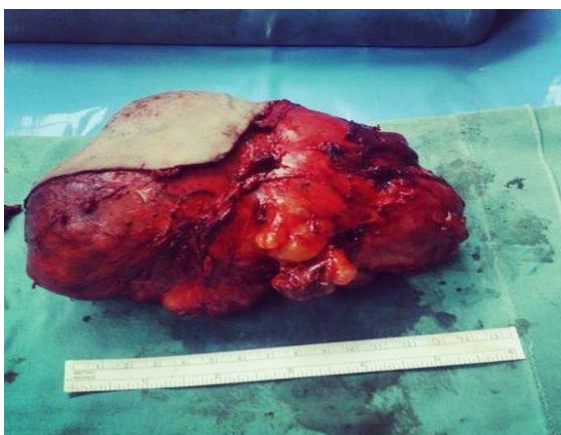


Figure 3: SCT with Mass of 705 Grams Removed

Histopathology report shows that the sacrococcyx features were compatible with grade 3 immature teratoma with focal ulceration of overlying skin; the coccyx was free of neoplastic infiltration.

DISCUSSION

The exact embryologic origin of SCT is not determined. It has been suggested that these neoplasms might result from disruption of blastogenesis; the same process that results in conjoined twins. The other theory is that it might originate from germinal cells, which are remnants of the primitive streak failed to differentiate into mesodermal, ectodermal, endodermal tissues in the embryonic disc. Some assisted reproductive techniques which involve embryologic manipulation showed that there is a possibility of disruption at various embryogenic stages which triggers SCT and other congenital tumors^{6,7}.

Teratoma should be differentiated from neurofibroma, meningocele and benign vascular tumors, such as lymphangiomas⁸.

SCTs should be removed surgically because of the malignant potential. If malignant transformation arises, en-block radical resection is indicated⁹.

SCT could cause dystocia during delivery, rupture of the SCT, hemorrhage, fetal death and injuries to the maternal birth canal^{10,11}.

MRI is a complementary modality in diagnosing SCT and is mandatory after ultrasound examination. MRI could reveal caudal or intra-pelvic mass which could be seen in the second and third trimester^{12,13}.

Endovascular embolization of giant SCT minimizes the blood loss during the surgical removal of the mass¹⁴.

Long-term follow-up is needed in the oncology unit with AFP assessment and radiological imaging¹⁵.

CONCLUSION

The prenatal diagnosis of SCT is important to predict possible complications. Those fetuses prenatally diagnosed with SCT or with associated anomaly should be subjected for a detailed ultrasound to rule out any other anomaly and detect the presence or absence of Non-immune Hydrops Fetalis. The outcome after birth in prenatally diagnosed SCT is affected by the presence of fetal hydrops or orthopedic impairment such as lower extremity weakness, swelling and urinary incontinence. Since these tumors are generally not malignant, most fetuses with SCT do well post surgical treatment after.

Author Contribution: All authors share equal effort contribution towards (1) substantial contribution to conception and design, acquisition, analysis and interpretation of data; (2)

drafting the article and revising it critically for important intellectual content; and (3) final approval of manuscript version to be published. Yes.

Potential Conflicts of Interest: None.

Competing Interest: None.

Sponsorship: None.

Submission Date: 19 August 2014.

Acceptance Date: 15 November 2014.

Ethical Approval: Department of Obstetrics & Gynecology, BDF Hospital, Bahrain.

REFERENCES

1. Kirkinen P, Partanen K, Merikanto J, et al. Ultrasonic and Magnetic Resonance Imaging of Fetal Sacrococcygeal Teratoma. *Acta Obstet Gynecol Scand* 1997; 76(10): 917-22.
2. Jelin E, Jelin AC, Lee H. Sacrococcygeal Teratoma with Spinal Canal Invasion Prenatally Diagnosed. *J Pediatr Surg* 2009; 44(4): E9-11.
3. Chou MM, Chen YF, Chen WC, et al. Prenatal Ultrasonographic Characterization of a Giant Fetal Sacrococcygeal Immature Teratoma with Four-Dimensional Ultrasound. *Taiwan J Obstet Gynecol* 2011; 50(3): 385-7.
4. Batukan C, Ozgun MT, Basbug M. First Trimester Diagnosis of Sacrococcygeal Teratoma Using Two and Three-Dimensional Ultrasound. *J Clin Ultrasound* 2011; 39(3): 160-3.
5. Roman AS, Monteagudo A, Timor-Tritsch I, et al. First-Trimester Diagnosis of Sacrococcygeal Teratoma: The Role of Three-Dimensional Ultrasound. *Ultrasound Obstet Gynecol* 2004; 23(6): 612-4.
6. Wee WW, Tagore S, Tan JV, et al. Foetal Sacrococcygeal Teratoma: Extremes in Clinical Presentation. *Singapore Med J* 2011; 52(6): e118-23.
7. Olson CK, Keppler-Noreuil KM, Romitti PA, et al. In Vitro Fertilization is Associated with an Increase in Major Birth Defects. *Fertil Steril* 2005; 84(5): 1308-15.
8. Cakmak M, Savaş C, Ozbaşar D, et al. Congenital Vulvar Teratoma in a Newborn. *J Pediatr Surg* 2001; 36(4): 620-1.
9. Szylo K, Lesnik N. Sacrococcygeal Teratoma- Case Report and Review of the Literature. *Am J Case Rep* 2013; 14:1-5.
10. el-Shafie M, Naylor D, Schaff E, et al. Unexpected Dystocia Secondary to a Fetal Sacrococcygeal Teratoma: A Successful Outcome. *Int J Gynaecol Obstet* 1988; 27(3): 431-8.
11. Villa JC, John V, Verghella V. Large Foetal Sacrococcygeal Teratoma Complicated by Preterm Premature Rupture of Membranes and Maternal Preeclampsia. *Rev Colomb Obstet Ginecol* 2007; 58(4): 322-7.
12. Danzer E, Hubbard AM, Hedrick HL, et al. Diagnosis and Characterization of Fetal Sacrococcygeal Teratoma with Prenatal MRI. *AJR Am J Roentgenol* 2006; 187(4): W350-6.
13. Avni FE, Guibaud L, Robert Y, et al. MR Imaging of Fetal Sacrococcygeal Teratoma: Diagnosis and Assessment. *AJR Am J Roentgenol* 2002; 178(1): 179-83.

14. Lahdes-Vasama TT, Korhonen PH, Seppänen JM, et al. Preoperative Embolization of Giant Sacrococcygeal Teratoma in a Premature Newborn. *J PediatrSurg* 2011; 46(1): e5-8.
15. Manzoni C, Canali R, Narciso A, et al. Sacrococcygeal Teratoma: Single Center Experience and Functional Long-Term Follow-Up. *ClinTer* 2011; 162(2): 99-106.