A case of a huge fetal sacrococcygeal teratoma with a vascular disruption sequence

Atis A, Kaya B, Acar D, Gezdirici A, Polat I, Aydin Y, Gedikbasi A
Kanuni Sultan Suleyman Training & Research Hospital, Obstetrics & Gynecology, Istanbul, Turkey

Objective
Fetal sacrococcygeal teratomas (SCT) occur in 1–2 per 20,000 pregnancies. Perinatal mortality of prenatally diagnosed SCTs ranges from 25% to 37%. Death occurs mainly in fetuses with fast growing, solid and highly vascularized teratomas, which can cause high-output cardiac failure. High-output failure leads to polyhydramnios, hydrops, intrauterine fetal demise and preterm birth. Vascular disruption defects refer to those involving the interruption or destruction of some part of the fetal vasculature. It can cause dysmorphology at nearly any stage of gestation, affect almost any tissue or structure. Here we present a rare case of huge fetal SCT causing fetal vascular disruption sequence.

Methods
A 30y old patient has referred to our perinatology clinic for a diagnosis of a mass at the back of the fetus in the sacral area at 16 th week of pregnancy. On detailed sonogram, it was found to be a huge sacrococcygeal teratoma having solid and cystic parts together. After the diagnosis, she was given genetic consultation. 2D ultrasound examination revealed a large mass of 60 x 40 mm starting from the sacral area with Voluson 730 Pro (General Electric, Zipf, Austria). No other fetal abnormalities were detected, the patient had a negative family history and she took no prenatal medications. There was no evidence of abnormal amnion. The patient returned later at 35 weeks and 2 days in labor with painful contractions and breech presentation. The previously identified mass had slightly increased in size to 155 x 92 mm. After a consulting examination with the neonatologist, pediatric surgeon, delivery was performed in our tertiary center. A cesarean section was performed and a female newborn (1830 g) was delivered an Apgar 6-7. On examination the baby found to have bilateral cleft lip and palate and lacked left forearm and some digits (figure 1, 2, 3). One week later, the baby died because of high output cardiac failure.

Results
Although most cases are benign, SCTs are associated with high morbidity and mortality rates because of preterm delivery and complications, such as malignant invasion and cardiac failure. Vascular disruption defects usually occur as a consequence of localized or general fetal hypoxia resulting from direct effects on the fetal vasculature or secondary to changes to maternal uterine or placental vasculature. In animals and humans, the most distal parts of the skeleton are the most easily affected by fetal hypoxia; thus abnormalities of the phalanges as well as facial abnormalities are the most frequent morphologic manifestations of fetal hypoxia. Similarly, in this case, the fetus was found to have limb anomalies with agenesis of left forearm and some digits and cleft lip and palate besides SCT. The SCT had a rich vascular supply so ischemia occurring in other parts of the body esp. distally may have caused the vascular disruptive fetal malformations.

Conclusion
This is the first SCT causing vascular disruption sequence although the mechanism is well known. After genetic consultation, we should keep the vascular accidents causing complex anomalies like this in mind.