Three cases of a sacrococcygeal teratoma

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Objective

We present three cases of a prenatally diagnosed sacrococcygeal teratoma (SCT). All of these cases had a different outcome. In the first case the baby was delivered by caesarean section because of a vascular steal syndrome by the teratoma with fetal anemia. After birth the neonate had surgery and the teratoma was removed with a good and functional result. In the second case there was severe cardiac failure with hydrops of the fetus at 19 weeks and the fetus died in utero. In the third case the SCT was a part of multiple malformations. An amniocentesis for fetal karyotyping was performed and showed a trisomy 13. The pregnancy was terminated.

Methods

We present three cases of a sacrococcygeal teratoma in our perinatology centre. Each case had a different outcome.

Results

Case I

A 33-year old gravida 4 para 3 was sent to our clinic at 19 weeks of gestation. Ultrasonographic evaluation concluded a large sacrococcygeal teratoma of 63x71x88 mm with signs of cardiac decompensation (reversed a-wave in the ductus venosus, tricuspid regurgitation) and signs of hydrops. Parents were counselled concerning pathology and absence of treatment options at this duration of pregnancy. The fetus died in utero.

Case II

A 27-year old gravida 3 para 2 was referred to our clinic at 29 weeks of gestation for a spinal mass, suspicious of spina bifida. Ultrasonographic evaluation showed a giant sacrococcygeal teratoma of 100 x 76 x 72 mm and a polyhydramnios with still high normal peak systolic velocity (PSV) of the middle cerebral artery (MCA). Corticoids were administered. At 31 weeks and 6 days PSV of the MCA rose above 1,5 M/M. A caesarean section was performed because of expected dystocia due to the large sacrococcygeal massa and a female neonate was born with a birthweight of 1840 gram. A postnatal MRI showed a complex teratoma (Altmann type I) with a large extrapelvic component and a small intrapelvic component. Surgery was performed nine days after birth. The arteria sacralis was preoperatively visualised by ultrasonography. First the arteria sacralis was clipped by laparoscopy to reduce the vascularisation of the tumor. The sacrococcygeal mass was resected with limited blood loss. Pathologic examination concluded an immature teratoma. Genetic analysis for the Currarino syndrome was negative.

Case III

A 23-year old primigravida was referred at 27 weeks of gestation because of suspicion of spina bifida on routine ultrasound examination. Expert ultrasonographic examination revealed multiple fetal malformations. There was a hypoplastic left eye, a sacrococcygeal mass, an omphalocele, a Dandy Walker malformation and a cardiac anomaly (transposition of the great vessels and a double outlet right ventricle). An amniocentesis revealed a trisomy 13. The pregnancy was terminated on parents' request at 28 weeks of gestation.

Discussion

A sacrococcygeal teratoma is the most common congenital germ cell tumour consisting of all three germ layers. They arise from the totipotent cells from the node of Hensen. The incidence is 1/40,000 with a female predominance (ratio 4/1).

Diagnose is made by ultrasonography and MRI. SCTs can be classified in a location-based classification (Altmann, type I-IV); an Altmann type I SCT develops only outside the pelvis (47% of the cases) and an Altmann IV SCT is located in the pelvis. This classification is descriptive and has no prognostic value. Pathologically, SCT are classified as mature of immature. Mature types are more cystic, but the correlation between sonographic appearance and malignant potential is weak. Ultrasound is useful for monitoring the evolution of the tumor and detecting fetal decompensation in order to optimize delivery timing.

A prognostic classification made by Benachi consists of three groups based on their diameter, vascularity and growth. Group A: diameter <10 cm, absent or mild vascularity and slow growth (<8mm/week). Group B: diameter ≥ 10 cm, pronounced vascularity or vascular steal syndrome and fast growth (>8mm/week). Group C: diameter ≥10cm, absent or mild vascularity and slow growth. Highly vascularised SCT are also fast growing and related to a poor outcome (group B). Group A and C have a good prognosis. Group C are large but slow growing SCT and predominantly cystic. The prognosis is good but the tumour size can give difficulties during delivery and skin alterations in the neonate. Drainage of the cystic component can be discussed. The perinatal mortality seems to be greater in the highly vascularised group. The high vascularised SCTs are also the fast growing-ones.

There are cases described where the vascular flow was interrupted by laser. This procedure however is often complicated by preterm birth and intra-uterine death.

An amniocentesis for foetal karyotyping should be performed, definitely in case of associated malformations.

Literature

- Sacrococcygeal teratoma. Weerakkody Y et al. Radiopaedia.org