Massive fetal intracranial teratoma with hydrocephalus detected at 33 weeks of gestation

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Submitted: 2010-02-07 Accepted: 2010-02-22 Published online: 2010-04-28

Key words: intracranial teratoma; hydrocephalus; macrocephaly; germ cell tumor; MRI; USG

Abstract
Fetal intracranial teratomas are rare neoplasms that can be readily detected via USG and MRI. While early discovery of such a condition may be helpful, the prognosis remains poor. We present a case of a massive intracranial teratoma and hydrocephalus initially detected via USG at 33 weeks of gestation.

CASE REPORT
A 40 year-old woman, gravida 1 para 0, was referred to the Department of Fetal-Maternal Medicine and Gynecology, Research Institute Polish Mother’s Memorial Hospital at 33 weeks of gestation due to symptoms of preterm labour and hydrocephalus on USG examination. Her gynecological history was significant for Cervical Intraepithelial Neoplasia (CIN) Grade I but otherwise unremarkable. During her stay she was tested for Toxoplasmosis, Parvovirus, Cytomegalovirus (CMV), and Syphilis infections; the tests were significant for toxoplasmosis IgG antibodies and the presence of IgM and IgG CMV antibodies.

Abdominal sonography performed at 33 weeks of gestation revealed a fetus in vertex position. USG measurements of Biparietal Diameter (BPD) and Head Circumference (HC) measured 10.6 cm and 38.0 cm, respectively, both lying significantly out of normal range (>2SD). Other parameters used in age and growth estimation corresponded to normal development. Examination of the head revealed a massive intracranial tumor (6.06 × 4.83 cm) with abundant cystic structures. Considerable enlargement of the ventricles and lack of brain parenchyma were also noted, Figure 1. Vascularisation of the intracranial mass was also visualized. Chromosomal analysis indicated a 46, XY karyotype.

On MRI examination at 33 weeks the intracranial tumor was situated in between the border of the frontal lobes and was classified as heterogeneous with the presence of many liquid spaces, Figure 2. The upper left and lower right parts of the mass were different in character and had different
structures. T2 weighted scans showed that the lower right part was of lower signal intensity, more heterogeneous and had a higher number of cysts. The maximal width of the ventricles in axial plane measured 45 mm on the right and 40 mm on the left. The lumen of the third ventricle was not visible.

The vast growth of the fetal head past its gestational age prompted delivery via cesarean section to avoid dystocia as a result of cephalopelvic disproportion. The cesarean section was planned and performed at 34 weeks and 4 days of gestation. The weight of the infant at birth was approximately 2500 g with an APGAR score of 9 after both 1 and 5 minutes. Shortly after birth a ventricular shunt was inserted to reduce intracranial pressure. CMV tests of the infant's serum were positive for IgG antibodies but negative for IgM.

The infant died after 4 weeks. The cause of death was due to compression of respiratory and circulatory centers in the brain stem. Postmortem examination indicated macrocephaly. The HC had grown from 38 cm at 33 weeks gestation to 42 cm at birth and to 50 cm at autopsy. The fetal brain was almost completely replaced by the excised teratoma which measured 15 × 13 × 8 cm with a weight of 456 g. Although the rim of the cerebral hemispheres were present, it was not possible to find the origin of the tumor because of its enormous size and diffuse distribution. Grossly, the mass had a partly cystic and partly solid appearance with large areas of hemorrhage, necrosis and cartilaginous-like folds, Figure 3.

Histopathological inspection of the tumor demonstrated a variety of tissue elements derived from the three germ layers. The mass was predominantly composed of primitive neuroepithelial tissue forming tubules, rosette-like structures and abundant fields of chorioid-plexus-like tissue. Islands of cartilage, foci of coarse-woven bone, bundles of smooth muscle, cystic spaces lined by cuboidal epithelium (some of the cells contained mucin), salivary glands and intestinal glands were also noted in several fields, Figure 4. In areas of immature neuroectodermal tissue mitotic figures were infrequent.

The definitive diagnosis was intracranial immature teratoma.

DISCUSSION
Congenital intracranial teratomas are a rare type of Germ Cell Tumor (GCT) that account for about 0.5% of all brain tumors and 2% of brain tumors in childhood. Males are most often affected. According to the World Health Organization, 3 histological variants of tera-
teratomas can be classified: mature, immature, and those with malignant transformation, none for which grading exists (Bolat et al. 2008; Köken et al. 2008; Lipman et al. 1985; Rosenblum et al. 2007). The most frequent site for congenital teratomas is the sacrococcygeal region, but it may also occur in the neck, gonads, mediastinum and, as in our case, intracranially (Bolat et al. 2008; Di Rocco et al. 2006; Pinto et al. 1999).

USG examination and MRI can be used to detect intracranial teratomas, but ultimately, the diagnosis of teratoma can only be confirmed via biopsy or histopathology on autopsy (Köken et al. 2008; Tsutsumi et al. 2008). The first indication of an intracranial mass is usually derived through routine USG examination in the second or third trimester (Saada et al. 2009). Presentation of a fetus with an intracranial teratoma may include hydrocephalus, enlarged biparietal diameter, and increased head circumference – all of these were present in our case. Concomitant anomalies may also include polyhydramnios, cleft palate, urinary tract malformations, pulmonary hypoplasia, hepatomegaly, adrenal hypoplasia and high output cardiac failure (Bolat et al. 2008; Di Rocco et al. 2006; Köken et al. 2008; Weyerts et al. 1993). The decline in serum alpha-fetoprotein levels has also been described as a useful diagnostic marker in identification of potential immature teratomas (Oi S et al. 1990).

Regardless of what type of neoplasm, fetal intracranial tumors present as a great challenge to physicians (Carstensen et al. 2006). The prognosis depends on the size, location, histological composition, the extent of destruction of brain parenchyma and the degree of obstruction of CSF (Pinto et al. 1999). Therefore, the prognosis of treatment, regardless of method, is limited by the presence and remainder of functional brain parenchyma.

While early diagnosis may aid in ensuring a safe delivery of the infant, the poor prognosis is unlikely to be changed (Köken et al. 2008). Though the child in our case survived 4 weeks, 91.1% of fetuses with an intracranial teratoma are stillborn or dead within the first week of life. The 1-year survival rate in one large series was 7.2% (Bolat et al. 2008). Moreover, most infants that survive have severe neurological sequelae (Canan et al. 2000; Cartensen et al. 2006; Di Rocco et al. 2006). Nevertheless, one benefit of early diagnosis is the pos-
sibility to plan for a cesarean section to avoid dystocia, among other complications, as a result of cephalopelvic disproportion (Lipman et al. 1985; Saada et al. 2009).

Treatment alternatives in the management of intracranial teratomas are inadequate and therefore, the most likely outcome of intracranial teratomas remains fatal. As a result, the termination of the pregnancy during early gestation should be discussed (Bolat et al. 2008; Di Rocco et al. 2006; Pinto et al. 1999).

REFERENCES