CASE REPORT

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

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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

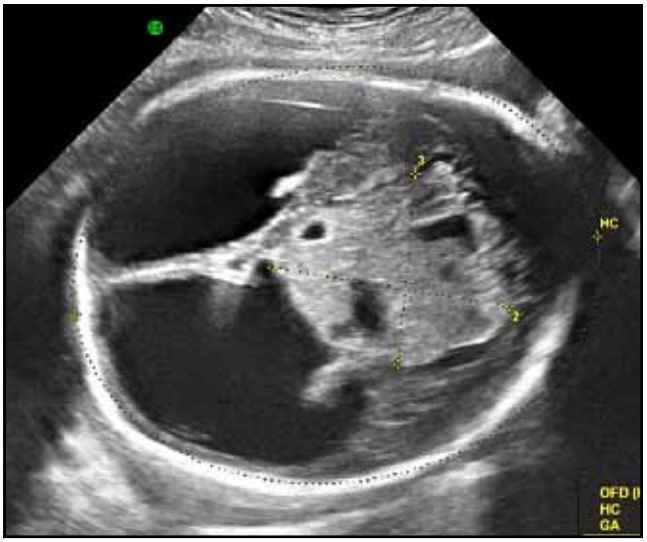

Fig. 1. USG examination at 33 weeks of gestation showing massive ventricular dilatation and an intracranial mass (6.06 × 4.83 cm), HC = 37.46 cm.

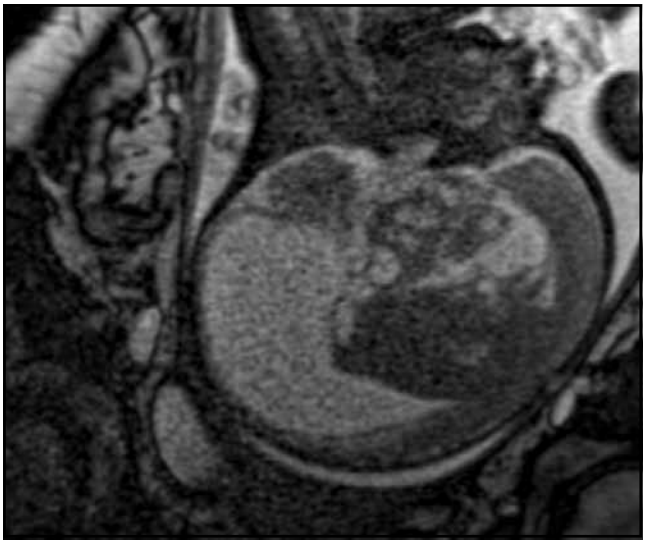

Fig. 2. MRI examination at 33 weeks of gestation showing massive ventricular dilatation and cystic structures within the intracranial tumor.

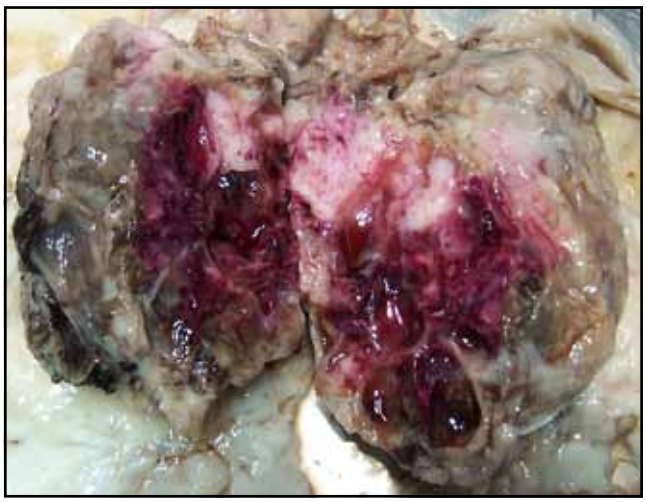

Fig. 3. Intracranial teratoma at post-mortem examination, 15 × 13 × 8 cm, 456 g.

structures. T2 weighted scans showed that the lower right part was of lower signal intensity, more heterogenic and had a higher number of cysts. The maximal width of the ventricles in axial plane measured 45 mm on the right and 40mm 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 2 500 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-

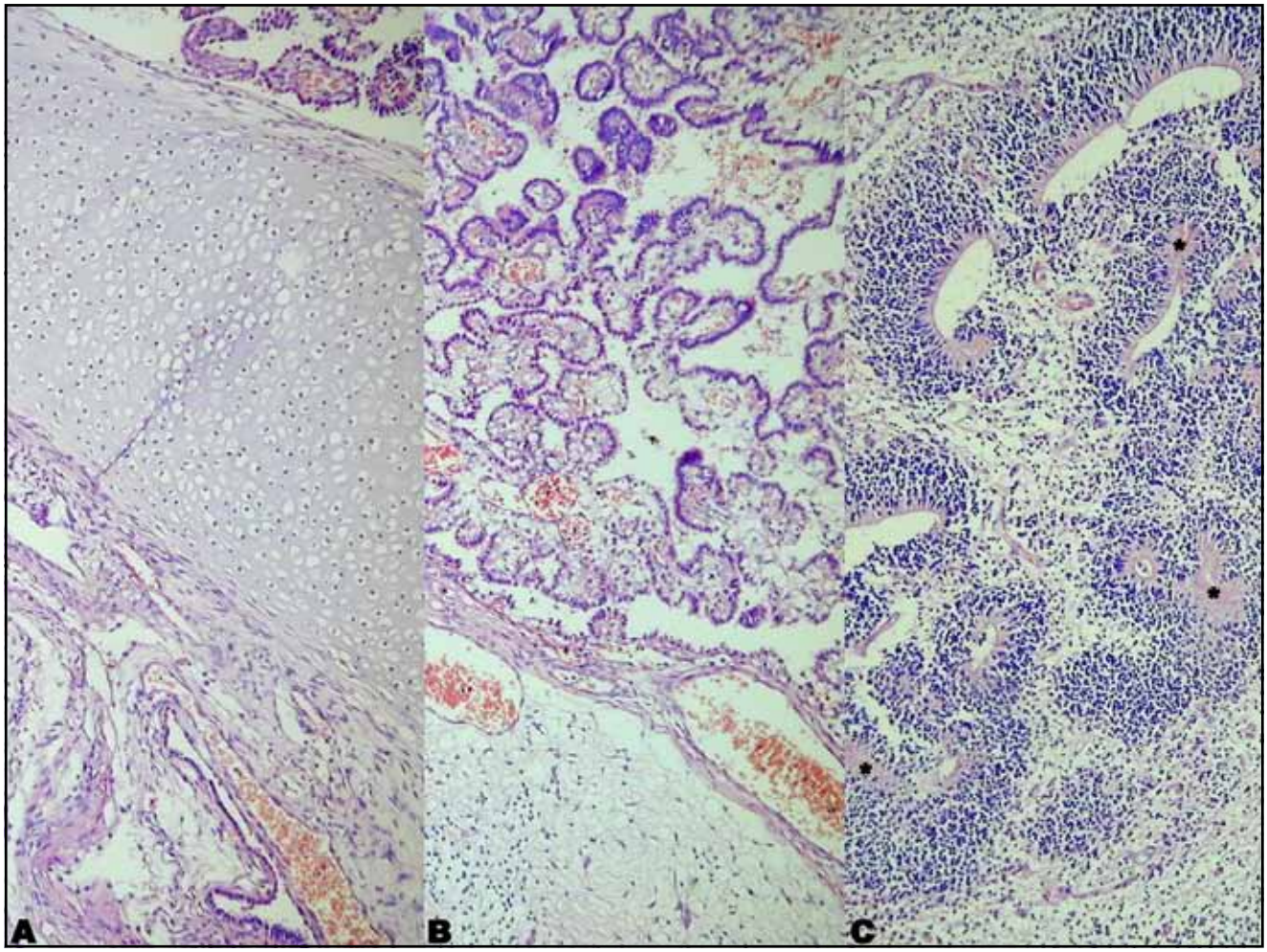

Fig. 4. Post-mortem histopathology. (**A**) Chondroid tissue. (**B**) Choroid-Plexus-like tissue (**C**) Neuroepithelial Rosettes (asterisk).

tomas 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 sequela (Canan *et al.* 2000; Cartensen *et al.* 2006; Di Rocco *et al.* 2006). Nevertheless, one benefit of early diagnosis is the pos-

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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).

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