A rare immature teratoma of the *tela chorioidea* of the third ventricle: late-onset, intrapartum ultrasound diagnosis and postnatal outcome

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Abstract We describe a previously unreported case of immature teratoma originating from the *tela chorioidea* of the third ventricle diagnosed during labor at term of pregnancy. Postnatal MR imaging and pediatric neurosurgery with postnatal outcome at 6 months of age are reported.

Keywords Immature intracranial teratoma · Hydrocephalus · Prenatal ultrasound diagnosis · Magnetic resonance imaging · Pediatric neurosurgery · Histology

Introduction

Congenital intracranial tumors are rare diseases comprising 0.4–3.1 % of all intracranial tumors, with teratomas representing 9–30 % of them [1]. Most teratomas are midline tumors located predominantly in the sellar and pineal regions [1] and are usually incidentally diagnosed during routine second or third trimester scan with very rare early diagnosis [1–6]. The tumor usually replaces the intracranial contents and cause distortion of normal cerebral anatomy being represented by a solid/cystic mass lesion associated to hydrocephalus. Teratomas involve at least one type of tissue from each of the three embryonic layers (usually accompanied by solid

areas) and are most often of benign origin, although Thurkow et al. [7] has reported an extremely rare case of malignant teratoma of the neck, with mature and immature metastatic lesions in the lungs, in an immature fetus. Teratomas of the head and neck account for only 6 % of all teratomas [8], with the majority involving the sacrococcygeal region in the fetus and the newborn [9]. Teratomas are composed of a variable admixture of skin, skin appendages, smooth muscle, cartilage, and respiratory and gastrointestinal epithelium [8]; embryonic tissue may be also present (in an “immature” teratoma) and is neural in type, as it often resembles a retinal anlage [9]. Due to the rapid growth of the tumor, the prognosis of teratomas is usually poor with a mortality rate around 90 % [10, 11].

We describe a previously unreported case of immature teratoma originating from the *tela chorioidea* of the third ventricle diagnosed during labor at term of pregnancy.

Case report

A 27-year-old with previous Cesarean section, underwent routine third trimester ultrasound examination at 31 weeks and 0 day of gestation. The scan was carried out using a real-time high-frequency ultrasound machine (Voluson E8, GE, Milwaukee, WI, USA) equipped with a 2D/3D transabdominal RAB 4-8D MHz probe. The fetus was in breech position and showed a normal growth with an estimated fetal weight of 1,969 g. Amniotic fluid was normal. As the woman requested a trial of labor, a follow up scan performed at 37 weeks and 6 days of gestation showed cephalic presentation and normal biophysical profile score. The woman went into spontaneous labor at 40 weeks and 0 day of gestation. A scan performed at hospital admission incidentally revealed an intracranial mass, partly solid and cystic, located mainly at the level of the midline echo and causing severe obstructing hydrocephalus and secondary macrocephaly. These findings

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Fig. 1 Transabdominal intrapartum scan showing a partly solid, cystic, non-homogeneous tumor starting from cerebral midline echo at the level of the midbrain (third ventricle) (a) and filling the right lateral ventricle (b). Also, note the presence of an obstructive hydrocephalus

elicited a dedicated fetal neurosonogram that was conducted using the 2D/3D transabdominal and transvaginal approach. The neurosonogram demonstrated an increased biparietal

diameter (BPD) (107 mm, >95th percentile for gestational age) and head circumference (HC) (311 mm, >95th percentile for gestational age), respectively. The midbrain (third

Fig. 2 Three-dimensional ultrasound: the volume of the tumor (52.981 cm³) was calculated using the VOCAL application

Fig. 3 **a** T2-weighted MR imaging performed at 1 day postnatal in the axial plane. Note that the tumor arises from the midbrain (third ventricle) and extends into the right lateral ventricle. Severe obstructing communicating hydrocephalus is confirmed. **b** Sagittal plane: the tumor extends caudally to reach the ethmoid-sphenoidal planum and the sellar diaphragm. The posterior-inferior portion reaches the interpeduncular

cistern, with resulting splaying of the cerebral peduncles. A cystic component of the lesion extends behind the *sella turcica* abutting the hind-brain cistern. Superiorly, the tumor involves the third ventricle and the Monro foramina, thus causing ventricular dilatation. The pituitary pedicle and the optic chiasma are not detectable

ventricle) was occupied by a large heterogeneous mass with solid and cystic content (Fig. 1), and volume calculated using VOCAL application was 52.981 cm³ (Fig. 2). Severe bilateral obstructed hydrocephalus was also observed. To prevent cephalopelvic disproportion, an intrapartum Cesarean section was performed following multispecialist counseling. A living female infant weighing 3,175 g was delivered. The 1- and 5-min Apgar score were 8 and 9, respectively. The umbilical pH was 7.23, and base excess was −4 MEq/L. The clinical examination of the newborn showed bulging and enlarged anterior fontanelle with sutures widely separated and a head circumference of 37.5 cm (>97th percentile). The eyes, ears, and neck were unremarkable. The brain ultrasound showed a large midbrain mass with hyperechogenic and hypoechogenic features measuring 37×33 mm. Angiographic study using color Doppler ultrasound revealed a forward flow pattern within the lesion. A communicating (bi-triventricular) obstructed hydrocephalus was confirmed. MR imaging was planned at day 1 postnatal life using a 1.5-T equipment (Intera Achieva®, Philips Eindhoven, The Netherlands) with a sense body coil. T2-weighted images were acquired in the three orthogonal planes using a single-shot fast spin-echo (SSFSE) sequence with a repetition time of 15,000 ms; echo time 120 ms; echo train length 140; acquisitions 2; matrix 139×256). Each pulse sequence lasted 30 s. T1- and T2-weighted imaging disclosed a large dishomogeneous tumor (4.4×3.9×3.3 cm), partly solid and cystic, filling the third ventricle and extending into the right lateral ventricle. Superiorly, the tumor involved the third

ventricle and the Monro foramina; caudally, the tumor extended until the sellar diaphragm. Severe hydrocephalus involving the third and the fourth ventricle was confirmed (Fig. 3). The newborn infant was transferred to a tertiary neurosurgery care center at 3 days of life and underwent a ventriculoperitoneal shunt placement at day 7. Radical excision of the tumor was uneventfully carried out a week later. Histology examination revealed the intracranial mass to be an immature teratoma originating from the *tela chorioidea* of the third ventricle (Fig. 4).

Fig. 4 Histological examination demonstrating an immature teratoma: the dominant tissue type in this tumor results from immature neuroepithelium in a fibrillary background (hematoxylin eosin 910 HPE)

At 6 months of postnatal life, the infant is currently undergoing chemotherapy and anticonvulsant therapy and the neurodevelopmental outcome appears normal.

Conclusion

Teratomas are rare tumors composed of tissue originating in all three embryonic layers, with an occurrence of 1:20,000 to 1:40,000. Only 5 % occur in the head or neck, and of these, malignant transformation has been reported in less than 5 % of cases. Although the first case of intracranial teratoma has been reported in the 1864 [12], only a few cases of congenital intracranial teratomas have been described: massive tumors replacing the intracranial contents, smaller ones causing hydrocephalus, intracranial teratomas with extension into the orbit or neck, and teratomas presenting as an incidental post-mortem finding in a stillborn fetus [10]. The knowledge of the anatomy of the third ventricle is essential to understand the clinical implication of an etheroplastic-growing mass located at its level and secondary complications like obstructing hydrocephalus and intraventricular hemorrhage. Nowadays, the prenatal diagnosis of intracranial tumors relies on ultrasound and antenatal/postnatal MR imaging or computed tomography (CT) scan. As of 2003, only two cases of congenital intracranial teratoma (CIT) have been reported by means of half-Fourier single-shot turbo spin-echo (FASTE) imaging. Marden et al. [13] described the first case of CIT using the true fast imaging in steady-state precession (FISP) MR sequence. According to Marden et al. [13], true FISP can be obtained in almost 20 s and provides superior contrast resolution compared with HASTE. Prenatal detection is crucial for timely counseling to the parents and to arrange appropriate obstetric management and neonatal assistance. To potentially prevent cephalopelvic disproportion, Cesarean section is performed in 60 % of cases [14]. Subdural hemorrhage [15] and fetus-in-fetu (FIF) [16] should be considered in the differential diagnosis of all congenital intracranial masses detected prenatally by ultrasound. Subdural hemorrhage is usually located in the supratentorial region, and maternal anticoagulant therapy, maternal abdominal trauma, and/or fetal coagulation disorders may be associated [17]. Fetus-in-fetu (FIF) is a rare condition where a degenerated fetiform mass is incorporated within the body of a fetus. Although most cases of fetus-in-fetu occur in the abdomen, Marnet et al. [16] reported a case of FIF of the third ventricle associated with a triventricular hydrocephalus resembling an intracranial teratoma.

Hydrocephalus may be the consequence of a hemorrhage caused by the tumor or by blockage of the ventricular draining system as it was in the current case. The prognosis of congenital intracranial teratomas is usually poor with only a 10 % survival rate [5, 14, 18, 19].

To the best of our knowledge and after electronic search of PubMed/Medline using “intracranial tumor,” “intracranial teratoma,” and “congenital brain teratoma” as keywords, this is the first observation to report the prenatal diagnosis of a late-onset congenital intracranial teratoma. Postnatal MR imaging and neurosurgery with histologic examination confirmed this lesion to be an immature teratoma originating from the *tela chorioidea* of the third ventricle.

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