

Congenital intracranial mature teratoma: the role of fetal MRI over ultrasound in the prenatal diagnosis and the perinatal management

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DESCRIPTION

A 30-year-old mother, gravida 2, para 0010, had a normal pregnancy while performing the regular prenatal examinations. From her medical history, there was nothing noteworthy. In the third trimester ultrasonography, at 31 weeks' gestation, a large hypoechoic oval mass was observed in the growing fetal brain parenchyma of the right cerebral hemisphere deviating the midline to the left. Normal intracranial structures were identifiable, and there was no ventricular dilatation. The differential diagnosis included a cerebral cyst versus a fetal cranial neoplasm. Fetal MRI showed a round mass with fine borders and smaller cystic lesions, located at the right temporal lobe, setting the suspicion of teratoma (figure 1).

After discussion with the parents, they chose to proceed with the pregnancy without any intervention. The patient came to spontaneous onset of labour at 36 weeks. The vaginal delivery was successful without any complication. A female infant was delivered, weighing 3260g and with head circumference measured 38.2cm (>90th percentile). Physical examination revealed a large head with wide fontanels. No other congenital anomaly was observed and neurological examination was normal.

Immediately after the birth, at first postnatal day, a new MRI of the newborn was performed. The mass was hyperintense in T2-weighted MRI scan, with well-defined borders, cystic formations

and direct continuation from the encephalic parenchyma (figure 2). In addition, the mass showed contrast enhancement in T1-weighted MRI. At that time, a right temporal craniectomy and total excision of the tumour was considered necessary due to mild ventricular dilatation. The histopathological result from the intraoperative tissue of the mass confirmed the diagnosis of fetal mature teratoma. The infant is under a 5-year follow-up without any indication of recurrence or significant neurological deficits.

Congenital intracranial tumours are rare and account for approximately 0.5%–1.5% of all child-hood brain tumours, however teratomas are the most common type representing the majority of brain tumours during pregnancy. Congenital intracranial teratomas present high frequency of fetal and postnatal death, with overall survival rate being estimated at 7.8%.

They are seen in various sites within the central nervous system: cerebral hemispheres, pineal, hypothalamic area, suprasellar region and third ventricle. Different forms have been described, as regards to size (huge/small), presentation (causing hydrocephalus or incidental finding) and location (intracranial/extracranial).

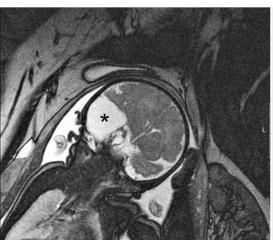
The average maximum tumour size is 10 cm, with mean age of ultrasound diagnosis at 32 weeks' gestation and no differentiation regarding gender. Most teratomas are detected on routine prenatal ultrasound examination. Major signs are macrocephaly,



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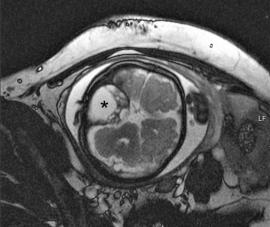
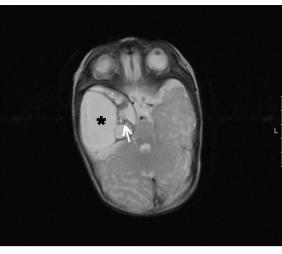


Figure 1 Fetal MRI: T2-weighted coronal MRI (on the left) and T2-weighted axial MRI (on the right) depicting the oval-shaped hyperintense mass (black asterisk) surrounded by cyst-like areas, located in the right temporal lobe of the fetus and slightly deviating the midline to the left.





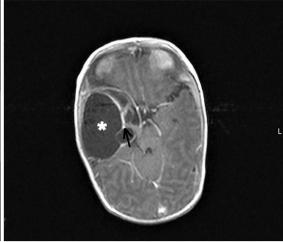


Figure 2 MRI scan after the delivery of the newborn: T2-weighted MRI (on the left) and T1-weighted MRI with contrast (on the right) depicting a heterogenic mass (white and black asterisk) with well-defined borders and cyst-like areas of variable size (white arrow) and contrast enhancement (black arrow).

intracranial mass and hydrocephalus. The sonographic and MRI appearance of the intracranial teratoma is usually that of an irregular heterogenous mass with hyperechogenic and hypoechogenic features, with both solid and cystic areas and/or calcified components, distorting brain anatomy. ^{4 6}

The use of MRI, in the perinatal period, is the imaging modality of choice for indicating the morphological features of teratoma, for estimating the feasibility of a possible surgical resection, for allowing the surgeon to better advise the parents on the potential sequelae of surgery and the overall prognosis and thus, for optimising perinatal care.^{6–8}

Learning points

- Congenital intracranial mature teratoma is a rare entity with difficult diagnosis, challenging management and poor prognosis.
- ► Fetal MRI is the imaging modality of choice for early recognition of teratoma and for optimisation of perinatal management.

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