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CASE REPORTS

Intracranial Extracerebral Ectopic Brain Tissue (Glioneuronal Heterotopia): Rare Case Report

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Abstract Intracranial extracerebral ectopic brain tissue, also called glioneuronal heterotopia, is a rare developmental disorder. These lesions are predominately seen at the base of the brain in the supratentorial region and are frequently associated with craniofacial anomalies. We report prenatal ultrasonographic and magnetic resonance imaging (MRI) findings of isolated extracerebellar ectopic brain tissue in the posterior fossa of a 22 weeks fetus.

Case Report

A 27-year-old primi at 22 weeks gestation was referred to us for a routine antenatal scan. Ultrasound examination showed a well defined round echogenic mass located in the midline of the cisterna magna posterior to a normal vermis (Fig. 1). The cerebellum was anatomically normal with a normal transverse cerebellar diameter for gestational age. On color doppler examination, the mass was not vascular (Fig. 2). 3D images were also acquired which showed that the mass was located in the region of the cisterna magna (Fig. 3). The size of the cisterna magna was within the normal range. No other structural cerebral or extracerebral abnormalities could be seen. Biometric parameters were normal for gestational age. Fetal cerebral MRI showed a well defined round and solid mass, posterior to the vermis, with a signal similar to that of normal brain tissue on T2-weighted imaging (Fig. 4) and with no hemorrhagic changes on T1-weighted imaging. Vermian and hemispheric cerebellar anatomy were normal. Imaging findings were suggestive of ectopic/heterotopic cerebral tissue.

Discussion

The main abnormalities of the posterior fossa that are observed on antenatal ultrasound examination include a small posterior fossa associated with a neural tube defect (Chiari II malformation), an increase in the size of the fluid-filled space of the posterior fossa, small cerebellar biometry and a heterogeneous group of rare focal parenchymal cerebellar abnormalities [1]. In this case, the posterior fossa was normal in size. Cisterna magna was also within normal limits with normal cerebellar biometry.

True ectopia in the posterior fossa should be distinguished from the more frequent focal cerebellar cortical dysplasia or heterotopia (typically small clusters of cells located within the cerebellum), or heterotopic cortex and heterotaxy [2]. These intraparenchymal abnormalities are identified in the cerebellar white matter and are mainly recognized as incidental postmortem findings that may

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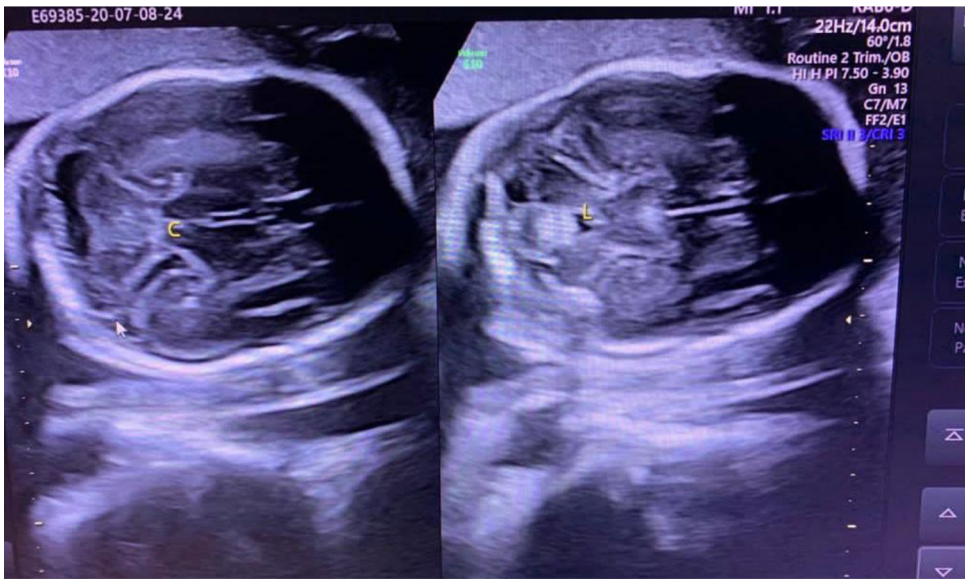

Fig. 1 Cranial ultrasound shows a well defined round echogenic mass (L) is seen in the retrocerebellar location in the region of the cisterna magna. The cerebellum (C) is within normal limits for age

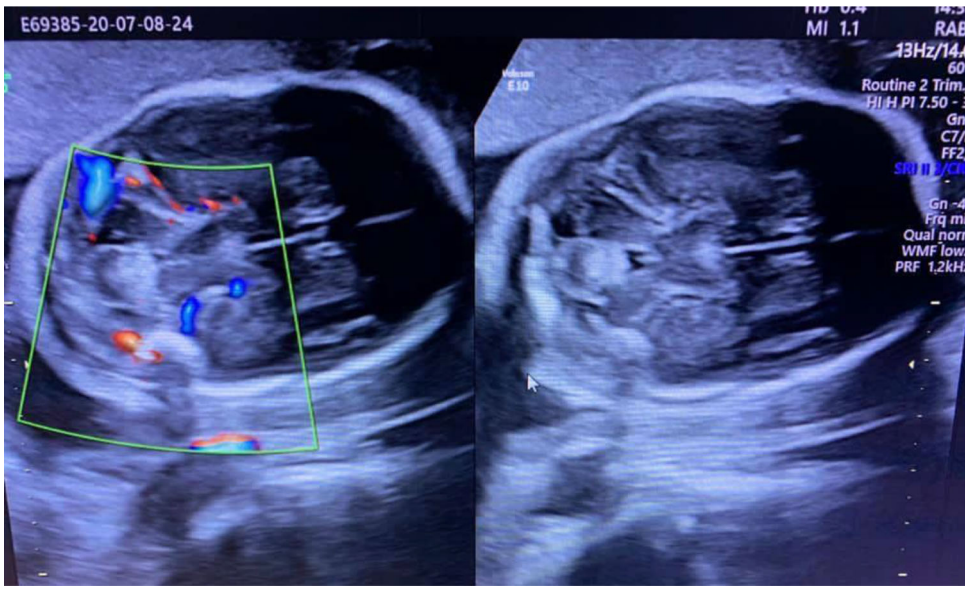

Fig. 2 On cranial ultrasound with color doppler, the mass does not show any internal vascularity. Normal vascularity was seen in the rest of the posterior fossa

represent normal variants [2]. In this case, the abnormality was a true ectopia of extracerebellar brain tissue.

These extremely rare developmental disorders have been reviewed by Oya et al. [3] and are grouped under the term 'intracranial extracerebral glioneuronal heterotopia' (IEGH). Oya et al. reviewed ten cases in the literature. All reviewed cases were located in the supratentorial region, mainly in the anterior and middle cranial fossa. This case is atypical since the lesion was located in the subarachnoid spaces of the infratentorial region and was not associated with any other abnormality.

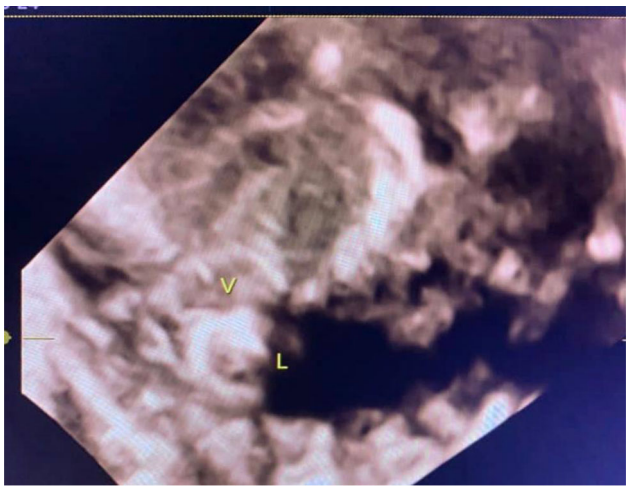

Fig. 3 Cranial ultrasound with 3D acquisition shows the mass (L) in the retrocerebellar location close to the vermis (V)

Summary

Intracranial extracerebral ectopic brain tissue (also called glioneuronal heterotopia) is a rare developmental disorder. The abnormality can be detected on routine fetal ultrasonagraphy and can be confirmed with an MRI which shows a mass with signal characteristics similar to normal brain tissue.

Declaration

Conflict of interest There is no conflict of interest.

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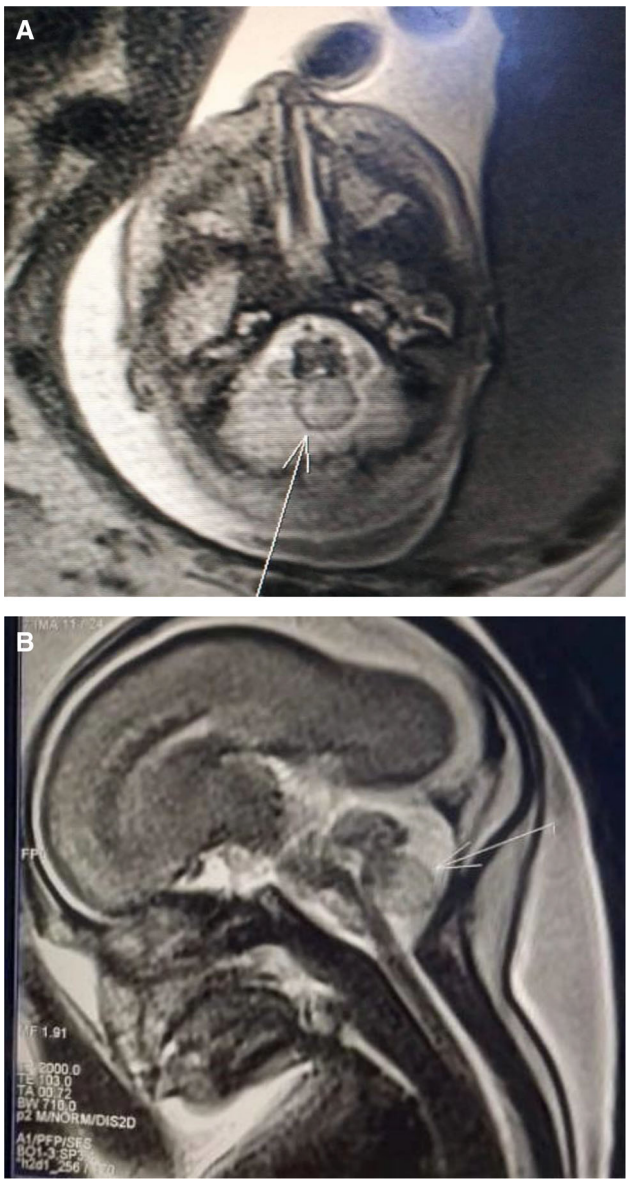

Fig. 4 Fetal magnetic resonance imaging at 22 weeks showing a solid mass (arrow) in the infratentorial subarachnoid spaces on T2 weighted axial (**A**) and sagittal (**B**) images with a signal similar to normal brain tissue