Corpus callosum agenesis with interhemispheric cyst: a neuroimage to remember

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DESCRIPTION

An 18-month-old boy presented with global developmental delay since early infancy, with developmental age of 6 months without any dissociation along different domains. Vision and hearing were apparently normal. He was born to non-consanguineous parents at term with a smooth perinatal transition. There were no antenatal complications, although level II sonography was not done. He was hypotonic with an age-appropriate head circumference (47 cm). MRI of brain revealed a rare peculiar developmental malformation (callosal agenesis and an interhemispheric cyst communicating with lateral ventricles; [figure 1](#)). He was initiated on rehabilitative measures and parents were counselled regarding the prognosis.

Agenesis of corpus callosum (ACC) is a rare malformation reported in children with developmental delay.¹ The clinical phenotype may vary from normalcy to epilepsy and intellectual disability, and depends on the nature of associated anomalies—interhemispheric cyst, hydrocephalus, Dandy-Walker malformation or interhemispheric-fissure lipoma.^{2–3} Although the index child did not have any dysmorphism or associated anomalies, some children with ACC may have dysmorphic features such as hypertelorism, polysyndactyly, hallux duplication and so on.⁴ Barkovich classified ACC with interhemispheric cyst into type 1 cysts (diverticula of the lateral or third ventricles), and type 2 cysts (loculated cysts, not communicating with ventricles).⁵ Antenatal ultrasound is a good screening tool for identification of congenital cerebral malformations, which in our index child, was unfortunately not done. In

the postnatal period, although ultrasound may be a useful screening tool, MRI is the modality of choice, where type 1 cysts are isointense to cerebrospinal fluid (CSF), while type 2 are hyperintense on both T1-weighted and T2-weighted images. In our index child, neuroimaging was suggestive of a type 1 cyst. This case highlights the importance of neuroimaging in children with developmental neurological disorders. The identification of this peculiar malformation is necessary since it clearly delineates the diagnosis and avoids unnecessary investigations. Also, in selected cases, these cysts may require shunting depending on their natural history.⁶

Learning points

- ▶ Neuroimaging is essential in developmental delays even if head size is normal, more so if neurological examination is abnormal.
- ▶ Association of interhemispheric cyst with callosal agenesis is rare.
- ▶ In cases of callosal agenesis with interhemispheric cyst, polymalformative syndromes such as acrocallosal syndrome should be considered if facial dysmorphism and digital anomalies are noted.

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Figure 1 Axial (A–C), coronal (D,E) and sagittal (F) T2-weighted MRI of the brain showing an interhemispheric cyst (arrows) communicating with lateral ventricle (type 1 cyst) with non-visualisation of corpus callosum.

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