Interhemispheric Cysts with Agenesis of the Corpus Callosum Requiring Open Fenestration

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We present the case of a male infant diagnosed with an interhemispheric cyst and agenesis of the corpus callosum on a 15-week gestational ultrasound. He was born at term and followed with serial magnetic resonance imaging (MRI). Figure 1a and 1b demonstrate multiloculated interhemispheric cysts with agenesis of the corpus callosum and cortical abnormalities consistent with pachygyria/polymicrogyria obtained at 37 days of age. Progressive headaches and further imaging that demonstrated expansion of the cysts necessitated a neurosurgical referral at 16 months of age. At this time, his gross and fine motor milestones were delayed (11- to 12-month stage), but the remainder of his neurological exam was normal. Given the growth observed on serial imaging (Figure 1c and 1d), his parents consented to open fenestration of the cysts.

We elected to perform an open approach rather than an endoscopic approach for two main reasons. Firstly, an open approach provided us with a better ability to extensively perforate the walls of cysts than an endoscopic approach would have allowed. Secondly, we are unable to use MRI image guidance with an endoscopic approach at our centre. Image guidance provided us with an improved ability to accurately locate the deeper cysts. Overall, we felt that these advantages outweighed the less invasive advantage associated with an endoscopic approach.

The patient underwent an MRI image-guided left parasagittal craniotomy for fenestration of the multiloculated interhemispheric cyst. The superior cyst was initially accessed, and a portion of its wall resected. Its fluid appeared yellow and turbid in nature. The midline cyst was then fenestrated, demonstrating more clear fluid. The trigone of the left lateral ventricle was visualised through the inferior cyst and was successfully entered. The anterior cyst could not be safely accessed. The patient had returned to his neurological baseline by postoperative day 16 and was discharged. His postoperative MRI (Figure 1e and 1f) demonstrates a reduction in the size of the cysts fenestrated.

The superior cyst's wall was sent for pathological analysis and its architecture consists of a collagenous structure lined by a flattened to low cuboidal epithelium (Figure 2). Nuclei are blunt oval with delicate chromatin. No squamous features are present. Microvillus-like projections are noted focally. The cyst lining expresses cytokeratins (CKCAM5.2) and S-100 protein. Glial fibrillary acidic protein (GFAP) and epithelial membrane antigen (EMA) are more sparse and patchy in expression. This unusual pattern implies choroid/ependymal lineage allowing for mixed epithelial and glial features.

Interhemispheric cysts have reportedly been associated with agenesis of the corpus callosum, although the pathophysiological mechanism remains to be elucidated. Barkovich et al. classified these cysts according to morphological features on imaging, as communicating (Type I) or not communicating (Type II) with the ventricular system.[1] To our knowledge, there have been 13 previous reported cases with histological analysis available: 8 arachnoidal,[2-4] 3 ependymal[2,5,6] and 2 choroidal epithelial.[7,8] Two cases of ependymal cysts expressed GFAP[5,6] with one additionally expressing S-100 and cytokeratine.[5] Other cases did not report immunohistochemistry results. Of these cases, seven were symptomatic,[2,4,7,8] five were incidental findings[2,5,6] and one was unspecified.[3] Surgical management for these lesions includes cysto-peritoneal shunting[3,5,6] or open cyst fenestration with either a cystoventriculostomy[2,4] or cystocisternostomy.[2]

To our knowledge, this is the first reported case of an interhemispheric cyst and agenesis of the corpus callosum with both choroidal and ependymal histopathological features. Additionally, this case highlights the importance of continued surveillance in patients with these lesions, as they have the potential to expand or result in symptoms necessitating surgical intervention.

Disclosures

The authors have no conflicts of interest to disclose.

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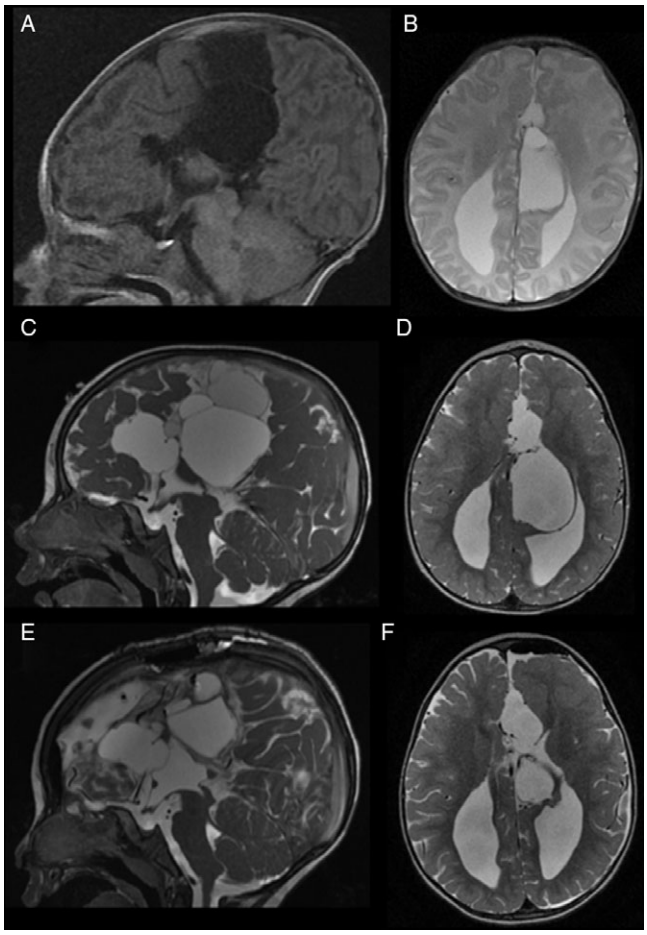

Figure 1: *(A) T1 weighted sagittal and (B) axial T2 weighted axial MRI at day 37 of life demonstrating three interhemispheric cysts to the left of midline (superior 1.6 × 1.2 cm, inferior 2.7 × 1.7 cm, anterior 0.97 × 0.88 cm) with agenesis of the corpus callosum. (C) Sagittal FIESTA and (D) axial T2 weighted MRI at 18 months of gestational age redemonstrating the interhemispheric cysts with increase in size (superior 3.7 × 3.8 cm, inferior 5.3 × 3.2 cm, anterior 3.2 × 2.0 cm). (E) Sagittal FIESTA and (F) axial T2 weighted MRI completed 14 days postoperatively demonstrating fenestration of the superior and inferior interhemispheric cysts with reduction in size and mass effect (superior 1.3 × 1.8 cm, inferior 3.7 × 2.5 cm, anterior 2.3 × 3.7 cm).*

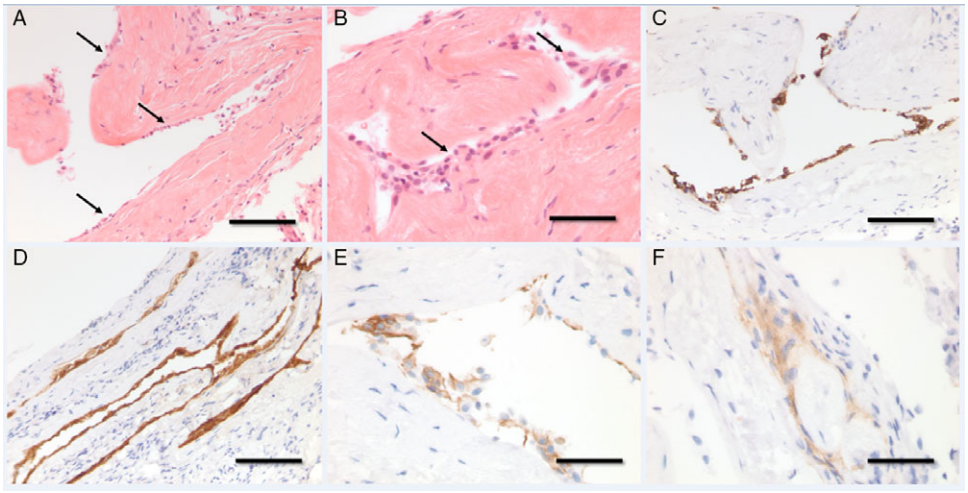

Figure 2: *(A) the cyst wall is collagenous with a delicate lining of simple to low cuboidal cell, H&E, bar = 100 um. (B) The lining is better preserved in small clefts or folds, H&E, bar = 50 um. The lining expresses a number of proteins including: (C) Cytokeratin, immunoperoxidase, bar = 100 um, (D) S-100 protein, immunoperoxidase, bar = 100 um, E) Glial fibrillary acidic protein (GFAP), immunoperoxidase, bar = 50 um, (F) Epithelial membrane antigen (EMA), immunoperoxidase, bar = 50 um.*

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