Interhemispheric cistern lipoma associated with malformations of cortical development, hypogenesis of the corpus callosum, and abnormal vasculature

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To the Editor: Intracranial lipomas (ICLs) are very rare congenital malformative lesions accounting for about 0.1% to 1.7% of all intracranial tumors.[1,2] Herein we describe a rare case of a lipoma affecting the cerebral interhemispheric cisterns in association with malformations of cortical development (MCD), hypogenesis of the corpus callosum, and abnormal vasculature.

A 5-year-old male patient was admitted to our hospital with a prior diagnosis of epilepsy for 2 years, which gradually aggravated even upon treatment with medication in the local hospital. The patient was born after a normal-term pregnancy and uncomplicated delivery and had been healthy except for epilepsy. His neurological examination was unremarkable. Magnetic resonance imaging results showed that the lesion was hyper-intense on both T1 and T2-weighted images, showing an interhemispheric lipoma as well as agenesis of the corpus callosum. Fluid attenuated inversion recovery imaging showed a thick cortex around the interhemispheric lipoma [Figure 1A–E]. The patient underwent surgery after indication assessment. The adipose mass in the interhemispheric cisterns and the left frontal lesions were resected.

The pathological findings showed that the mass was composed of typical adipose tissue, with some blood vessels within the mass, leading to a diagnosis of lipoma. In the brain tissue, mature fat, and blood vessels were visible on the surface of the meninges. The normal structure of the cerebral cortex and the white matter was disrupted by the proliferation of vasculature and fibrous tissue. Some vascular walls were thickened and showed hyaloid degeneration, around which mature fat was also visible. The neurons in the cerebral cortex were arranged in a disorderly pattern. Cortical atrophy and neuronal reduction were visible in a partial region [Figure 1F–K]. Therefore, the diagnosis of interhemispheric cistern lipoma associated with MCD, hypogenesis of corpus callosum, and abnormal vasculature was made.

ICLs are rare congenital malformations. The first description of ICLs was made in the year of 1818 through incidental findings at autopsy.[1] ICLs are commonly thought to be related to the abnormal persistence and mal-differentiation of the meninx primitiva, which is the mesenchymal precursor of the leptomeninges during the development of the subarachnoid cisterns.[3] The majority of the lesions are interhemispheric and more than 50% are located in the callosal cisterns. Other relatively rare localizations can be observed in the quadrigeminal plate, suprasellar/interpeduncular cisterns, the cerebellopontine angle cistern, sylvian cistern, and cortical surface.[1,4]

In the present case, the lipoma was located on interhemispheric cisterns, associated with hypogenesis of corpus callosum, malformations of cortical dysplasia, and abnormal vasculature. ICLs are known to be associated with adjacent hypoplastic corpus callosum. Other malformations associated with ICLs include absence of the septum pellucidum, hypoplasia of the vermis, spinal dysraphism, and myelomeningocele.[2] It is not clear whether cortical dysplasia and lipoma arise together or not and if abnormal vasculature is caused by lipoma or cortical dysplasia. In our case, the vascular and fibrous tissue were proliferated in the cerebral cortex and white matter, which may cause the disruption of the normal structure of the cerebral cortex and the white matter, including the reduction

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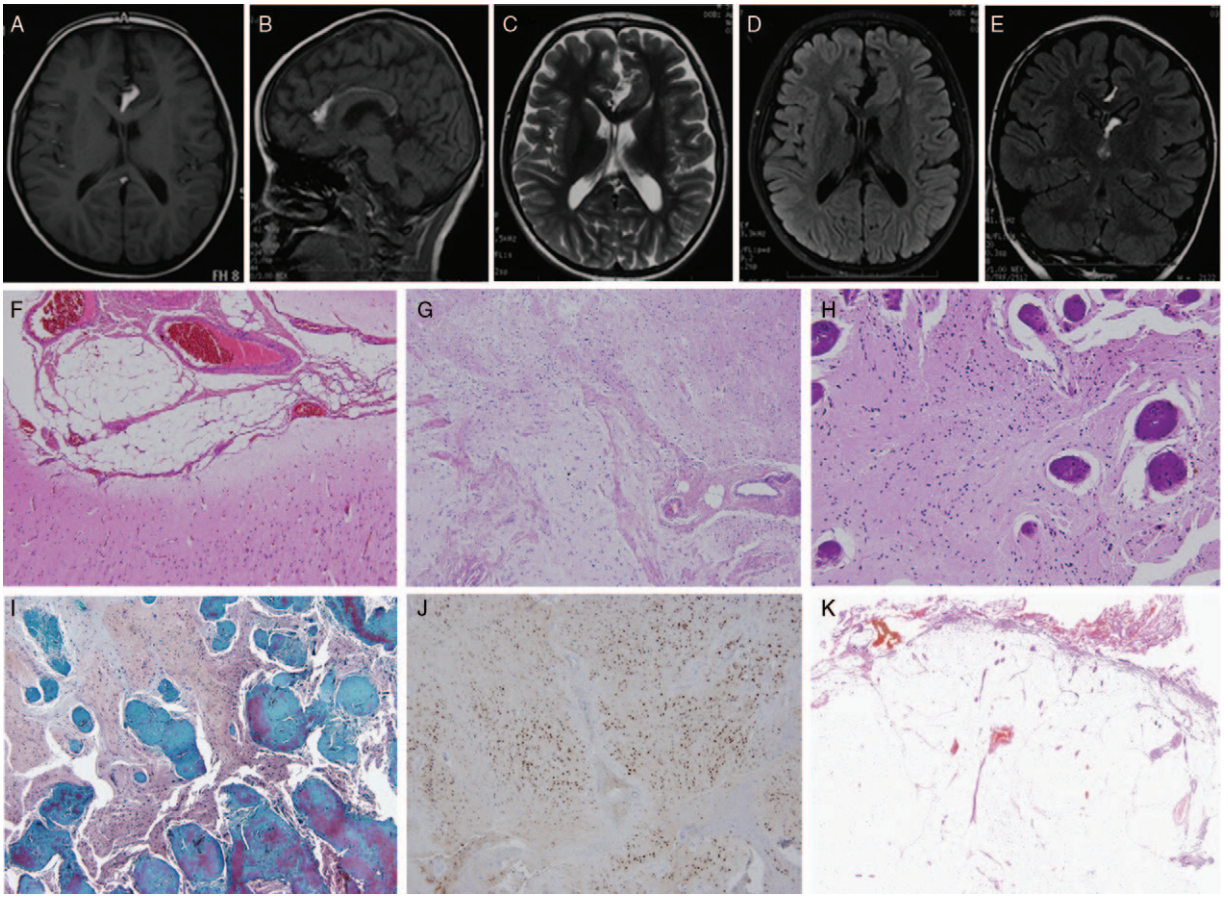

Figure 1: Radiological and pathological features of the patient with interhemispheric cistern lipoma. MRI results revealed T1-hyperintense (A, B), T2-hyperintense (C) signals, while FLAIR (D) showed low intensity. A coronal-uncompressed lipid FLAIR-sequence (E) revealed a high-signal lesion around the corpus callosum and in the cerebral falx with the characteristics of a lipoma. MRI results also showed dysmorphia and hypogenesis of the corpus callosum. (F-K) Histopathological findings of the lipoma and underlying cortex. Mature adipose tissue was visible on the brain surface (F, hematoxylin and eosin staining, original magnification ×100). The normal structure of the cerebral cortex was destroyed with proliferation of fibers and blood vessels (G, hematoxylin and eosin staining, original magnification ×100). Proliferation of large numbers of thick-walled vessels within the brain tissue shown by hematoxylin and eosin staining (H, original magnification ×100) and Masson trichrome staining (I, original magnification ×100). Neuron-specific nuclear protein immunohistochemical staining showed a disorder of neuron arrangement (J, immunohistochemical staining, original magnification ×100). Mature adipose tissue enclosed in a capsule, consistent with a diagnosis of lipoma (K, hematoxylin and eosin, original magnification ×40). FLAIR: Fluid attenuated inversion recovery; MRI: Magnetic resonance imaging.

and derangement of neurons according to the previous report.[4]

To date, the reports on molecular alterations in ICLs were rare. Encephalocraniocutaneous lipomatosis (ECCL) had been described in one patient with a proven neurofibromin 1 (*NF1*) mutation, and it was hypothesized that ECCL may result from somatic mosaicism for a second mutation in the normal *NF1* allele or another gene of the RAS-mitogen-activated protein kinase pathway.[5] However, no pathological molecular alteration, including *NF1*, sprouty-related, EVH1 domain-containing protein 1 gene, and gene of phosphate and tension homology deleted on chromsome ten, was found by next-generation sequencing using the 908 epilepsy-related gene panel in our case. Therefore, further studies need to be performed on ICL.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient's guardians have given consent for his images and other clinical information to be reported in the journal. The guardians understand that his name and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

None.

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