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Case Report

Combined choroid plexus cyst and choroid plexus lipoma in the lateral ventricle causing unilateral ventriculomegaly: a case report

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Abstract: Choroid plexus cysts (CPCs) are the most common neuroepithelial cysts, but rarely present with neurological signs or symptoms. Intracranial lipomas are generally seen at an interhemispheric location, and solitary choroid plexus lipomas (CPLs) have been reported in only a few cases. Here we report a symptomatic case of a 39-year-old woman with combined CPC and CPL on the lateral ventricle. She presented with a headache associated with a mass lesion and multiple cysts on her right lateral ventricle and subsequently developed unilateral ventriculomegaly. The mass demonstrated hypointensity on T1-weighted magnetic resonance imaging (MRI) and homogeneous enhancement on contrast-enhanced MRI. Histopathologically, the resected lesion was composed of multiple cysts lined with a single layer of cuboidal epithelial cells and numerous mature fat cells in the stromal component. The stromal component also included the distinct foci of arachnoid cells, suggesting that mature fat cells are derived from the arachnoid cell in the choroid plexus. To our knowledge, this is the first case of combined CPC and CPL.

Keywords: Choroid plexus cyst, lipoma, hydrocephalus, unilateral ventriculomegaly, neuroepithelial cyst

Introduction

Choroid plexus cysts (CPCs) are commonly found at autopsy and routine fetal anomaly scans using prenatal sonograms, with an incidence of approximately 0.1 to 3% of all pregnancies [1, 2]. Manifestations in children have also been reported, but CPCs are rarely seen in adults. Adult CPCs are commonly encountered at autopsy and some researchers believe that these are the result of cystic degeneration [3]. Although the majority of adult CPCs are asymptomatic, some symptomatic cases have been reported [4-8]. Common symptoms at presentation are typically due to developing ventriculomegaly.

An intracranial lipoma is a rare, benign tumor that accounts for 0.34% of all intracranial tumors [9]. An interhemispheric location is the most common site of this tumor (seen in 40 to 50% of cases), and dysgenesis of the corpus callosum has been observed as an associated anomaly of pericallosal lipomas. The basal cistern and sylvian fissure are the next most frequently affected sites, while lipomas are uncommonly located in the choroid plexus. The majority of choroid plexus lipomas (CPLs) are seen with pericallosal lipomas, and solitary CPLs have been reported in only 9 cases [10-12]. Similar to CPCs, CPLs are usually asymptomatic and are found with concomitant symptomatic anomaly.

Here, we describe a case of combined CPC and CPL on the lateral ventricle causing unilateral ventriculomegaly and conduct a literature review. To our knowledge, this is the first case of combined CPC and CPL.

Results

Clinical summary

A 39-year-old woman presented to our neurosurgical department with increased frequency

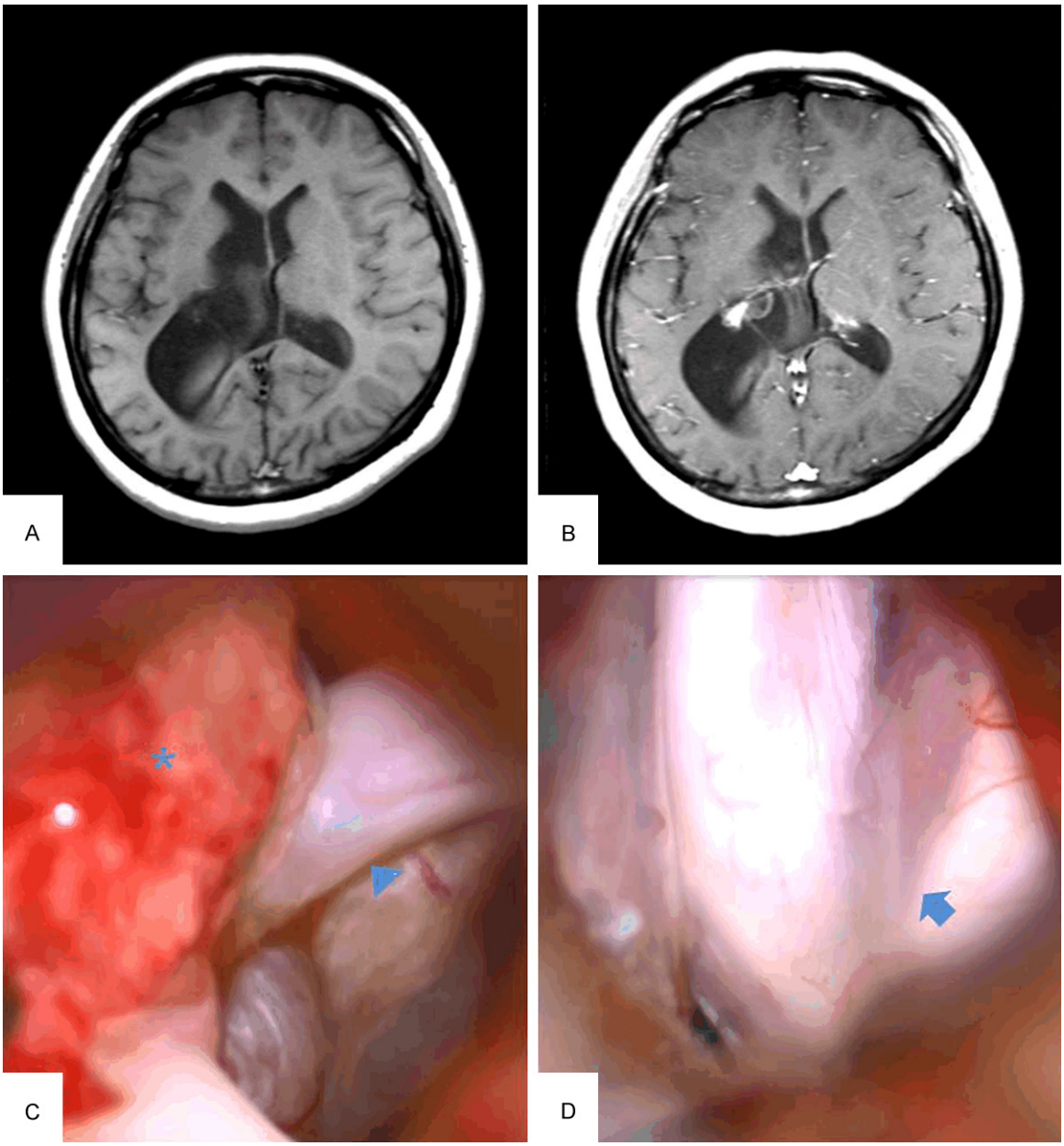

Figure 1. Preoperative magnetic resonance imaging (MRI) and intraoperative images. A. Axial T1-weighted MRI showing a right-sided unilateral ventriculomegaly and a hypointense mass in the right lateral ventricle. B. Gadolinium-enhanced MRI revealing a heterogeneously enhanced mass lesion with multiple cysts and thin bands crossing the right lateral ventricle. C. Intraoperative images showing a reddish tumor mass (asterisk) with cysts and thinning septal walls (arrowhead) attached to the lateral ventricle wall. D. Membranous tissue (arrow) deep inside the intraoperative view interfered with cerebrospinal fluid flow.

of headaches without nausea. Clinical examination was unremarkable. A cranial computed tomography (CT) scan showed a unilateral dilatation of the right lateral ventricle and a low density mass with cysts in the right trigone. Ventricular enlargement was most notable in the posterior and inferior horns of the right lateral ventricle. Gadolinium-enhanced magnetic resonance imaging (MRI) revealed a heterogeneously enhancing 1 × 1 cm mass with multiple cysts in the right trigone of the lateral ventricle. Cystic contents were homogeneously hypointense on T1-weighted and fluid attenuated inversion recovery (FLAIR) images and hyperin-

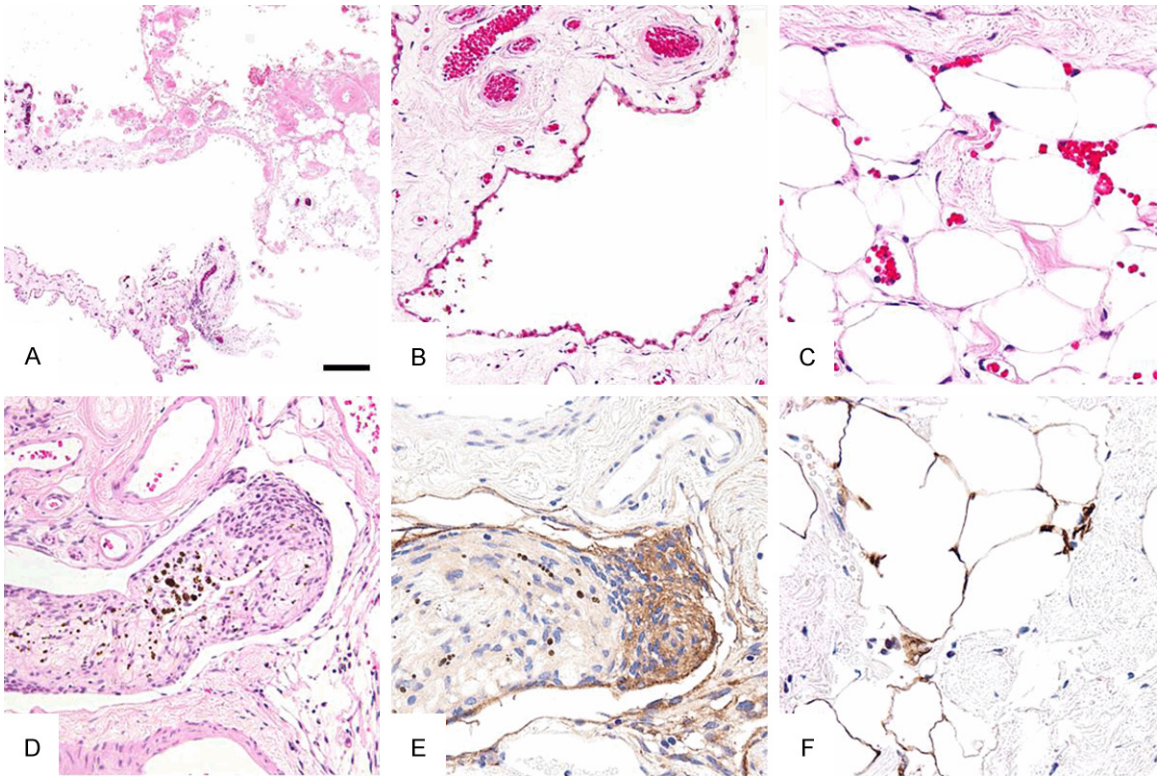

Figure 2. Microscopy findings. Micrograph at higher magnification revealed cysts and inflammatory cell infiltrates, lipid cells, and hyalinized vessels (HE, A). The cyst was lined by a single layer of cuboidal epithelial cells (HE, B). The basal part of the cysts included abundant fat cells with small nuclei (HE, C) and foci of arachnoid cells (HE, D) with immunoexpression of EMA (E). The fat cells expressed fatty acid synthase (F). The scale bar in (A) indicates 500 μm for (A), 100 μm for (B), and 50 μm for (C-F).

tense on T2-weighted images, suggesting that the cystic content contained cerebrospinal fluid (CSF)-like fluid. The mass was heterogeneously hypointense on T1-weighted, T2-weighted, and FLAIR images and isointense on diffusion-weighted images (**Figure 1A**). Thin bands enhanced by gadolinium crossed the dilated right trigone of the ventricle, suggesting they were internal septations within the cysts (**Figure 1B**). Angiography showed a slight tumor stain fed by the posterior choroidal artery.

The patient underwent parieto-occipital craniotomy for the suspected intra-ventricular tumor, and the mass was removed completely. Intraoperative findings demonstrated that a soft, reddish mass with multiple cystic components was firmly attached to the choroid plexus (**Figure 1C**), and a thin cyst wall with septal bands widely expanded in the right trigone of the ventricle. A part of the expanded wall was seated on the neighboring foramen of Monro and obstructed the CSF outflow pathway (**Figure 1D**). This part was only partially removed because of its tight adhesion with the ventricle wall and vessels, which resulted in CSF passage recanalization. The cyst contained a clear colorless fluid. The expanded right lateral ventricular wall was revealed to be harder and thicker than a normal wall. The postoperative course was uneventful, and the frequency of headaches was significantly decreased. Postoperative MRI confirmed the complete removal of the enhancing mass, the thin bands lying in the right trigone of the lateral ventricle, and reduction in ventricular size. The patient remained stable without deterioration in clinical symptoms or recurrence of the lesion on MRI at postoperative month 9.

Pathological features

The resected tissue consisted of multiple cysts, normal choroid plexus, and a stromal component (**Figure 2A**). Multiple cysts were lined by a single layer of cuboidal epithelial cells (**Figure**

2B). The stromal component located on the basal part of the cysts was composed of extensive mature fat cells as well as small to moderate-sized vessels with hyalinization and connective tissues (**Figure 2C**). Neither lined cells nor fat cells showed nuclear or cellular atypia. The stromal component had cell nests composed of meningeal or glial cells (**Figure 2D**). These cells were devoid of typical whorl formation and irregular cytoplasmic processes or nuclear atypia that are characteristic of meningioma and astrocytoma, respectively. There was no histopathological evidence of xanthogranuloma. Immunohistochemical analysis demonstrated that the lined epithelial cells showed immunopositivity for S100 and D2-40 and were weakly positive for the epithelial membrane antigen (EMA), which together with normal morphological findings demonstrated that they were normal choroid plexus epithelial cells. Expression of EMA and D2-40 was also observed in meningeal cells (**Figure 2E**). Glial cells expressed S100 and D2-40. The expression of the S100 protein and fatty acid synthase was found in fat cells (**Figure 2F**). No MIB-1 labeling was observed on these cells. The pathological diagnosis was combined CPC and CPL.

Discussion

CPC represents the most common fetal lateral ventricular mass, but is rarely seen in adults. CPC size is usually less than 2 cm in diameter and CPC is asymptomatic. On MRI, CPC commonly appears as a single cyst or multiple cysts with well-demarcated cystic walls that demonstrate contrast enhancement. The content fluid of the CPCs is similar to CSF, and it occasionally appears with higher signal intensity than CSF on MRI, indicating higher protein content [13]. The neuroimaging features of the presented case corresponded to CPC, except for the homogeneously enhanced nodule, which was assumed to be a tumor. Primary lateral ventricle tumors are relatively rare. The most common lateral ventricle tumors are meningioma, pilocytic astrocytoma, choroid plexus papilloma, oligodendroglioma, ependymoma, central neurocytoma, and subependymal giant cell astrocytoma. The majority of these tumors rarely accompany a cystic lesion. A lipoma in the ventricle is much rarer than the above tumors. Senser et al. reported 7 cases of solitary CPL, and all were asymptomatic and located in the trigone of the lateral ventricle without associated anomalies [11]. No cases displayed cystic formation. Intracranial lipoma generally appears hyperintense on T1-weighted images and shows no enhancement on contrast-enhanced MRI, whereas our case showed hypointensity on T1-weighted images with contrast enhancement. The MRI findings of our case may be due to the abundant vessels and the presence of cell nests composed of meningeal and mesenchymal cells.

A unilateral ventriculomegaly is often caused by a CPC on the neighboring foramen of Monro and contributes to the clinical presentation [5, 14, 15]. Our operative findings confirmed that the expanded cyst wall neighboring the foramen of Monro was thicker than that of the posterior horn side and stuck to surrounding structures. Although mobile cysts acting as a ball valve are often reported as the cause of unilateral ventriculomegaly, the stuck cyst wall directly blocked the CSF flow in our case [15].

The diagnosis of combined CPC and CPL was based on the presence of multiple cysts lined with a single layer of cuboidal epithelial cells and abundant mature fat cells in the stromal component. The stromal component included arachnoid cells, but the lack of features of the meningioma in them indicated that the fat cells were not part of metaplastic meningiomas. Most researchers believe that CPCs are formed by an infolding of the epithelium into the stroma [16]. On the other hand, CPLs are accepted to be due to an abnormal differentiation of the primitive meningeal tissues [17]. Several reports have illustrated the presence of foci of arachnoid cells in the normal choroid plexus stroma from which intraventricular meningiomas arise [18-20]. Microscopy of CPCs has generally revealed chronic inflammatory cell infiltrates cholesterol clefts, hemosiderin, and calcium; however, mature fat cells have been never observed in the normal choroid plexus or stromal component of CPC. The presence of extensive mature fat cells provided sufficient evidence for the diagnosis of lipoma in our case. Histopathology implicated the possibility that mature fat cells are derive from the arachnoid cells. The uncommonly differentiated fat cells might be associated with an infolding of the epithelium into the stroma and the consequent cyst formation.

We described the first case of a combined CPC and CPL in the lateral ventricle causing unilateral ventriculomegaly. The neuroimaging findings differed from typical cases of intracranial lipoma, and we suggest that CPL should be considered in the differential diagnosis of uncommon choroid plexus tumors. The histopathology in our case implied that the lipoma differentiated from meningeal cells in the choroid plexus.

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Disclosure of conflict of interest

None.

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