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CASE REPORT

Suprasellar arachnoid cyst due to ectopic choroid plexus: case report

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Abstract
Arachnoid cysts (ACs) are malformations that account for about 1% of all intracranial lesions. The aetiology and progression of these lesions have been debated, with one possible explanation being the production of cerebro-spinal fluid (CSF) by ectopic choroid plexus (CP). To our knowledge, only seven cases of ACs incorporating CP have been reported, and we believe this to be the first reported case of a suprasellar AC containing ectopic CP. A 1-year-old boy presented with developmental delay and macrocephaly. MRI scan revealed hydrocephalus due to a suprasellar AC. An endoscopic ventriculocisternostomy was undertaken. Intra-operatively, intra-cystic, pink frond-like tissue resembling choroid plexus was identified. Histologically, the cyst wall was composed of fibrous tissue, with layered arachnoid cells, while the frond-like tissue was composed of papillary structures in keeping with normal choroid plexus tissue. We postulate that the rest of the ectopic CP may have been trapped within the double layered arachnoid fold of the diencephalic leaf of Liliequist's membrane which may drive the formation and development of certain suprasellar ACs.

Keywords Arachnoid cyst · Suprasellar · Ectopic choroid plexus · Ventriculostomy

Introduction

Arachnoid cysts (ACs) are malformations that account for about 1% of all intracranial lesions. The aetiology and progression of these lesions have been debated, with the production of cerebro-spinal fluid (CSF) by ectopic choroid plexus (CP) proposed as a possible mechanism. This is, to our knowledge, the first reported case of a suprasellar arachnoid cyst containing ectopic CP. Herein, we present our case and review the literature to discuss the pathophysiological mechanisms that are thought to lead to this rare occurrence.

Case report

A 1-year-old boy presented with delayed neurological development and macrocephaly. There was no history of preceding trauma or infection. Visual testing was normal. Endocrine evaluation revealed hypothyroidism. MRI scan revealed hydrocephalus due to a suprasellar mass. The mass was in keeping with a suprasellar AC, measuring 3.45 (AP) by 3 cm (TV) by 3 cm (CC). The cyst extended into the base of the sella turcica. T1 and T2 MRI sequences (Fig. 1) were in keeping with that of CSF signal. The anterior wall of the cyst demonstrated a "nodule" of hypodense, non-enhancing tissue on both T1 and T2 sequences. Given the presence of hydrocephalus and endocrine abnormalities, an endoscopic ventriculocisternostomy was proposed and undertaken. Intra-operatively, the cyst roof was bulging through the foramen of Monroe. Following fenestration of the roof of the cyst, intra-cystic, pink frond-like tissue resembling CP was identified. The floor of the cyst was then widely fenestrated, caudal to which an incomplete/perforated mesencephalic leaf of Liliequist's membrane could be identified along

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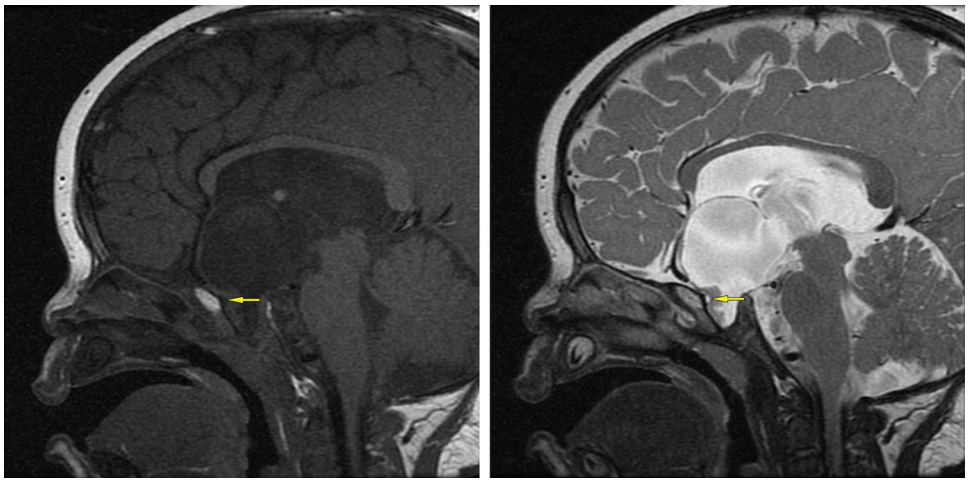


Fig. 1 Left: T1 MRI sequence demonstrating a large suprasellar cyst. Arrow: small nodule of hypodense tissue within the cyst. Right: T2-weighted MRI sequence. Arrow: hypodense tissue within the suprasellar cyst

with neurovascular structures coursing through the prepontine cistern (see Fig. 2). The cyst wall and ectopic tissue were biopsied. Histologically the cyst wall was composed of fibrous tissue, with layered arachnoid cells, while the frond-like tissue was composed of papillary structures lined by exuberant cuboidal epithelium, in keeping with normal CP tissue. (See Figs. 3 and 4) The child had an uneventful post-operative course. Follow-up MRI revealed resolution of his hydrocephalus with partial collapse of the cyst. The child displayed significant improvement in his neurodevelopment. His endocrine function however did not improve.

Discussion

The majority of ACs are primary lesions, of congenital origin as a result of anomalous splitting and duplication of the endomenix during the early stages of embryonal development of the arachnoid pathways [3, 15]. Histological findings in these lesions demonstrate splitting or duplication of the arachnoid membrane into an inner and outer leaflet at the edge of the cyst cavity [5, 28].

Secondary, or acquired, ACs are less common, and occur [3, 25] as a result of trauma, surgery, infection, or intracranial haemorrhage [16, 20, 25]. Suprasellar ACs in particular have been associated with trauma in infancy [14]. Fox and Al-Mefty [7] were the first to propose that ACs may develop in the suprasellar area due to the occlusion of the normal perforations in Liliequist's membrane, "by adhesions after subarachnoid haemorrhage, infection and other causes of inflammation". Choi and Kim [3] speculate that trauma may rather induce an abnormal splitting or duplication of the immature endomenix in infancy, given that the subarachnoid space is not yet fully developed, with areas still "closed without cerebrospinal fluid".

The mechanism by which ACs expand is poorly understood [15], with several postulates, including osmotic gradients caused by higher protein or sodium content in the cyst, [23] one way valve mechanisms, [11, 13, 29] direct secretion of fluid by arachnoidal cells lining the cyst wall [8], or by fluid production by ectopic CP [3, 12].

To our knowledge, only seven cases of ACs incorporating CP have been reported [12, 26, 27]. In Schummann et al.'s [26] review of these cases, however, they noted that four of

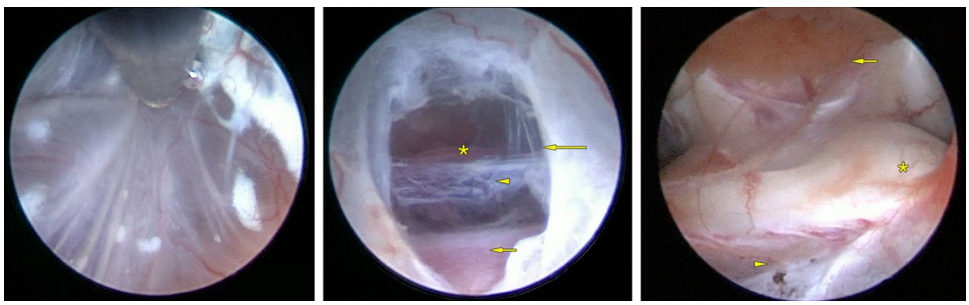


Fig. 2 Left: Endoscopic view from within lateral ventricle revealing roof of arachnoid cyst. Middle: following fenestration of both the roof and floor of the cyst. Asterisk: contents of prepontine cistern. Arrowhead: incomplete mesencephalic leaf of Lilliquist's membrane. Small arrow: basilar tip behind the posterior wall of the cyst. Large arrow: septostomy in the floor of the cyst. Right: visualization of contents of cysts. Arrow: large tuft of normal appearing choroid plexus. Asterisk: posterior clinoid process. Arrowhead: floor of suprasellar cyst

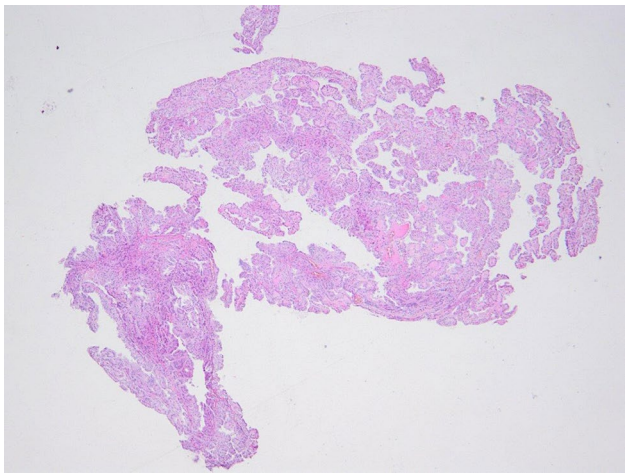


Fig. 3 A low-power view demonstrates numerous finger-like projections lined by cuboidal epithelium. (H&E 40×)

these described cysts were possibly lined with epithelium of ependymal origin, thus making the present case the fourth reported case of an AC arising due to entrapped CP, and the first case description of a suprasellar AC containing ectopic CP.

As postulated by Schummann et al., [26] two scenarios exist to explain this unique situation: either the ectopic CP is incorporated into an already developing AC, or the CSF production from the ectopic CP becomes entrapped between two layers of arachnoid which then induces membrane splitting and cyst growth.

The diencephalic leaf of Liliequist's membrane has been shown by Zhang [30] to be a double layered fold of arachnoid mater, which can allow for the formation of a truly intra-arachnoid diverticulum [17]. We postulate that the rest of the ectopic CP may have been trapped within this intra-arachnoidal diverticulum and, following CSF production by the ectopic tissue, formed a truly intra-arachnoid cyst. This morphology would be in keeping with a suprasellar type 1 AC according to the classification of Andre et al. [1]. The position of the basilar artery bifurcation behind the posterior wall of the cyst and the clinical presentation of hydrocephalus are typical for the type 1 suprasellar AC [1, 21].

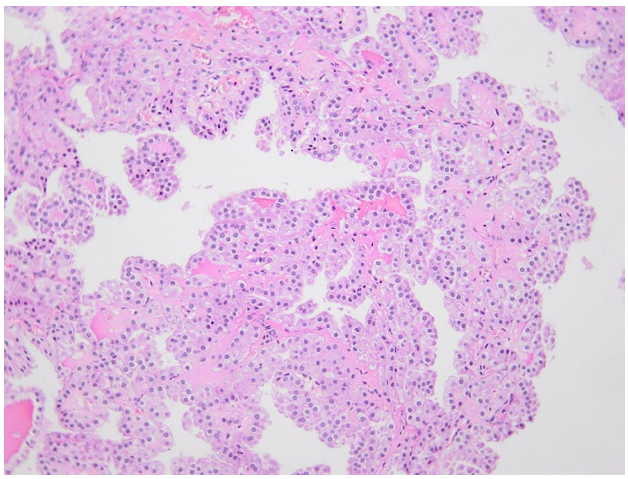


Fig. 4 High-power view (H&E 200×), demonstrating arachnoid villi lined by cuboidal epithelium compatible with choroid plexus

The presence of CP in an extra-ventricular location is thought to occur as a result of an abnormality in embryonic development from either anomalous migration of CP tissue or metaplasia from ependymal rests [2], and has been used to explain the occurrence of extra-ventricular CP cysts [9], choroid papillomas [2, 18, 24], and ACs [26].

In the current case, given the hydrocephalus and neurodevelopmental delay, surgery would generally be indicated. In asymptomatic cases, however, or cases in which there is only endocrine dysfunction or isolated gaze disturbances, several authors [4, 10] have suggested a conservative approach as many of these cysts will not evolve and may even resolve spontaneously [6, 19, 22]. We postulate that given the presence of CSF producing CP within the cyst, progression is likely and that as suggested by Shuman et al. [26] in cases of reclosure of a fenestrated cyst, ectopic CP tissue would predispose to a faster recurrence.

Conclusions

The mechanism by which ACs expand is poorly understood, and fluid production by ectopic CP should be considered as an aetiological factor. We suggest that during the endoscopic treatment of suprasellar ACs, a careful inspection of the cyst cavity is performed as even small pieces of ectopic choroidal tissue may drive the progression or rapid recurrence of suprasellar ACs.

Declarations

Ethics approval The Human Research Ethics Committee of the University of the Witwatersrand approved the presentation of the case report.

Conflicts of interest The authors declare no competing interests.

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