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Case Report

Bilateral Middle Cranial Fossa Arachnoid Cysts With Temporal Lobe Agenesis. Case Report In Adult.

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ABTRACT

Arachnoid cysts (ACs) represent approximately 1% of all intracranial space-occupying lesions. They are benign collections of fluid that develop within the arachnoid membrane, containing clear, colorless fluid resembling CSF. A 21-year-old man was admitted in consultation for some headaches. On admission, he was in good health. The neurological evaluation was normal. No neurocognitive disorders were noted, and routine laboratory investigations were within the normal range. The standard neuropsychological assessment was normal. The electroencephalogram was normal. Eye fundoscopy showed no papilledema. Bilateral middle cranial fossa arachnoid cysts with temporal lobe agenesis in adults are very rare. The diagnosis is made by a brain MRI. Prophylactic surgery in asymptomatic cases is not advisable. These patients must be followed by the practitioner.

Key Words: Arachnoid cysts, CSF, papilledema, MRI, neuropsychological assessment

INTRODUCTION

Arachnoid cysts (ACs) represent approximately 1% of all intracranial space-occupying lesions. They are benign collections of fluid that develop within the arachnoid membrane, containing clear, colorless fluid resembling CSF. Most of the time, they appear as single, sporadic lesions. The majority of individuals where ACs have been identified are male, and the middle fossa is the most common location [1, 2, 3].

Bilateral temporal ACs are rare. Bilateral temporal lobe agenesis associated with middle cranial fossa arachnoid cysts, on the other hand, is an exceedingly rare condition, and only a few cases have been reported so far [4–7]. A case is reported.

CASE REPORT

A 21-year-old man was admitted in consultation for some headaches. On admission, he was in good health. The neurological evaluation was normal. No neurocognitive disorders were noted, and routine laboratory investigations were within the normal range. The standard neuropsychological assessment was normal. The electroencephalogram was normal. Eye fundoscopy showed no papilledema.

Axial brain T1-weighted MRI sequence shows a bilateral middle cranial fossa cystic lesion CSF-like signal with temporal lobe agenesis. Transversal MRI section, T2-weighted sequence showing bilateral middle cranial fossa ACs. Sagittal MRI section, T1-weighted sequence showing temporal arachnoid cyst with temporal lobe agenesis. (**Figure1**). The diagnosis was bilateral middle cranial fossa ACs with temporal lobe agenesis. The patient was managed conservatively. The patient and his family were sensualized and informed of some precautions, especially brain-head trauma. With a 3-year follow-up, he was satisfied and presented only some headaches in physical effort.

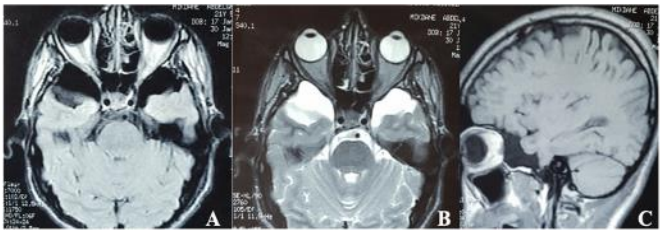

Figure 1: **(A)** axial brain T1-weighted MRI sequence showing bilateral middle cranial fossa arachnoidal cysts. **(B)**Axial MRI section, T2-weighted sequence showing bilateral middle cranial fossa arachnoid cysts. **(C)** Sagittal MRI section, T1-weighted sequence showing temporal arachnoid cyst with temporal lobe agenesis. Bilateral middle cranial fossa arachnoid cysts with temporal lobe agenesis in 21-year-old man.

DISCUSSION

In 1964, Robinson described ten cases with unilateral temporal lobe defects and coined the term 'temporal lobe agenesis syndrome' [2]. Bilateral temporal lobe agenesis associated with middle cranial fossa ACs is an exceedingly rare condition,

and only a few cases have been reported in literature [1,4,5,6,7]. Most ACs are found in the first decade of life. They represent 1% of all intracranial lesions, and in recent years, with the development of neuroradiology, the clinical detectability of ACs has increased. Amelot A. and Al. retrospectively reviewed 240 temporals intracranial ACs managed over 25 years in their pediatric neurosurgical unit. No bilateral temporal ACs were reported in this casuistic. The ACs are typically considered congenital lesions [1], but a small proportion is believed to be secondary to trauma, inflammation, or bleeding [1-4, 8, 9, 10].

Two hypotheses are reported: the first stipulates that middle cranial fossa cysts represent a passive collection of cerebrospinal fluid related to temporal lobe agenesis. The second theory considers temporal lobe defects as a consequence of the compression of ACs as the primary developmental malformation of the arachnoid membrane.

Arachnoid cysts are fluid-filled sacs that grow on the brain and spine. They are not tumors, and they are not cancerous [1]. Most cases of middle cranial fossa cysts are asymptomatic. Most ACs appear at birth or after childhood head trauma, and the vast majority of cysts don't cause symptoms. Children can exhibit signs of increased intracranial pressure, seizures, and focal or cognitive neurological deficits, depending on the cyst's location. Acute clinical symptoms can be caused by rupture or bleeding inside the cyst [1, 5, 8, 10]. Middle fossa arachnoid cysts in this area of the brain can cause problems with vision, hearing, movement, and balance. They can lead to fatigue, weakness, or paralysis, usually on one side of the body. Some children have neurological symptoms. These may include delays in development and changes in behavior. Vision and hearing abnormalities and behavioral changes are less common [5].

Neuroimaging: Diagnostic tools for the investigation of ACs are computerized tomogram (CT) with contrast studies and brain MRI. On CT, ACs are observed as extra-axial cysts with the density of CSF, without any contrast enhancement. Adjacent calvarial remodeling is common, as is a hypoplasia of the adjacent brain parenchyma, especially in middle cranial fossa ACs. Brain MRI signals are similar to CSF in T1- and T2-weighted imaging, with no enhancement on gadolinium with the normal signal from the contiguous. Rarely, radioisotope scintigram and cine phase-contrast MR imaging were used.

The ACs are divided into types I–III according to the Galassi classification [9]. Type I: small, spindle-shaped, limited to the anterior aspect of the temporal fossa. Type II: medium-sized lesion, shape roughly triangular or quadrangular, with a straight inner margin. It occupies the anterior and middle parts of the temporal fossa and extends superiorly along the Sylvian fissure. Type III. The lesion occupies the temporal fossa almost entirely and extends over a wide area of the cerebral hemispheres, splitting the opercula of the Sylvian fissure. The temporal lobe is severely atrophic.

Management: Patients who do not demonstrate signs of increased intracranial pressure or focal neurological signs are also considered for conservative management [10]. Some studies claim that ACs are to be regarded as congenital findings that should not be surgically treated unless they cause severe symptoms [5]. While others suggest a connection between temporal ACs and neuropsychiatric symptoms, motivating surgical treatment to possibly alleviate these symptoms [9, 11, 12].

It is largely acknowledged that surgery is not indicated for asymptomatic patients with incidental findings, whereas patients with clear signs of raised ICP or temporal seizures are candidates for surgery. Untreated, symptomatic ACs can lead to permanent brain damage, severe pain, and movement disorders. Blood vessels on the cyst wall can tear and bleed into the cyst, which can make it grow larger. When blood vessels tear and blood pools outside of the cyst, a hematoma can form [7]. Leaking fluid: If trauma or injury damages the cyst, a CSF leak can result. The fluid can leak into other parts of the brain, causing severe damage. Different procedures are performed, including cyst excision, stereotactic aspiration, cyst fenestration, cystocisternostomy, ventriculocystostomy, and cystoperitoneal shunt treatment [1, 3, 8]. Considering the developing CNS during childhood, reductions of large space-occupying ACs followed by restorations of the structural integrity of the developing brain are very desirable.

CONCLUSION

Bilateral middle cranial fossa arachnoid cysts with temporal lobe agenesis in adults are very rare. The diagnosis is made by a brain MRI. Prophylactic surgery in asymptomatic cases is not advisable. These patients must be followed by the practitioner. The patient and his family must be

sensualized and informed of some precautions, especially brain-head trauma.

Conflict of interest - nil

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