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Clinical case

Midline retrocerebellar arachnoid cyst in a young adult. Case presentation

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SUMMARY

Introduction: Arachnoid cysts are benign extra-axial fluid collections indistinguishable from cerebrospinal fluid. They constitute 1% of non-traumatic intracranial space-occupying lesions. Most are located supratentorially, are diagnosed incidentally during childhood, and do not produce clinical manifestations.

Case presentation: A 27-year-old female patient presented with a headache accompanied by vision loss and gait instability. Physical examination revealed truncal ataxia, increased height of support, and incipient papilledema. A non-contrast and contrast-enhanced computed tomography (CT) scan of the head showed a retrocerebellar space-occupying lesion with a cystic appearance, no contrast enhancement, and a mass effect causing obstructive hydrocephalus. An arachnoid cyst was suspected. The patient underwent



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surgery involving fenestration, partial resection of the cyst walls, and communication of the cyst with the subarachnoid space. The symptoms resolved postoperatively. A follow-up CT scan showed complete resolution of the cyst. The patient recovered well without complications. Histopathological examination confirmed the diagnosis of an arachnoid cyst. There was no recurrence of the lesion.

Conclusions: Although most patients with arachnoid cysts remain asymptomatic, some may present with complex manifestations depending on the location and size of the lesion. Infratentorial cysts frequently cause hydrocephalus with secondary intracranial hypertension. Other cystic lesions should be considered in the differential diagnosis. While endoscopic techniques are currently the gold standard, open surgery remains effective in treating these lesions.

Keywords: Arachnoid cyst; Infratentorial; Ataxia; Papilledema; Hydrocephalus; Fenestration.

ABSTRACT

Introduction: Arachnoid cysts are benign extra-axial collections containing fluid that is indistinguishable from cerebrospinal fluid. They account for 1% of non-traumatic intracranial space-occupying lesions. Most are located supratentorially, are diagnosed incidentally during childhood, and do not produce clinical symptoms.

Case presentation: A 27-year-old female patient presented with headache, accompanied by visual loss and gait instability. Physical examination revealed truncal ataxia, a widened base of support, and incipient papilledema. A simple and contrast-enhanced computed tomography (CT) scan of the brain showed a retrocerebellar space-occupying lesion with cystic appearance, no contrast enhancement, and a mass effect causing obstructive hydrocephalus. A diagnosis of arachnoid cyst was considered. She underwent surgery, including fenestration, partial resection of the cyst walls, and communication with the



subarachnoid space. Symptoms resolved after surgery. Follow-up CT showed disappearance of the cyst. The patient recovered favorably without complications. Pathological study confirmed the diagnosis of arachnoid cyst. No recurrence was observed.

Conclusions: Although most patients with arachnoid cysts remain asymptomatic, some may present complex symptoms depending on the location and size of the lesion. Infratentorial cysts frequently cause hydrocephalus with secondary intracranial hypertension. Differential diagnosis should include other cystic lesions. Although endoscopic techniques are currently the standard, open surgery remains effective for treating these lesions.

Keywords: Arachnoid Cyst; Infratentorial; Ataxia; Papilloedema; Hydrocephalus; Fenestration

SUMMARY

Introduction: The arachnoid cysts have benign extra-axial collections that contain a liquid indistinguishable from the liquid. We represent 1% of non-traumatic intracranial expansive lesions. Most of it is located supratentorially, and is diagnosed incidentally in childhood and does not produce clinical manifestations.

Case report: Female patient, 27 years old, with symptoms of headache, loss of vision and instability when walking. On physical examination, he presented truncal ataxia, increased base of support and incipient papilledema. A simple computerized tomography (CT) of the skull with contrast revealed retrocerebellar lesions with a cystic appearance, without contrast enhancement, with mass effect causing obstructive hydrocephalus. Suspeitou-se of arachnoid cyst. The patient was subjected to surgery with fenestração, partial resection of the walls and communication of the cyst with the subarachnoid space. After the procedure, the symptoms will disappear. An evolutionary CT scan showed the



disappearance of the cyst. The patient evolved without complications. The pathological examination confirmed the diagnosis of arachnoid cyst. There is no trace of the injury.

Conclusions: Most patients with arachnoid cysts remain asymptomatic, some may present complex manifestations depending on the location and size of the lesion. Infratentorial cysts frequently cause hydrocephalus with secondary intracranial hypertension. Other cystic lesions should be considered a non-differential diagnosis. Despite endoscopic techniques being currently the best, open surgery continues to be effective in treating these injuries.

Keywords: Arachnoid Cystus; Infratentorial; Ataxia; Papilledema; Hydrocephalus; Fenestration.

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Introduction

Arachnoid cysts are benign, extra-axial, intra-arachnoid collections with walls formed by thickened arachnoid membranes and containing a fluid indistinguishable from cerebrospinal fluid (CSF). They represent approximately 1% of all non-traumatic intracranial space-occupying lesions. (1) Most are congenital, the true arachnoid cysts, and result from developmental abnormalities of the arachnoid. (1,2) Some develop as a result of other injuries and are called secondary arachnoid cysts. (1-4)

Regarding the theories that explain the formation of these lesions, the embryonic theory is the most widely accepted. (1,4) Also mentioned are the foldings and thickenings of the arachnoid, the association with hereditary genetic diseases, (1) focal brain hypoplasia, alterations in the secretion of substances from the subarachnoid space (ESA) to the



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venous system, (4) rupture or tearing of the arachnoid material during gestation, as well as the entrapment of remnants of the choroid plexus, arachnoid granulations, or neuroepithelium. (1)

The clinical manifestations in patients depend primarily on their age and the location of the cyst. In adults, they may present with a seizure, balance and gait disturbances, manifestations of intracranial hypertension (ICH), hearing impairment, visual disturbances, and vertigo. Diagnosis can be made prenatally using ultrasound. In neonates and infants, transfontanellar ultrasound is a reliable method. Computed tomography (CT) allows for defining the location of the cyst, its relationship to neurovascular structures, the presence of hydrocephalus, and the degree of compression resulting from the lesion. Currently, magnetic resonance imaging (MRI) is the diagnostic method of choice. (1-4) Treatment is divided into two main modalities: conservative for asymptomatic patients and surgical for those with clinical manifestations. (1,4,5)

This article aims to present to the scientific community and discuss the fundamental elements of a surgical case of an intracranial arachnoid cyst.

This article will serve as a reference for the management of this disease, as it discusses its fundamental aspects. A surgical case of this rare disease, which is usually asymptomatic and requires no treatment, is reported. This is a successful case treated without the use of advanced surgical techniques, resulting in complete postoperative resolution of clinical manifestations and disappearance of the lesion.

Case presentation

History of the present illness

A 27-year-old female patient, right-handed, of mixed race, from an urban area, with no apparent medical history, presented to the emergency department with a four-week history of holocranial, throbbing headache that worsened with physical exertion and was



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difficult to control with analgesics (paracetamol and dipyrrone). She also reported vision loss and gait instability. She did not experience disorientation, fever, seizures, memory loss, or muscle weakness. She had no relevant family history.

Physical examination positive

BP: 120/80 mmHg, HR: 83 beats per minute, RR: 15 breaths per minute. Painful expression. Conscious. Oriented to time, place, and person. Clear and coherent speech. Attention and memory intact. No long tract motor deficits. No nuchal rigidity. Pupils isocoric and reactive to light, truncal ataxia, increased height of support, and incipient papilledema. No other signs of focal neurological deficit. GCS 15/15 points

Diagnostic evaluation

Hematological studies were performed and showed no abnormalities: hemoglobin 115 g/L, white blood cell count $6.5 \times 10^9/L$, erythrocyte sedimentation rate 10 mm/h, and platelet count $250 \times 10^9/L$. A non-contrast and contrast-enhanced CT scan of the head (Figure 1) revealed a midline, retrocerebellar, infratentorial space-occupying lesion (SOL) measuring 42 x 37 mm in diameter. The lesion was hypodense with cerebrospinal fluid attenuation (CSF) (10 HU), with regular and well-defined borders. It exhibited a mass effect, compressing and displacing the fourth ventricle anteriorly, causing obstructive hydrocephalus. There was no perilesional edema or nodular lesion. No enhancement was observed after contrast administration. A brain MRI was not performed due to unavailability.





Fig.1. Plain CT scan of the skull. Cystic lesion of the posterior fossa. Obstructive hydrocephalus.

A differential diagnosis was performed to rule out other cystic lesions of the posterior fossa. A neoplastic lesion was ruled out: these are not common in the patient's age, the lesion did not present with a mural nodule, associated edema, or contrast enhancement. Megacisterna magna was ruled out because it does not present with hydrocephalus, and Dandy-Walker syndrome was ruled out because there was no cystic dilation of the fourth ventricle or agenesis of the cerebellar vermis. A retrocerebellar infratentorial arachnoid cyst of the midline was considered as the presumptive diagnosis.

Therapeutic intervention

The patient was admitted to the Neurosurgery service. Given her clinical and hemodynamic stability, acetazolamide 250 mg orally was administered every 8 hours. The following day, she underwent surgery. A medial suboccipital approach was performed via bilateral suboccipital osteoclastic craniotomy (Figure 2) with the patient in the prone position. A Y-shaped durotomy was performed with electrocoagulation of the occipital sinus. Once the occipital surface of the cerebellar hemispheres was exposed, a thin-walled, transparent cystic lesion was observed in the midline (Figure 3). The cyst was fenestrated, draining the clear, colorless fluid, and partially resecting its posterolateral walls and opening it to the subarachnoid space (Figure 4).



Fig. 2. Marking of the midline occipitocervical incision (left). Exposure of the external surface of the occipital squama (center). Bilateral suboccipital osteoclastic craniotomy (right).

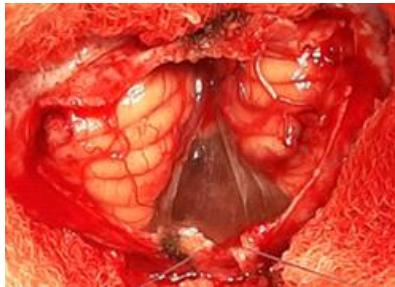


Fig. 3. Y-shaped durotomy. Dural traction points. Occipital surface of cerebellar hemispheres laterally and arachnoid cyst in the center.

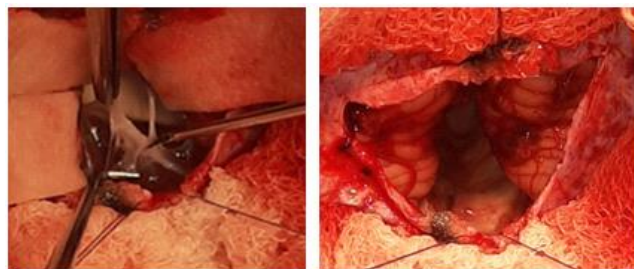


Fig. 4. Fenestration of the cyst with partial resection of its posterolateral wall (left). Cavity after resection (right).

Monitoring and results



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The postoperative course was favorable, with regression of symptoms and no complications. Postoperative neuroimaging studies confirmed the disappearance of the cyst, decompression of the posterior fossa structures, and restoration of cerebrospinal fluid dynamics without hydrocephalus. (Figure 5) The patient was successfully reintegrated into society. Follow-up visits have shown no recurrence of the cyst.



Fig. 5. Postoperative non-contrast CT scan of the skull. Disappearance of the lesion. No hydrocephalus.

Ethical aspects

The study adhered to the ethical codes set forth in the Declarations of Helsinki and Nuremberg. Data confidentiality was protected, and no experimental therapeutic interventions were performed.

Discussion

The first description of cerebral arachnoid cysts was made in 1831 by Richard Bright, who described them as serous cysts associated with the arachnoid membrane. Primary (congenital) arachnoid cysts are present at birth and result from developmental abnormalities in the brain that occur during the first weeks of gestation. (1,4,6,7) The



embryonic theory explains that during hydric dissection (a normal process in arachnoid development), there is incomplete separation within the mesoderm. This leads to the formation of isolated compartments delimited by intra-arachnoid septa. Cerebrospinal fluid (CSF) becomes trapped within these compartments. (1,4) This is the most widely accepted theory of arachnoid cyst formation and offers an explanation for their origin in almost all locations, including the case presented.

Another theory that could also explain the lesion in this case is focal brain hypoplasia. According to this theory, the ESA occupies the space left by an area of brain tissue that does not develop normally. One indication of this is the fact that, after evacuation of the cystic contents, there is no collapse of the cavity, suggesting that the surrounding brain may be hypoplastic. Furthermore, giant posterior fossa cysts are often accompanied by an elevated tentorium, straight sinus, and torcula. (1,4) In the present case, we did not observe immediate collapse of the cavity after decompression. However, the restoration of CSF circulation and the disappearance of the postoperative cavity observed after surgery demonstrate that a compressive effect existed, regardless of the possibility of hypoplasia in the surrounding cerebellar tissue.

Approximately two-thirds of intracranial arachnoid cysts are located in the supratentorial space, most of them in the temporal fossa. In the case analyzed, the lesion is located in the posterior fossa, where the remaining third is found. In this latter compartment, those located in relation to the vermis and cisterna magna are most frequent (up to 12% of the total). They can also be located, in descending order of frequency, in the cerebellopontine angle, the quadrigeminal plate, and the prepontine space. (1)

Most arachnoid cysts are diagnosed in the first two decades of life, particularly in the first two years and during preschool age. There is a male predominance (males are four times more likely to have arachnoid cysts than females). (1,2,8) The diagnosis is almost always made incidentally. (1,2) In adults, there is also a higher prevalence in males. At this age,



middle fossa arachnoid cysts are the most frequent and predominate on the left side. The significant technological development of recent years and the increasing access to imaging studies (CT and MRI) have led to an increase in the diagnosis of arachnoid cysts. Consequently, some authors consider that they constitute more than 1% of intracranial space-occupying lesions. (9)

We present a case of a symptomatic infratentorial arachnoid cyst in a young woman. Given that these are considered congenital lesions, and that symptoms are essentially caused by irritation and/or compression of adjacent structures, we propose that cyst growth occurred in this case. Several mechanisms explain this phenomenon. The most widely accepted, demonstrated in vivo, is the valvular mechanism: fluid enters the cyst from the upper esophageal sphincter (UES) during cardiac systole without outlet. (1,4,8) In this case, we cannot rule out the possibility that an external event caused decompensation of the lesion, either by activating any of the growth mechanisms or by causing the loss of communication between the cyst and the UES. In any case, the space conflict in the posterior fossa was not old: there was no evidence of bone deformity, and the patient was asymptomatic.

Most arachnoid cysts remain stable throughout life. (1-7) It is unusual for a cyst diagnosed incidentally in an adult to grow or produce any symptoms. (9) The development of symptoms secondary to infratentorial arachnoid cysts is uncommon in young people and adults. (8) In adults, the symptoms and signs can be difficult to relate to the cyst itself and depend, as mentioned above, on its location. (1-4) In the case analyzed, compression with anterior and lateral displacement of the infratentorial structures (cerebellum and fourth ventricle) occurred. Compression of the fourth ventricle caused an obstruction in the CSF circulation and the resulting hydrocephalus. Compression, primarily of the cerebellar vermis, caused the postural and gait disturbances. These are the most common manifestations of a posterior fossa arachnoid cyst. (1,4,9)



The appearance on MRI is similar to that of CSF in both T1 and T2 sequences. (1) Some experts claim that pulsatile CSF movement within the cyst has been observed using MRI. The use of cine-MRI allows visualization of CSF flow between the ESA and the interior of the cyst. (1,8) This study provides more detailed information on the anatomical relationships and size of the cyst, as well as the condition of adjacent structures. (1,2,4,8) Although MRI is currently the diagnostic method of choice for arachnoid cysts, (1,2,4,8) non-contrast and contrast-enhanced CT of the head is an essential and sufficient tool for diagnosis. On CT, the cysts are almost always extra-axial, have cerebrospinal fluid density, and do not enhance with contrast. This study allows for the identification of associated bone deformities, compression of adjacent neurovascular structures, or the presence of hydrocephalus. (1,4) It is a rapid study that allows for immediate decision-making.

In the case presented, an MRI was not performed because the study was unavailable. A simple and contrast-enhanced CT scan of the head was performed. The differential diagnosis was made considering the patient's history and the imaging characteristics of the lesion. Both congenital cysts (megacisterna magna, epidermoid cyst, Dandy-Walker syndrome) and acquired cysts (astrocytoma, hydatid cyst, cysticercosis, porencephalic cyst) were considered. The presumptive diagnosis was ultimately confirmed by the histopathological study.

Conservative treatment with periodic imaging follow-up is intended for asymptomatic patients. Surgery aims to control or eliminate symptoms, although it may not reduce the size of the cyst. Several surgical techniques exist: puncture and evacuation, resection of the cyst walls, fenestration, and shunting. All surgical techniques can be performed via open surgery or with endoscopic support. Endoscopic treatment allows for minimal invasion of intracranial structures. It is considered by many to be the procedure of choice, as it has similar advantages to invasive techniques. (1-11)

The choice of surgical technique depends on the characteristics of the lesion and the



neurosurgeon's preference and experience. (1,4) Since minimally invasive techniques were unavailable, a direct approach was used with fenestration and resection of the posterolateral walls of the cyst. The anterior wall of the cyst, being attached to the cerebellar tissue, was preserved to avoid potential neurovascular damage. This technique allowed communication between the cyst and the physiological pathways of cerebrospinal fluid (CSF) circulation, minimizing the possibility of recurrence and achieving decompression of the surrounding structures.

Conclusions

Although most cysts remain stable throughout life, some produce complex clinical manifestations. These vary depending on the patient's age, location, and size of the cyst. CT and MRI are the gold standard studies for diagnosing these lesions. Surgical treatment is reserved for symptomatic cysts. While endoscopic techniques are currently preferred, open surgery is an effective and efficient option for the surgical treatment of arachnoid cysts.

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Conflict of interest

The authors declare no conflicts of interest.

Authorship contribution

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