

Lesión como malformación de Chiari secundaria a quiste aracnoideal de la cisterna cuadrigémina

Chiari malformation-like lesion secondary to arachnoid cyst of the quadrigeminal cistern

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Resumen

Se presenta el caso de una mujer de 31 años de edad con cefalea y cervicalgia que se agravaba con la maniobra de Valsalva, presentando además mareos, encontrándose una malformación de Chiari secundario a un quiste aracnoideo cuadrigeminal. Después de efectuada la resonancia magnética diagnóstica, la paciente fue sometida a descompresión del agujero magno y extirpación del quiste cuadrigeminal, seguido por la resolución tanto de la malformación de Chiari y el quiste. Los síntomas desaparecieron después de la cirugía y han permanecido completamente resuelto hasta la actualidad. En pacientes adultos que presentan signos y síntomas de una malformación de Chiari debido a la compresión de la médula por las las amígdalas cerebelosas, la presencia de un quiste aracnoideo de cisterna cuadrigeminal es una rara patología asociada que puede ser tratada quirúrgicamente.

Palabras clave: Quiste aracnoideo, Malformación de Chiari, Cisterna cuadrigeminal.

Abstract

We report a rare case of a 31-year-old woman with headache and pain manifested by cervicalgia that worsened with the Valsalva maneuver and dizziness, who was found to have a Chiari malformation secondary to a posterior fossa arachnoid cyst. After magnetic resonance imaging (MRI), the patient was submitted to foramen magnum decompression and arachnoid cyst removal that were followed by resolution of both the Chiari malformation and the cyst. The symptoms disappeared after surgery and have remained completely resolved to the present day. In adult patients who present with signs and symptoms of Chiari malformation due to direct medulla compression by the tonsils, a quadrigeminal cistern arachnoid cyst is a rare associated pathology that can be treated surgically.

Key words: Arachnoid cyst, Chiari malformation, Quadrigeminal cistern.

Introduction

Arachnoid cysts are commonly found in the Sylvian fissure, sella region, cerebral convexity and posterior fossa. Most patients are asymptomatic or present with seizure, focal deficit, normal pressure hydrocephalus, lower cranial nerve paresis or trigeminal neuralgia^{1,2}.

Arachnoid cysts are usually located in the posterior fossa. The quadrigeminal area between the incisural notch of the tentorium and the collicular plates is the third most frequent infratentorial location of arachnoid cysts. An arachnoid cyst in the quadrigeminal cistern can extend anteriorly and compress the quadrigeminal plate, causing hydrocephalus due to aqueductal stenosis^{2,3,4}.

Arachnoid cysts of the posterior fossa represent a rare lesion in the posterior fossa, the symptomatology of which depends on the extent and severity of compression on the brain stem, cerebellum and/or aqueduct².

We present a case of severe compression of the cerebellum causing herniation of the tonsils with symptoms very similar to those of a Chiari malformation.

Case Report

A 31-year-old woman was admitted to our clinic with headache and pain manifested by cervicalgia that worsened with the Valsalva maneuver and dizziness. During the examination, she was alert, conscious with a Glasgow coma score of 15 and had no motor deficits. Neurological examination of the eyes, ears, nose and throat did not reveal any cause of the headache. The fundoscopy examination was normal and there was no loss of any sensory modality in the fifth cranial nerve distribution. The remaining cranial nerves were all normal and there was no sign of nystagmus. The routine hematology examination was normal.

Magnetic resonance imaging (MRI) revealed a large quadrigeminal arachnoid cyst causing severe compression of the cerebellum. There was also compression of the entire brain stem and aqueduct of less intensity, with no dilation of the third or lateral ventricles. The intensity of the cerebrospinal fluid (CSF) of the cyst was the same in all sequences.

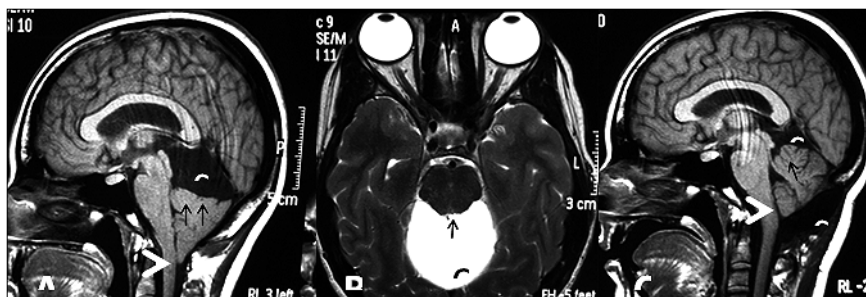


Figure 1. A-C: A) Preoperative sagittal T1-weighted MRI and B) axial T2W revealing the cyst (c) into quadrigeminal cistern, causing distortion of the tectum and flattening on colliculus (arrow in B); projecting downwards and determining severe compression of superior surface of the cerebellum (double arrows in A). See tonsillar herniation into foramen magnum (white arrowhead in A). C) Postoperative sagittal T1W MRI confirm a very important reduction of mass effect by cyst (c), and regression of tonsillar herniation (white arrow). Note decompression on the cerebellar folia (arrow). A residual fluid collection remains on surgical approach (c').

The imaging suggested a quadrigeminal arachnoid cyst as a cause of a bilateral herniation of the cerebellar tonsil (Figure 1).

Surgery was undertaken due to the frequency of the daily headaches and dizziness owing to the herniation of these tonsils. The cyst was totally removed by midline suboccipital craniotomy in the prone position. The cisterna magna was opened to reveal the bilateral cerebellar lobules and tonsils displaced by the cyst with a transparent, thin glossy membrane in the middle, emerging over the superior part of the vermis. Postoperatively, she was monitored in an intensive care unit for 2 days and discharged 5 days after the operation, with an improvement in her symptoms. Magnetic resonance imaging, performed six months later, revealed diminution of the cyst size and a small extradural collection at the site of the approach without any further complaints on the part of the patient (Figure 1). Histological examination confirmed the diagnosis of an arachnoid cyst of the quadrigeminal cistern.

Discussion

The prevalence and natural history of arachnoid cysts in adults have yet to be fully determined. With the increasing use of MRI and CT there has been a corresponding increase in the number of incidentally diagnosed arachnoid cysts. The studies of groups of adults with arachnoid cysts have estimated cyst prevalence to be between 0.3% and 1.7%. Another study with a population of children undergoing brain MRI

found that the prevalence of arachnoid cysts in children was 2.6%, slightly higher than in adults^{4,5}.

Arachnoid cysts are more frequent in males than in females. The most common locations are the middle fossa 34% [most commonly on the left side (70%)], retrocerebellar position (33%), and over the convexity (14%)⁵.

Arachnoid cysts of the posterior fossa represent a rare group of intracranial lesions. Quadrigeminal arachnoid cysts are also known as paramesencephalic or paracollicular arachnoid cyst, arachnoid cysts of the cisterna ambiens, tentorial notch arachnoid cysts, and parapineal arachnoid cysts^{2,6,7}. The retrocerebellar and cerebellopontine angle cysts are more common than quadrigeminal arachnoid cysts, which are the third most common infratentorial cysts². Quadrigeminal arachnoid cysts account for 5% to 10% of all intracranial arachnoid cysts⁷.

Infratentorial arachnoid cysts are mostly congenital, although they may be acquired pathologies. Inflammation, trauma and diverticulum from ventricles are also causative mechanisms^{1,8,9}. In our case, no inflammatory or traumatic causes were evident and histological examination found no hemorrhagic or inflammatory cells, so the cyst was considered to be of congenital origin.

The symptomatology depends on the severity and extent of compression on the brainstem, aqueduct and cerebellum². In infants, obstructive hydrocephalus due to obstruction of the aqueduct with intracranial hypertension is the most common clinical presentation^{2,6}. In adults we can find symptoms such as diplopia caused by lateral rectus pal-

sy, secondary to intracranial hypertension¹⁰ or trochlear nerve⁹, nistagmus, visual disturbances¹¹ hearing deficit, clonus⁹ spasticity¹⁰ hemiparesis, paraparesis¹⁰, signs of memory deficit, gait disturbances and sphincter incontinence linked to normal pressure hydrocephalus have been described⁸.

Our patient presented with dizziness, headache and cervicgia triggered by the Valsalva maneuver and coughing, suggesting a Chiari malformation. MRI confirmed a large cyst compressing the cerebellum, with downward displacement of the tonsils, a Chiari I-like syndrome, since a malformation is defined as a structural defect in the body due to abnormal embryonic or fetal development. Postoperatively, her symptoms of headache and dizziness improved

after decompression. MRI and CT imaging can provide the best information on the infratentorial, midline, supracerebellar location of the quadrigeminal cistern arachnoid cyst, with the same attenuation values as the CSF, and with no contrast enhancement^{4,5}.

Cyst excision is the most appropriate treatment with resection of the cyst wall. Ventriculoperitoneal shunting is sometimes required before craniotomy to remove the arachnoid membrane of the cyst. Internal cyst drainage (fenestration) or shunting the cyst to the peritoneal cavity are other options^{2,3,11}. Opening the posterior wall of the third ventricle to provide communication has also been recommended to avoid shunt dependency¹². The introduction of neuroendoscopy has provided a

minimally invasive modality for the surgical treatment of quadrigeminal arachnoid cysts⁷. Total cyst excision was achieved, without ventriculoperitoneal shunting since the patient did not develop hydrocephalus. The patient's symptoms disappeared after surgical removal of the cyst and she has remained asymptomatic to the present day.

Quadrigeminal arachnoid cysts resulting in a symptomatic acquired tonsillar herniation are rare. The management of these patients should be directed at decompressing the posterior fossa and includes the removal of the walls of the arachnoid cyst.

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