Case Report

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Spontaneous clival meningocele

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Abstract

The occipital bone is an uncommon location for meningoceles protrusion. This condition occurs generally after a severe traumatism or surgical procedure. However, in some rare cases, the herniation can happen spontaneously. Nontraumatic clival meningoceles present an extremely rare entity and correspond to a herniating pachymeningeal collection containing cerebrospinal fluid through a zone of fragility in the clivus. Clinical presentation ranges from simple headache or rhinorrhea to severe complications such as recurrent bacterial meningitis or nerve compression. Computed tomography provides an analysis of the bone and magnetic resonance imaging provides a superior contrast resolution, helping to distinguish among the various types of clival lesions. We report the case of a young woman with a long history of idiopathic intracranial hypertension, who presented with a worsening headache. Magnetic resonance imaging confirmed a clival meningocele without other complications and the patient was put under medical surveillance.

Keywords

Clivus, meningocele, computed tomography, magnetic resonance imaging

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Introduction

A meningocele protruding spontaneously through the clivus is not a very common finding.[1] The symptomatology is not always appealing, especially in the absence of a previous surgery or a traumatic context. Only few isolated cases of clival meningoceles without any impact on the cranial nerves have been described so far as patients generally present in the emergency room (ER) with nerve paralysis.[2]

The diagnosis is based on magnetic resonance imaging (MRI) imaging by visualization of the osseous defect, which is not always evident.

Many different therapeutical options can be proposed according to the degree of the functional impact. In some patients, the treatment can be symptomatic until resolution of symptoms or even medical abstention if the patient is asymptomatic.[3]

Case report

A 35-year-old female patient with a history of idiopathic intracranial hypertension presented with severe headache. Clinical examination did not show any neurological deficiency and ophthalmological assessment found bilateral papillary edema. A cerebral MRI 1.5 T was then performed.

T2 weighted sequences demonstrated signs of increased intracranial pressure, including an enlarged Meckel's cave (Figure 1(a) and (c)) and prominent subarachnoid space around the optic nerves (Figure 1(b)).

Dehiscence of the posterior wall of the clivus was present, communicating the prepontine cistern with the intraclival cavity, compatible with a clival meningocele (Figure 2).

The decision of clinical surveillance was proposed over an invasive surgery. Given her young age, our patient was treated with clinical vigilance, based on an adjustment of her medical treatment with an emphasis on healthy lifestyle and dietary measures.

Discussion

Cranial meningoceles are rare with an estimated prevalence of 0.05% with eight trans clival cerebrospinal fluid (CSF) collection and four clival meningocele reported cases so far.[4]

They can be either congenital or acquired, more frequently, subsequent to trauma (80%–90% of cases),[2] granulomas, tumors, systemic disease,[5] infections, and in 3%–4% of all cases, as a result of long-term remodeling, secondary to raised intracranial pressure.[4] The most likely explanation

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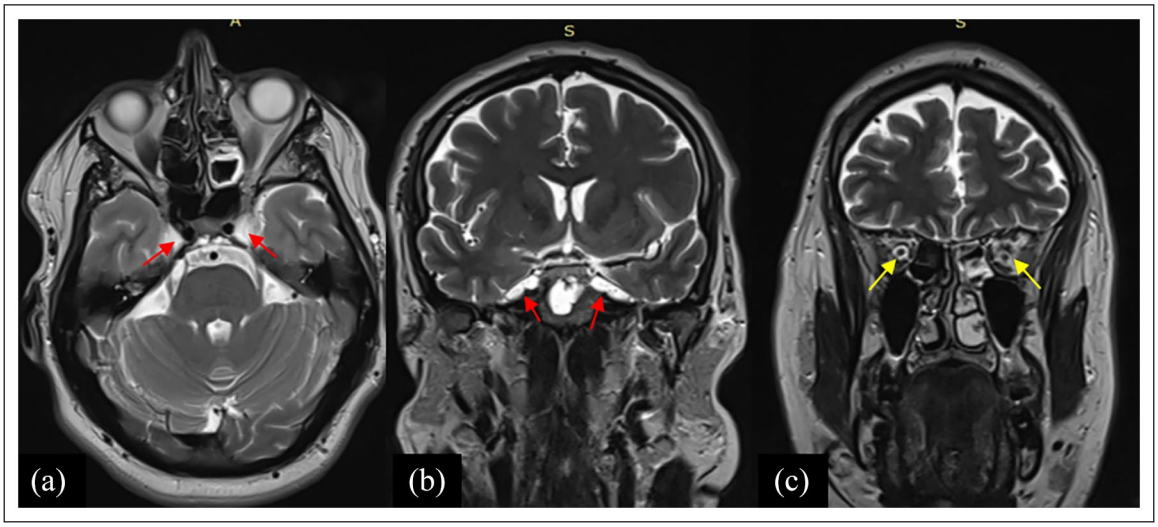

Figure 1. (a) T2 weighted axial and (b and c) coronal demonstrating a minimal bilateral dilation of perioptic subarachnoid spaces (yellow arrows) and an enlarged Meckel's cave (red arrows) confirming the intracranial hypertension.

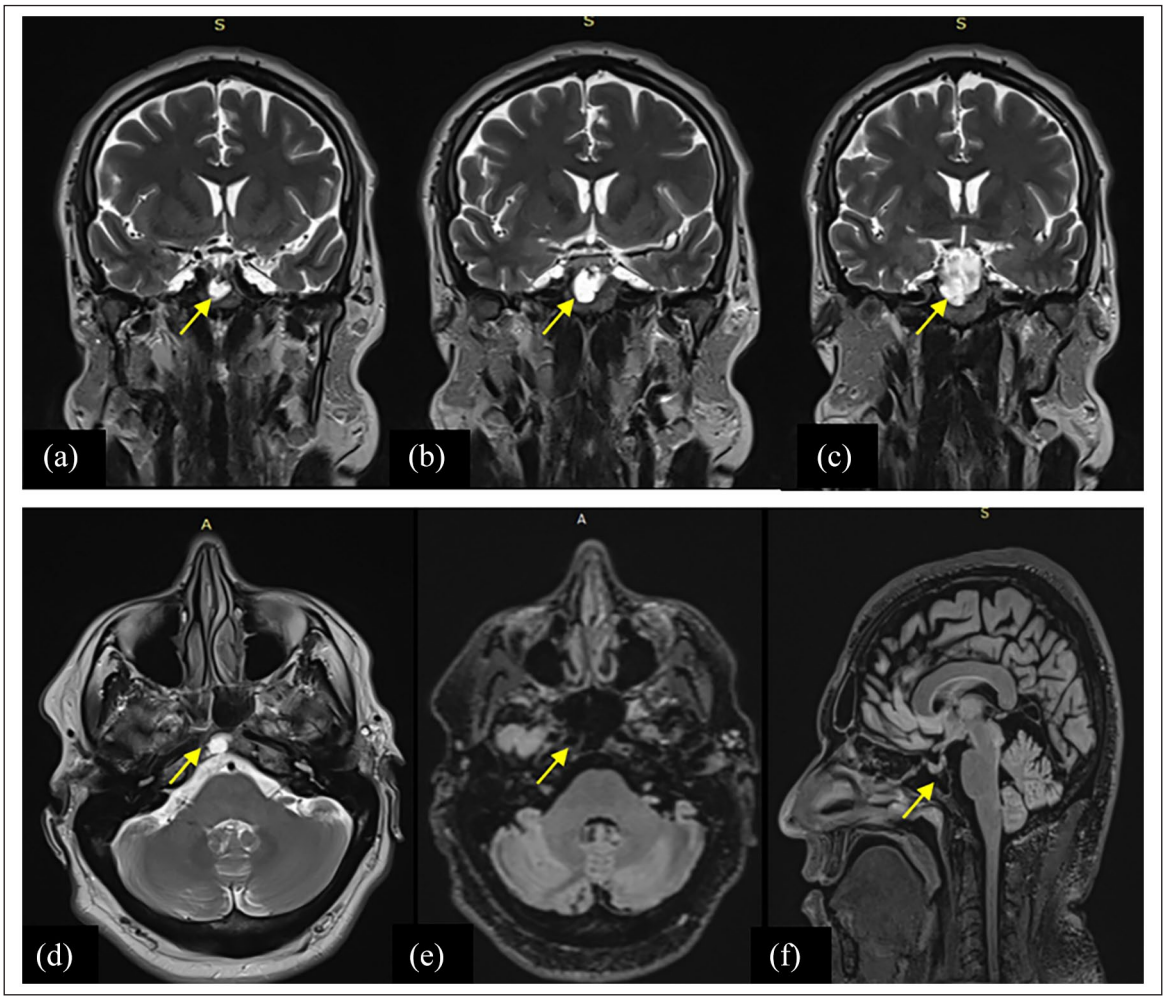

Figure 2. (a–c) Coronal and (d) axial T2. (e) Axial and (f) sagittal T2 FLAIR weighted images showing a dehiscence of the clival posterior wall with a communicating liquid isointense to CSF between the prepontine cistern and the clival cavity suggestive of a clival meningocele (arrow).

for spontaneous meningocele rests in the anatomic development. The basisphenoid and basiocciput bones join to form the clivus which we subdivide into the upper, middle, and lower thirds.[6] This spheno-occipital synchondrosis may not fuse in adult life and be responsible for weakening of the bone. Excessive aerial expansion of sphenoidal sinus causes further fragility to some points of the bone around this junction, leaving a thin wall.[7] Added to this anatomic vulnerability are functional factors like continuous or high CSF pressure that lead to a permanent defect of the skullbase with meningeal herniation and CSF leaks.

Clival meningoceles are space-occupying lesions that can have multiple complications, including recurrent meningitis, intracranial hypertension, and nerve compression due to local swelling.[4] CSF pressures along with the hydrostatic pulsatile forces may lead to the development of pit holes on the middle fossa at the sites of arachnoid villi with herniation of dura/arachnoid and even brain tissue. Arachnoid granulations eroding a pneumatized part of the skull can be significant causing CSF rhinorrhea.[8] Superficial siderosis of the central nervous system (CNS) is another rare reported complication of clival meningocele. It consists of hemosiderin deposition secondary to repetitive hemorrhaging into the subarachnoid space caused by this dural abnormality. The cardinal manifestation can be deafness and cerebellar ataxia.[9]

The diagnosis is based on cross-sectional imaging modalities. Computed tomography (CT) is very useful for the identification of a bony defect. Sometimes the osseous defect can be very small and may not appear on thin-slice CT, leading to misdiagnosis, as the lesion may seem to be confined to the bone. In this case, CT-cisternography demonstrates the communication of intrathecal contrast.[10]

MRI visualizes the meningeal collection herniating through the bone defect, particularly in T2-weighted sequences, and allows to rule out other possible diagnosis. Meningoceles will follow all MRI signal characteristics of CSF: T1 low signal and T2 high signal.

This cystic lesion carries some similarities with other probable benign pathologies such as cholesterol granuloma, which presents as a circumscribed mass replacing normal clival marrow: hyperintense on T1WI due to blood and cholesterol crystals, hyperintense centrally on T2WI, with a hypointense peripheral rim of hemosiderin, and non-enhancing on post contrast images.[7]

Another differential diagnosis to consider is ecchordosis physaliphora, a benign intradural hamartomatous lesion derived from residual notochord. It shows two components on T1WI: a hypointense intraclival component and an isointense intradural component. The lesion is hyperintense on T2WI and does not enhance after intravenous administration of gadolinium.[11]

We might cite as well chordomas, a locally aggressive, slow-growing malignant tumor arising from notochordal elements. They appear hypo- to isointense on T1WI and hyperintense on T2WI due to fluid signal intensity. They generally enhance heterogeneously with a "honeycombing" appearance.[11]

Besides, if the identified lesion presents as a homogeneous well-circumscribed lesion on MRI with a connection between the lesion and subarachnoid space, consideration must also be given to the possibility of herniated brain parenchyma, demonstrating an enhancement similar to a normal brain parenchyma, which would distinguish it as a meningoencephalocele.

Surgical correction of basal skull defects in patients with meningocele is recommended, for the prevention of both meningitis and further progression of the herniation.[12]

In cases of clival defect, the endonasal endoscopic approach gives easier access to the area of interest, and is therefore recommended over the transcranial approach. The principle of this surgery is based on packing of the sphenoid sinus with sufficient multilayer closure such as fascia lata graft, fat tissue bio-glue,[8] or a vascularized nasoseptal flap[9] that would fill in all the surface defects. This direct technique provides satisfactory results in patients with symptomatic transclival meningoceles.[8,13,14]

However, only few cases describe vigilant surveillance as a therapeutical option, especially in patients with small bone defect with minimal herniation,[15] requiring close observation until an invasive procedure is needed. This spared the reported patients an invasive procedure and meningoceles resolved spontaneously.[3,15–17]

Conclusion

Spontaneous clival meningoceles have been rarely reported in the literature. MRI is the key to diagnosis, showing the zone of defect and the communication with subarachnoid spaces. This benign condition can cause functional impairment in some cases, and expose to a high risk of CNS infections, in which case surgical intervention should be recommended.

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