Ependymoma mimicking cavernous malformation of the third ventricle: case report.

Ependimoma simulando cavernoma do terceiro ventrículo: relato de caso.

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INTRODUCTION

Ependymomas were firstly described by Virchow in 1863¹ and firstly identified as a separate histological entity by Bailey and Cushing in 1926⁶. Ependymomas are neuroepithelial tumors arising from the ependyma and underlying glia of cerebral ventricles, the spinal cord central canal, or cells of the terminal ventricle⁴,⁵. These tumors constitute approximately 6 - 12% of all intracranial tumors in children (commonly infratentorial)⁷ and are even less prevalent in adults, where they constitute approximately 2% of intracranial tumors (commonly supratentorial)⁸,⁹. They are more common in children younger than 3 years of age (30% of all intracranial tumors).

The majority of ependymomas of the central nervous system (CNS) are found in the intracranial compartments. More than 70% of these tumors occur infratentorially, usually in the fourth ventricle in the midline. Supratentorial ependymomas originating from the wall of the ventricles and from brain parenchyma arise with almost equal frequency. Ependymomas of the third ventricle are very rare⁴.

HISTORY AND EXAMINATION.

A 35-year-old woman was admitted to the Neurosurgery Unit with a 6 months history of intermittent progressive headache. She had no other complaint or significant medical history. The headache was not associated with nausea, vomiting, visual alterations or deteriorating mental state. Neurological examination was normal.
NEUROIMAGING
Computed tomography (CT) scan showed noncommunicating hydrocephalus, secondary to a mass lesion in the third ventricle. Magnetic resonance imaging (MRI) demonstrated a 2.9 X 2.8 X 1.9 cm third ventricle lesion isointense with the gray matter on the T1-weighted sequences, with nonhomogeneous contrast-enhancement after gadolinium, and T2-weighted sequences with a hypointense perilesional ring caused by hemossiderin deposition. A developmental venous anomaly (DVA) at pontomesencephalic sulcus was associated with the ventricular lesion (Fig. 1). The clinical presentation and neuroimaging appearance led to an initial diagnosis of cavernous malformation (CM).

OPERATION
The patient underwent ventriculoperitoneal shunt and the postoperative course was uneventful. The patient had a complete resolution of her headache. The postoperative shunt MRI confirmed resolution of hydrocephalus and confirmed an initial diagnosis of CM. The patient was submitted to the preoperative evaluation for the elective neurosurgical procedure. A transcallosal transchoroidal approach was performed. Complete microsurgical dissection of the lesion by meticulous decompression and identification of the “plane” between the tumor and ependyma was done. The operative findings did not suggest a CM. The lesion was sent for histopatological examination.

PATHOLOGICAL FINDINGS
Tissue was fixed in 10% buffered formalin and processed. Histopathological and immunohistochemistry (Table 1) examination featured a diagnosis of ependymoma (Grade II). Hematoxylin and eosin demonstrated neoplastic neuroepithelial cells with ependymal rosettes and perivascular pseudorosettes. Postoperative Course. The patient developed a transitory Parinaud’s syndrome secondary to the dissection of the posterior third ventricle. She experienced no other complication and was discharged home on hospital Day 7.

Table I – Antisera used for immunohistochemical study

<table>
<thead>
<tr>
<th>Antisera</th>
<th>Source</th>
<th>Result</th>
</tr>
</thead>
<tbody>
<tr>
<td>EMA</td>
<td>NOVOCASTRA: CLONE GP1.4</td>
<td>Negative</td>
</tr>
<tr>
<td>S-100 protein</td>
<td>DAKO: Polyclonal</td>
<td>Diffusely positive</td>
</tr>
<tr>
<td>GFAP</td>
<td>DAKO: CLONE 6F2</td>
<td>Diffusely positive</td>
</tr>
<tr>
<td>KI 67</td>
<td>DAKO: CLONE MIB-1</td>
<td>Inconclusive</td>
</tr>
</tbody>
</table>

EMA: epithelial membrane antigen; GFAP: glial fibrillary acidic protein

DISCUSSION
The third ventricle is an unusual place for the development of ependymomas and this frequency is disproportionately less than the relative ependymal surface area, when compared with the fourth ventricle. Ependymomas are much more common in the fourth ventricle despite the ependymal surface area of third ventricle being greater than that of the fourth ventricle. It is believed that ependymomas arise from ependymal cell rests that tend to occur where the ventricles are sharply angled. These areas in CNS include the anterior spur of aqueduct, adjacent to the trigone of lateral ventricle, the area near the foramen of Luschka, and the terminal filum. Perhaps there may be a paucity of such cell rests in the third ventricle. Similar, the parenchymal ependymomas arise from rests of ependymal cells remaining within the brain parenchyma during embryological development.
Treatment of intracranial ependymomas may include surgery, radiation therapy, and chemotherapy. Surgery is the mainstay of the therapy and total resection is recommended when it is possible. Preoperative placement of a shunt is often required for intraventricular lesions. Third ventricular ependymomas in adults patients are considered a poor prognostic factor with 10-year survival rate of 35% (60% for lateral ventricles).

Histopathological analysis is the “gold standard” for definitively characterizing an intracranial lesion suspected of being a brain tumor. Neuroimaging classification of brain tumors is frequently reported to be unreliable, especially for gliomas. M. Julià-Sapé, et al., reported the accuracy of routine MRI studies in the classification of brain tumors and they provided detailed performance measures for MRI such as sensitivity and specificity. These authors found sensitivity 66.7% (95% CI 20.8-93.9), specificity 99.5% (95% CI 98.1-99.9), positive predictive value 50% (95% CI 15.0-85.0) and negative predictive value 99.7% (95% CI 98.6-100.0) to the diagnosis of ependymomas.2

CM may be associated with other vascular malformations, such as DVA, angiomas and capillary telangiectasias. Because of recent advances in brain imaging, they are now being diagnosed with increased frequency. MRI is the diagnostic method of choice of CM and is an extremely sensitive imaging method for detection of DVA. The coexistence of a CM and a DVA is the most common mixed vascular malformation. Much discrepancy exists in the literature concerning the prevalence of the coexistence of MCs and DVAs. Several authors report an association about 25% of the cases.10

Misdiagnosis of third ventricle lesions is not that rare. Some authors have pointed out that third ventricle MCs may have a unique appearance on MRI when compared with MCs locate elsewhere. The imaging characteristics of this patient showing a “popcorn” pattern in the third ventricular lesion and the coexistence of a DVA at the ponto-mesencephalic sulcus led us to an initial diagnosis of CM. Histopathological and immunohistochemistry examination featured a diagnosis of ependymoma (Grade II).

Despite a very persuasive radiological evidence of CM, we must emphasise that histopathological analysis is crucial in establishing the correct diagnosis. We report a third ventricular ependymoma mimicking CM with histopathological diagnosis after microsurgical lesion resection. Ependymomas should not be omitted from the differential diagnosis of tumors of third ventricle. To the best of our knowledge, this is the first reported case of ependymoma in the third ventricle mimicking a CM and coexisting with DVA.

References


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