Caso clínico

Epidermoid cyst mimicking an intrinsic brainstem tumor

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Abstract

Objective: To describe an atypical clinical and radiological presentation of a brainstem epidermoid cyst in a child and to provide a review of the medical literature on brainstem epidermoid cysts in children.

Material and method: Review of medical records and operative notes of an unusual case of a patient with a brainstem epidermoid cyst. MEDLINE literature search using the terms brainstem, epidermoid cyst and children.

Results: Gross total resection of the cyst was achieved. The patient had an uneventful recovery.

Conclusion: Epidermoid cysts are rare tumors of the brain and children. The management of these tumors can be quite challenging. A good clinical and neuroradiological evaluation preoperatively is fundamental for a successful surgical treatment. Surgical resection should be as radical as possible without putting the patient's neurological status into risk.

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Quiste epidermoide simulando un tumor intrínseco del tronco cerebral

Resumen

Objetivo: Describir una presentación atípica clínica y radiológica de un quiste epidermoide del tronco cerebral en un niño y hacer una revisión de la literatura médica sobre los quistes epidermoides del tronco cerebral en los niños.

Material y método: Revisión de historias clínicas y notas operativas de un caso inusual de un paciente con un quiste epidermoide del tronco cerebral. Búsqueda bibliográfica de los términos tronco encéfalo, quiste epidermoide y niños.

Resultados: La resección quirúrgica completa del quiste se logró. El paciente tuvo una recuperación sin complicaciones.

Conclusión: Los quistes epidermoides son tumores poco frecuentes en el cerebro y su manejo implica algunos riesgos. Una buena evaluación clínica y radiológica antes de la operación es...
Introduction

Epidermoid cysts are uncommon benign tumors that represent 1% of all Central Nervous System tumors. They were first described by the French pathologist Cruveilhier in 1829 and are quite rare in the pediatric age group. These tumors arise from inclusion of epidermal elements at the time of neural groove closure, which occurs between the 3rd and 5th weeks of embryonic life. The occurrence of these tumors in the brainstem is also very unusual. To our knowledge, only 13 pediatric cases of true brainstem epidermoid tumors have been reported in the literature. Herein, we describe an atypical clinical and radiological presentation of a brainstem epidermoid cyst in a child.

Case report

A five-year-old boy was admitted with a 3-month history of progressive left hemiparesis, headache, nausea, drowsiness and strabismus. Since the initial Computerized Tomography (CT) scan showed severe triventricular obstructive hydrocephalus, an endoscopic third ventriculostomy (ETV) was performed. After surgery, the patient was alert and awake, but continued to have right VI nerve palsy, left hemiparesis and slight gait ataxia. He showed no clinical or laboratory signs of infection. Brain Magnetic Resonance Imaging (MRI) revealed an atypical lesion in the pons involving the basilar artery and filling up the preptontine and cerebellopontine angle cisterns (Fig. 1). This atypical presentation led us to perform an open biopsy.

The patient underwent a right lateral suboccipital craniotomy. After exposure, the lesion was found displacing the V, VII, VIII and IX cranial nerves posterolaterally. Shortly following cyst fenestration, a thick, yellow-greenish, pus-like fluid drained from the lesion cavity. Typical epidermoid cyst content, with a pearly white wall, was seen and removed. The use of microsurgical techniques made it possible to gradually separate the capsule of the cyst from the surrounding structures and resect it completely. A postoperative MRI scan which included diffusion-weighted sequences showed total removal of the lesion. Histopathological examination revealed an epidermoid cyst. The postoperative period was uneventful and the patient recovered well, except for a remaining mild hemiparesis. The one-year follow-up MRI revealed no recurrence of the lesion.

Discussion

Epidermoid cystic tumors are extremely rare in children: only 13 pediatric cases have been described in the medical literature. The management of these tumors has involved some controversies over diagnostic work-up and therapeutic
Table 1 - Summary of cases of epidermoid cysts in children.

<table>
<thead>
<tr>
<th>Case number</th>
<th>Authors</th>
<th>Year</th>
<th>Patient age (years)</th>
<th>Location</th>
<th>Surgical resection</th>
<th>Outcome/recurrence</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Bhatia et al.⁵</td>
<td>1978</td>
<td>3.5</td>
<td>Pontomedullary</td>
<td>Subtotal</td>
<td>Died</td>
</tr>
<tr>
<td>2</td>
<td>Leal and Miles⁶</td>
<td>1978</td>
<td>3.5</td>
<td>Medullary</td>
<td>Subtotal</td>
<td>Died</td>
</tr>
<tr>
<td>3</td>
<td>Schwartz and Balentine⁷</td>
<td>1978</td>
<td>14</td>
<td>Pontine</td>
<td>Subtotal</td>
<td>Died</td>
</tr>
<tr>
<td>4</td>
<td>Weaver and Coulon⁸</td>
<td>1979</td>
<td>1</td>
<td>Pontine</td>
<td>Subtotal</td>
<td>Died</td>
</tr>
<tr>
<td>5</td>
<td>Fournier et al.⁹</td>
<td>1992</td>
<td>14</td>
<td>Pontomedullary</td>
<td>Subtotal</td>
<td>Died</td>
</tr>
<tr>
<td>6</td>
<td>Radhakrishnan et al.¹⁰</td>
<td>1992</td>
<td>13</td>
<td>Pontomedullary</td>
<td>Not attempted</td>
<td>Post-mortem diagnosis</td>
</tr>
<tr>
<td>7</td>
<td>Kuzeyli et al.¹¹</td>
<td>1996</td>
<td>2</td>
<td>Pontine</td>
<td>Subtotal</td>
<td>Not mentioned</td>
</tr>
<tr>
<td>8</td>
<td>Caldarelli et al.³</td>
<td>2001</td>
<td>16</td>
<td>Pontine</td>
<td>Subtotal</td>
<td>Residual intrapontine lesion and in the petrous apex; asymptomatic 3 years afterwards</td>
</tr>
<tr>
<td>9</td>
<td>Caldarelli et al.³</td>
<td>2001</td>
<td>1.5</td>
<td>Pontomedullary</td>
<td>Gross total</td>
<td>Tumor regrowth after 18 months (resected); child asymptomatic after 3 years</td>
</tr>
<tr>
<td>10</td>
<td>Ziyal et al.²</td>
<td>2004</td>
<td>5</td>
<td>Medullary</td>
<td>Gross total</td>
<td>Not mentioned VI nerve palsy, partial VII nerve palsy and no recurrence after 16 months</td>
</tr>
<tr>
<td>11</td>
<td>Takahashi et al.⁴</td>
<td>2007</td>
<td>10</td>
<td>Pontomedullary</td>
<td>Gross total</td>
<td>Asymptomatic with no tumor growth over 11 months</td>
</tr>
<tr>
<td>12</td>
<td>Golomb and Whitehead¹³</td>
<td>2006</td>
<td>10</td>
<td>Pontine</td>
<td>Not operated</td>
<td></td>
</tr>
</tbody>
</table>

considerations. The usual clinical manifestations of these tumors are cranial nerve palsies, which are sometimes associated with signs of raised intracranial pressure, due to obstructive hydrocephalus. Although not common, aseptic meningitis resulting from the spillage of the highly irritative contents of the cyst may also occur.³

Typically, epidermoids appear hypointense on T1-weighted MRI scans, but might also have an intermediate intensity between brain and cerebrospinal fluid (CSF) on T1, whereas they are usually hyperintense on T2-weighted sequences.²,³ However, signal intensity can be variable depending on the relative amount of lipid, cholesterol, keratin and proteins. Furthermore, the cysts normally have a regular contour and seldom show contrast enhancement or peritumoral edema.² A clear distinction between the tumor and the adjacent parenchyma is also frequently observed.

In our case, the radiological presentation was unusual. The patient's MRI did not show the usual regular contour of epidermoid cysts, leading us to perform an open biopsy, despite the initial diagnosis was of an infiltrative tumor of the brainstem, which is, in general, not managed surgically, provided that, in such cases, no therapeutic benefit is gained by sampling or resection¹²; however, we opted for an open biopsy because of the atypical radiological aspect of the lesion.

This finding stresses the importance of performing neuroradiological investigation to determine the characteristics of the tumor and its relationship to the brainstem. MRI is distinctly superior to CT.⁶,¹² However, neuroradiological findings may be atypical in cases of intra-axial lesions and should be distinguished from other diagnostic possibilities, such as arachnoid cysts, dermoid tumors, lipomas, teratomas, cholesterol granulomas, and, particularly in this case, from diffusely infiltrative gliomas of the brainstem.³ Out of the 13 cases previously mentioned, only 9 were studied by using MRI.²,⁴,⁸,¹¹

The use of modern MRI techniques, such as spectroscopy and Diffusion Weighted Imaging (DWI), might be helpful in establishing the diagnosis.³ For instance, DWI may differentiate epidermoid cysts from other cystic tumors, since the former would appear very hyperintense on the images, while the latter are usually hypointense or mildly hyperintense (bright). On the other hand, DWI is unable to distinguish epidermoids from less-hyperintense abscesses.

Table 1 summarizes published data on epidermoid cysts in pediatric patients. According to the table, epidermoids occur more often in girls and are located predominantly in the pons, extending to the medulla oblongata. High mortality rates have been observed: 6 out of 13 patients died, mainly due to dense adhesions of the tumor to important neurovascular structures. Owing to tumor location, only four cases were treated with gross total resection.

There appears to be a consensus about surgical resection as the best therapeutic option for treating epidermoid cysts; the optimal extent of resection remains controversial. Some authors⁷,⁹,¹⁰,¹² have advocated subtotal resection, arguing that total resection implies unnecessary high risk of neurological deficits and mortality, since the residual lesion often stays stable or grows slowly. Nevertheless, Ziyal et al.² pointed out that surgical resection should be as radical as possible, and reported a recurrence rate as high as 54%. In our case, total resection was possible because the cyst membranes were loosely attached to the antero-lateral aspect of the brainstem, cranial nerves and vascular structures.

Conclusions

Epidermoid cysts are uncommon brain tumors which rarely occur in children. Since their management involves some pitfalls, a good clinical and neuroradiological preoperative evaluation is fundamental for a successful surgical treatment. Surgical resection should be as radical as possible, but without putting the patient's neurological status at risk.
REFERENCES