Pineal region chordoid meningioma

Meningioma cordóide da região pineal

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ABSTRACT

Background: Meningiomas correspond to only 8% of pineal region tumors. Chordoid meningioma (CM) comprises only 0.5 to 1.0% of all intracranial meningiomas and is even rarer in the pineal region. In the literature, we found only six cases of pineal region CMs reported. Clinical presentation: We describe a case of one patient with headache, confusion, slurred speech and gait disturbance. The ophthalmologic examination revealed vertical gaze paresis and near-light dissociation of papillary reflex. MRI disclosed a large pineal region tumor with heterogeneous contrast enhancement. The tumor was totally removed by an occipital transtentorial approach (OTA) in a “three-quarter prone” position. Histological analysis disclosed a CM. Conclusion: Chordoid meningioma is a very rare variant of meningioma and the pineal region is a poorly described site. This study contributes to the understanding of this heterogeneous entity, to consider the CM as a differential diagnosis of pineal region tumors and to provide proper management of affected patients.

Keywords: Chordoid; Meningioma; Pineal region.

RESUMO

Objetivo: Meningiomas correspondem a apenas 8% dos tumores da região pineal. O meningioma cordóide (CM) representa 0,5 a 1,0% de todos os meningiomas intracranianos, sendo ainda mais raro na região pineal. Na literatura, apenas seis casos de CMs são relatados. Apresentação clínica: Descrevemos o caso de um paciente com cefaleia, confusão mental, disartria e ataxia de marcha. O exame oftalmológico revelou paresia do olhar vertical e dissociação do reflexo pupilar photomotor e de convergência. A RM revelou uma lesão expansiva na região da pineal com impregnação heterogênea pelo contraste. O tumor foi totalmente ressecado pela abordagem occipital transtentorial lateral em posição três-quartos prona. A análise histológica revelou um CM. Conclusão: Meningioma cordóide é uma variante rara de meningioma pouco descrita na região pineal, mas que deve ser considerada como diagnóstico diferencial das patologias localizadas nessa região. Este estudo contribui para um melhor entendimento do diagnostico e manejo destes tumores.

Palavras Chave: Cordóide; Meningioma; Região Pineal

INTRODUCTION

Chordoid meningioma (CM) is a very rare variant of meningioma with a unique chordoid appearance. It is classified as a WHO Grade II meningioma subtype and has a higher tendency to recur than common meningiomas1,5,17. Eventually, it can be part of Castleman’s disease, especially in young patients3. Usually, CM occurs in the supratentorial region and the location in the pineal region is extremely atypical and poorly documented in the literature6,9,10,13. Hereby, we report a case of pineal region CM in a 44-year-old male. Also, a literature review about the lesion and its treatment was undertaken.

CASE PRESENTATION

A 44-year-old male patient was referred to our neurosurgical unit from another institution with a 3-month history of progressive headache, confusion and drowsiness. He had undergone two stereotactic brain biopsy procedures of a pineal region lesion with inconclusive histological diagnosis and a ventriculoperitoneal shunt to treat hydrocephalus.
On neurologic examination he presented confusion, difficulty to walk and slurred speech. The reflexes were hyperactive with a bilateral ankle clonus. Ocular abnormalities included vertical gaze paresis and near-light dissociation of pupillary reflex (Parinaud’s syndrome). Laboratory analyses were negative for hematological abnormalities, including dysgammaglobulinemia. A brain magnetic resonance imaging (MRI-scan) showed an enlarging pineal mass when compared to the previous studies (Fig. 1). The serum and cerebrospinal fluid tumor markers including, alpha-fetoprotein, beta human chorionic gonadotropin were normal. Based on these radiological findings, negative pineal tumors markers and inconclusive histological diagnosis, surgical resection was indicated.

The patient was positioned in a “three-quarter prone” position and submitted to a right occipital transtentorial approach (OTA) as previously described1 After gentle right occipital lobe retraction, the tentorium was divided lateral to the straight sinus. The arachnoid overlying the quadrigeminal plate and vein of Galen was sharply opened. The soft pinkish tumor enlarging the space between the precentral cerebellar plate and vein of Galen was sharply opened. The soft pinkish tumor enlarging the space between the precentral cerebellar vein and the internal cerebral vein was observed (Fig. 2). After debulking of the core of the tumor, it was separated from the deep venous system. This tumor presented a well-defined cleavage plan to the anterior third ventricle walls, tectorial plate, splenium of the corpus callosum and, thalamus. Endoscopic inspection of surgical cavity at the end of procedure showed a total resection.

The patient had an uneventful postoperative course and was discharged on postoperative day four. Pathologic analysis showed a chordoid meningioma (Fig. 3). Immunohistochemical study confirmed the histology. The tumor Ki67/MIB-1 labeling index was 1%.
On two year follow-up, the patient’s symptoms and neurological deficits resolved completely, except for a left arm tremor. The MRI-scan showed no evidence of tumor recurrence (Fig. 4). Two years after surgery, patient has no signs of recurrence.

**DISCUSSION**

Meningiomas in general correspond to approximately 8% of the pineal region lesions. A wide variety of tumors can affect pineal region, the most common being germ cell tumors, gliomas, and pineal cell tumors. Meningiomas seem to arise from arachnoid cap cells of the falcotentorial junction or from the velum interpositum in the roof of the third ventricle.

Chordoid meningioma (CM) is an atypical tumor described by Connors et al., in 1980 and initially associated with retarded somatic and sexual development, hepatosplenomegaly, anemia, and dysgammaglobulinemia (Castleman’s disease). The term “chordoid meningioma” was first used by Kepes et al., in 1988 to describe the distinctive histological chordoid appearance of the tumor.

Hence, several cases of CM have been described with the establishment of the classification of this tumor as a WHO grade II meningioma in 1993, corresponding to approximately 0.5-1.0% of all intracranial meningiomas. In the 2007 WHO review classification, the CM joined the clear cell and the atypical meningioma in the grade II category.

To our knowledge, only six cases the pineal region CMs were previously reported (Table 1). Small series and case reports of CMs located on other intracranial sites have been described showing higher incidence in younger age group with a slight female predominance.

Table 1: Reported cases of pineal region chordoid meningiomas

<table>
<thead>
<tr>
<th>Author &amp; Year</th>
<th>Age (yrs), Sex</th>
<th>Symptoms &amp; Signs</th>
<th>MRI-Scan</th>
<th>Approach</th>
<th>Resection</th>
<th>MIB-1</th>
<th>Follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kobata et al.13, 1998</td>
<td>15, F</td>
<td>Visual deficit, seizures, anemia</td>
<td>T1W – iso/hypo T2W – hyper Gd T1W – homog</td>
<td>OTA</td>
<td>Total</td>
<td>NI</td>
<td>2 yrs No recurrence</td>
</tr>
<tr>
<td>Haque et al.5, 2002</td>
<td>34, M</td>
<td>ICH, hearing loss, gait ataxia, Parinaud’s sign</td>
<td>T1W – NI T2W – NI Gd T1W – homog</td>
<td>OTA</td>
<td>Total</td>
<td>8.4%</td>
<td>2 wks No recurrence</td>
</tr>
<tr>
<td>Ida et al.7, 2007</td>
<td>44, M</td>
<td>H/A, visual deficit, gait ataxia</td>
<td>T1W – hypo T2W – hyper Gd T1W – heterog</td>
<td>NI</td>
<td>Subtotal</td>
<td>20.1%</td>
<td>8 m Recurrence</td>
</tr>
<tr>
<td>Fukushima et al.6, 2008</td>
<td>37. F</td>
<td>ICH, diplopia, Parinaud’s sign</td>
<td>T1W – NI T2W – hypo Gd T1W – heterog</td>
<td>NI</td>
<td>Subtotal</td>
<td>2.1%</td>
<td>6m NA</td>
</tr>
<tr>
<td>Present case</td>
<td>22, F</td>
<td>H/A, fever, anemia</td>
<td>T1W – hypo T2W – hypo Gd T1W – heterog</td>
<td>OTA</td>
<td>Total</td>
<td>1%</td>
<td>2 yrs NA</td>
</tr>
<tr>
<td></td>
<td>44, M</td>
<td>H/A, confusion, gait ataxia, slurred speech, Parinaud’s sign</td>
<td>T1W – hypso/iso T2W – hyper Gd T1W – heterog</td>
<td>OTA</td>
<td>Total</td>
<td>1%</td>
<td>1 y No recurrence</td>
</tr>
</tbody>
</table>

ICH (Intracranial hypertension); H/A (Headache); Iso (Isointense); Hyper (Hyperintense); Hetero (Heterogeneous); Homo (Homogeneous); OTA (Occipital transtentorial approach); NI (Not informed); NA (Not applicable).

The clinical signals and symptoms of pineal region CMs are thought to be a result of mass effect associated with a peritumoral lymphoplasmacellular infiltrate and include obstructive hydrocephalus (headache, vomiting, papilledema,
and gait disturbances), altered mental changes and ocular abnormalities.

Diagnosis of a pineal tumor is made based on MRI. This technique allows the understanding of anatomic position and, therefore, the relationship of the tumor with the surrounding neural structures and major intracranial vessels to assess the proper approach. On MRI-scan the CM is hypointense on T1-weighted images and is isointense to hyperintense on T2-weighted images with variant contrast enhancement. The pineal meningiomas usually displace the deep venous system ventrally and inferiorly, thereby providing a clue to the diagnosis.

Nowadays, it is difficult to differentiate pineal region meningiomas from others pineal tumors only by through image appearance. Though, it is imperative to establish a tissue diagnosis and evaluate blood and/or tumor markers to provide proper management.

Although stereotactic biopsy can achieve correct diagnostic in about of 94% in pineal lesions, it was negative in our case after two attempts.

The majority of the patients presenting with tumor obstructive acute hydrocephalus secondary to pineal tumors are managed in our department with endoscopic third ventriculostomy and tumor biopsy. Stereotactic biopsy is reserved for patient with pineal region masses without hydrocephalus.

Surgery is the standard treatment for meningiomas located in all intracranial sites and postoperative radiotherapy is reserved for cases of subtotal resection or malignant subtypes.

Current selection of surgical approach to pineal tumors depends primarily on the relationship of the tumor to the deep venous system and other surrounding structures. The OTA is selected when the tumor displaces the internal cerebral veins ventrally and inferiorly, and did not expand dorsally to the splenium of the corpus callosum. Also the angle of the tentorium is taken under consideration.

Safe and effective surgical strategies are essential for current clinical management of pineal tumors and correct histological diagnosis is fundamental to decision making.

Histologically, CMs show eosinophilic, vacuolated, spindle-shaped and polygonal cells that formed trabeculae and cords in a myxoid background. A peripheral lymphoplasmacellular infiltrate have been described in some cases.

In the event of overwhelming chordoid presence, the immunohistochemical study becomes of ultimate importance. Immunohistochemical pattern of positivity for vimentin and epithelial membrane antigen (EMA), in conjunction with negativity for glial fibrillary acidic protein (GFAP), S-100 protein and cytokeratin helps in differentiating other morphologically simulating tumors such as chordoma, chordoid glioma, epithelial hemangioblastoma and metastatic mucinous carcinoma.

Despite of the benign origin of meningiomas, the CMs reveal a more aggressive behavior. A higher proliferation index (MIB-1), chordoid pattern predominance and longer follow-up period increase tumor recurrence rates. The chance of recurrence ranges from 0 to 42% according to the series reported in literature. The mean time of recurrence can be as long as 10.4 years. Radiation therapy is usually reserved for subtotal resection or tumor recurrence. In our case, because the completeness of resection, low proliferation index a close follow-up was preferred and patient did not present signs of recurrence in MRI scan two years after surgical procedure.

CONCLUSION

CM is a very rare variant of meningioma and the pineal region is a poorly described site. This study contributes to the understanding of this heterogeneous entity, to consider the CM as a differential diagnosis of pineal region tumors and to provide proper management of affected patients.

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