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Case Report

Successful endoscopic endonasal management of a transclival cerebrospinal fluid fistula secondary to *ecchordosis physaliphora* – An ectopic remnant of primitive notochord tissue in the clivus

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1. Introduction

Ecchordosis physaliphora (EP) is a rare congenital jelly-like lesion, small in size, found more often on the dorsal midline surface of the clivus. Thought to be derived from ectopic remnants of primitive notochord tissue, it is usually asymptomatic and found only incidentally in post-mortem or radiological studies [1]. Few symptomatic presentations, however, have already been described in the literature [2].

We present an unusual case of EP associated to transclival CSF fistula which was successfully managed by an endoscopic endonasal skull base surgery technique. We subsequently present a brief discussion about EP and its main differential diagnosis.

2. Case report

A 54-year-old female patient presented to the emergency unity of a tertiary hospital with an acute history of high fever, severe headache, neck stiffness, vomiting and confusion. Lumbar puncture was promptly performed and CSF analysis confirmed the hypothesis of acute bacterial meningitis. Proper treatment with antibiotic and corticosteroid regimens was started.

After recovering normal mental status, she referred a 6-month history of intermittent, right-sided, watery rhinorrhea. She denied co-morbidities and use of medications. Her physical, neurological and otolaryngological examinations were otherwise unremarkable at that moment.

She was then submitted to a complete radiological investigation. Computed tomography-cisternography (CT-C) images demonstrated a cystic lesion inside the sphenoid sinus. The lesion communicated with the subarachnoid space trough a subtle osseous defect in the superior part of the clivus (Fig. 1A). Further computed tomography (CT) images evidenced that the tiny hole had regular margins and there was no bone erosion (Fig. 1B). On magnetic resonance (MR) scans the intrasphenoidal lesion was hypointense and hyperintense on T1 and T2-weighted images, respectively (Fig. 1C and D). The lesion did not show enhancement after contrast administration on CT and MR images.

Based on these benign clinical and radiological characteristics, a diagnostic hypothesis of transclival CSF fistula secondary to *ecchordosis physaliphora* (EP) was made.

An endoscopic endonasal surgery was performed. During the procedure, after intrathecal fluorescein injection, a fluorescein-colored cystic lesion was clearly visualized on the posterior wall of the sphenoid sinus. The lesion was excised and two small bone

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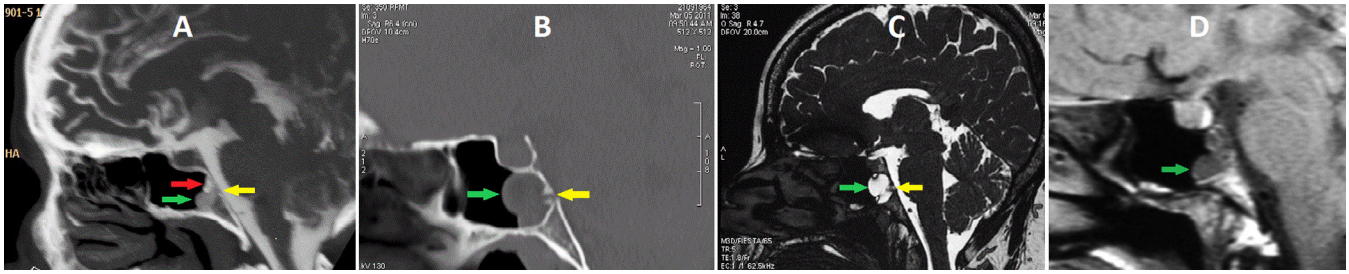

Fig. 1. Computed tomography (CT) and magnetic resonance (MR) images: (A) Midline sagittal CT-cisternography scan demonstrates a cystic lesion inside the sphenoid sinus (green arrow); contrast medium passage from the subarachnoid space to the interior of the sphenoid sinus (red arrow) through a subtle osseous clival defect (yellow arrow) can also be seen. (B) Sagittal CT image shows an enlarged cystic lesion inside the sphenoid sinus (green arrow). A clearly defined osseous defect in the clivus (yellow arrow) can also be seen. (C) Sagittal T2-weighted MR image (FIESTA sequence) demonstrates a homogeneous and hyperintense cystic lesion inside the sphenoid sinus (green arrow) and a small orifice in the clivus (yellow arrow), communicating the pre-pontine cistern within the mentioned sinus. (D) The clival lesion was hypointense on T1-weighted images and revealed no enhancement after Gadolinium (Gd) administration. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

defects were found in the upper third of the clivus. The orifices were corrected using hydroxyapatite bone cement (BoneSource®) and additionally repaired with a pedicled nasoseptal flap. A layer of polyethylene glycol hydrogel sealant (Duraseal®) was finally applied over the mucosal flap (Fig. 2A–D).

Histopathological examination of the lesion revealed a well circumscribed aggregate of cells, without surrounding tissue invasion or bone involvement (Fig. 2E). The cells were immersed in a myxoid and amphophilic matrix and presented numerous and large intracytoplasmic mucin-containing vacuoles – the so called physaliphorous cells (Fig. 2F). No architectural disarray, mitotic figures, cellular atypia or pleomorphism could be seen in the whole specimen (Fig. 2E and F).

Immunohistochemical assays supported the hypothesis of a notochord derived lesion. The tissue was positive for pancytokeratins (AE1 and AE3 clones) and epithelial membrane antigen (EMA) (Fig. 2G and H) and negative for S-100 protein.

The patient did well after surgery and has been asymptomatic during a 2-year follow-up period. Recent radiologic exams evidenced no lesion recurrence.

3. Discussion

Ecchordosis physaliphora (EP) is a benign notochord-derived lesion most often found in the dorsal midline surface of the clivus. It was first described by Luschka, in 1856. However, it was only almost 40 years later that Ribbert experimentally demonstrated its embryological origin and coined the name “ecchordosis physaliphora” [3,4].

EP is rarely symptomatic and is usually found only incidentally in 0.4–2% of all post-mortem or radiological studies [1]. However, some few symptomatic cases have already been reported in the literature [2].

Similar cases of EP associated to CSF fistula had been described only twice before [3,5]. MacDonald et al. described the case of a 66-year-old woman with CSF fistula secondary to a radiologically suspected EP. She was operated via microscopic transsphenoidal approach. The diagnosis was supported by the anatomopathological study [5]. For their turn, Alli et al. reported a 52-year-old woman who also presented with the hypothesis of CSF fistula secondary to EP. She was primarily submitted to an endoscopic endonasal surgical procedure, which was not sufficient for the complete resection of the lesion and prevention of CSF leakage. A microscopic transsphenoidal approach was therefore necessary to treat the patient. The hypothesis of EP was not confirmed after surgery [3] (Table 1).

The main differential diagnosis of EP is chordoma. It is also a notochord derived tumor which is considered the malignant counterpart of EP by some authors [1,3,5]. Differentiating EP from chordomas is often challenging and has very important prognostic and therapeutic implications [3,5]. As no single feature may reliably differentiate both lesions, all clinical, radiological, histological and immunohistochemical characteristics must be taken into account to the correct diagnosis [6].

In the case presented, the hypothesis of EP was initially suspected by radiological findings. Radiology is very useful in demonstrating the indolence and benignity of EP [1,4,6]. Computed tomography (CT) scans usually evidence a subtle and small soft tissue mass posterior to the clivus. Although not always present, an osseous stalk-like projection connecting the lesion to the dorsal aspect of the clivus is considered the pathological hallmark of EP by some authors [1,3,4]. Focal bony dissolution may be seen around EP, depending on its size and on its relationship to the clivus [1].

Magnetic resonance (MR) is very useful in characterizing EP lesions. MR scans often evidence homogeneous masses that present hypointense and hyperintense signals on T1 and T2-weighted images, respectively [1]. Because EP is a scarcely vascularized lesion, enhancement after contrast administration is usually absent [1,4].

Chordomas, for their turn, are considered locally aggressive and highly recurrent neoplasms. Patients with chordomas normally present with much more aggressive symptoms, like focal neurological deficits and severe headache. Distant metastases are also not rare at the moment of diagnosis [1,5]. Radiological images often show heterogeneous masses with irregular intratumoral calcification, bone destruction and surrounding tissue invasion [4]. MR scans may evidence large heterogeneous masses with low T1 and T2-weighted signals [1]. Enhancement after contrast administration on CT and MR scans should always be considered as a fundamental feature in the differentiation of EP from chordomas. Chordomas are highly vascularized lesions and present important heterogeneous enhancement after contrast administration [1,2,4].

Primitive notochord tissue and its variants present striking similar histological and immunohistochemical features [5]. Physaliphorous cells, which are characterized by large mucin-containing intracytoplasmic vacuoles, are typically present in all these lesions. The cells are often immersed in a myxoid amphophilic matrix. Staining positive for vimentin, low molecular weight cytokeratins, epithelial membrane antigen (EMA) and S-100 protein is also common for all these tissues [3,5,6]. Because of all that, these features may point to the embryological origin of the tumors, but are not useful in differentiating benign from malignant masses [2,3,5].