The Laryngoscope

How I Do It

Transnasal Endoscopic Resection of Symptomatic Ecchordosis Physaliphora

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INTRODUCTION

Ecchordosis physaliphora (EP) is a hardly symptomatic condition, most often diagnosed incidentally due to a notochordal remnant and typically occurring between the dorsum sellae and the sacrococcygeal region. We report two cases of transclival EP treated by pure endoscopic approach at two university hospitals.

CASE REPORT

Case 1

A 51-year-old female with a 3-year history of recurrent cerebrospinal fluid (CSF) leakages, misdiagnosed as unilateral allergic rhinorrhea, was referred to the Department of Otorhinolaryngology of the University of Brescia. A computed tomography (CT) scan demonstrated the presence of a fluid collection in the right sphenoid sinus, with evident remodeling of the posterior wall focally interrupted on the midline (Fig. 1A and 1B). Magnetic resonance imaging (MRI) was performed and confirmed in T2-weighted and constructive interference in steady state (CISS) sequences, a fluid collection compatible with CSF. The clival bony defect was associated with a lobulated, nonenhanced mass measuring 12 $\times$ 6 mm. The lesion, which appeared nonhomogeneously hyperintense on T2-weighted images and showed no diffusion-weighed image (DWI) restriction, occurred in proximity to the basilar artery and right abducens nerve (Fig. 1C and 1D).

Case 2

A 39-year-old female, with a long-lasting history of diplopia due to left abducens nerve palsy, was referred to the Department of Otorhinolaryngology of the University of Insubria (Varese). Since the age of 17, the patient had been complaining of mild though persistent left-side retro-orbital pain, as well as diplopia during left far lateral eye movements. The clinicians suspected a Tolosa-Hunt syndrome and treated her with several cycles of steroidal therapy, whose benefits proved very short-lasting. Over the previous 3 months, the patient's clinical picture had been deteriorating significantly due to bouts of sudden sixth-cranial nerve palsy.

A CT scan and a contrast-enhanced MRI revealed a 15-mm lobulated mass at the level of the infrasellar recess, through the posterior wall of the sphenoid sinus and upper clivus. It appeared hypointense on T1-weighted images and hyperintense on T2-weighted images; no contrast enhancement was observed. The lesion was displacing the prepontine cistern and exerting pressure on the left abducens nerve.

Surgical Procedure

Both patients underwent a transphenoidal-transclival endoscopic approach. A unilateral naso-septal flap (NSF), pedicled on the septal branches of the sphenopalatine artery, was harvested. The subsellar recess was reached by removing the sphenoidal intersinusal septa. The sphenoidal floor and upper clivus were drilled out between vertical portions of cavernous internal carotid arteries.

As soon as blunt instruments detached the thin residual bony cortex covering the lesion, a lobulated

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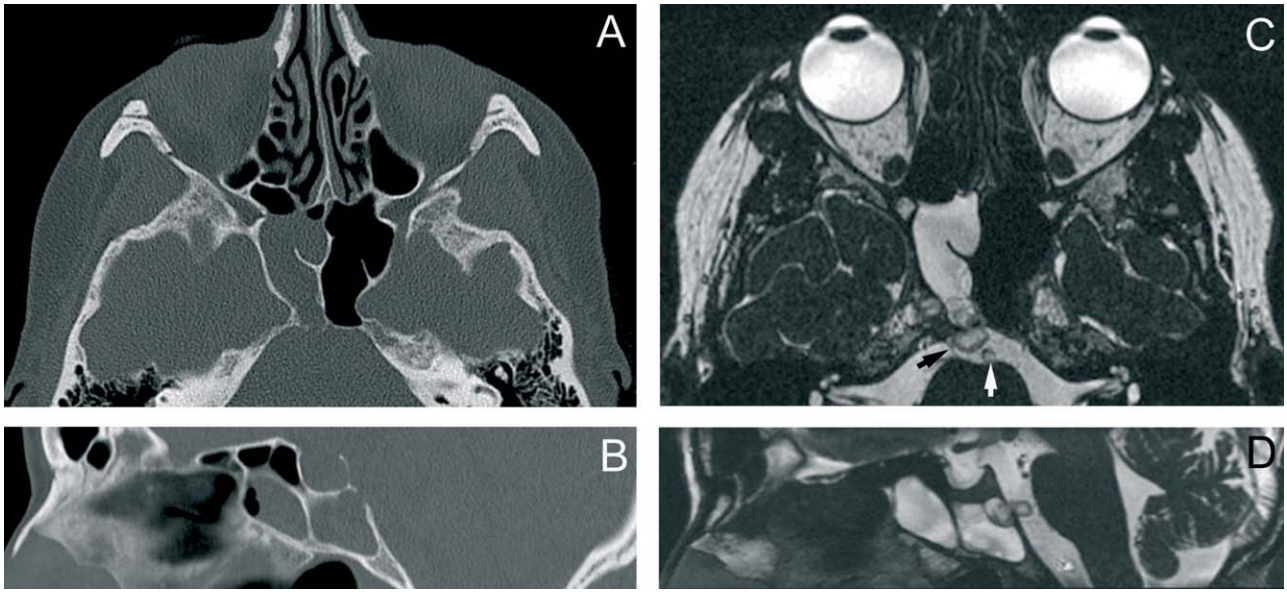

Fig. 1. Case 1: Preoperative axial (A) and sagittal (B) CT scan images showing bone remodelling of the posterior wall of the right sphenoid sinus and upper clivus. Preoperative axial (C) and sagittal (D) T2-weighted MRI: The multilobulated lesion appears to be in contact with the basilar artery (white arrow) and the right abducens nerve (black arrow). [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

cystic mass was identified at the level of the sphenoclival junction. Its thin capsule, once opened, caused a high-pressure gush of CSF, with direct communication between the sphenoid sinus and the prepontine arachnoidal cistern (Fig. 2). The removal of the transclival extradural portion allowed us to observe a well-defined round-shaped median dural defect (Fig. 3), addressed to cause the hourglass-like appearance of the lesion at the MRI.

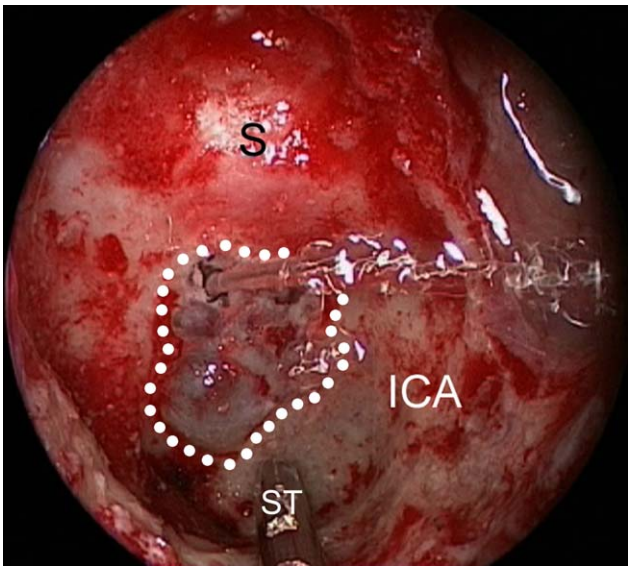

Fig. 2. Endoscopic view by 0° endoscope of case 2. Once the lesion (white dots) is opened, high-pressure CSF-leakage from the prepontine cistern is clearly evident. ICA = left internal carotid artery in its vertical cavernous portion; S = sella; ST: suction tip. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

The skull base defect was repaired by performing a modified "gasket-seal"[1] technique: fat tissue was used in one case and the iliotibial tract in the latter case. In both instances a fragment of adequately shaped septal cartilage was used to fasten the grafts to the residual skull base. By gently pushing the cartilage buttress within the defect, we allowed for the fat and fascial grafts to overlap with the borders of the defect, thus properly sealing it. The NSF was then placed overlay, in order to expedite the healing process and reduce the risk of further CSF leakage. No lumbar drainage was performed.

RESULTS

The standard nasal packing was removed after 48 hours and both patients were able to stand within 4 days. Hospitalization time was 7 and 8 days, respectively, and the postoperative course was uneventful in both cases. Final histological exams confirmed the lesions to be consistent with notochordal remnants. The first patient no longer showed any symptom, while the retro-orbital discomfort combined with the left sixth cranial nerve palsy of the second case showed significant improvements; as to the diplopia, resolution was nearly total. The MRIs we performed 12 months after surgery demonstrated the radical nature of both procedures (Fig. 4).

DISCUSSION

EP is a rare condition typically detected by serendipity, which occurs in 0.5% to 2.0% of autopsies[2] and in

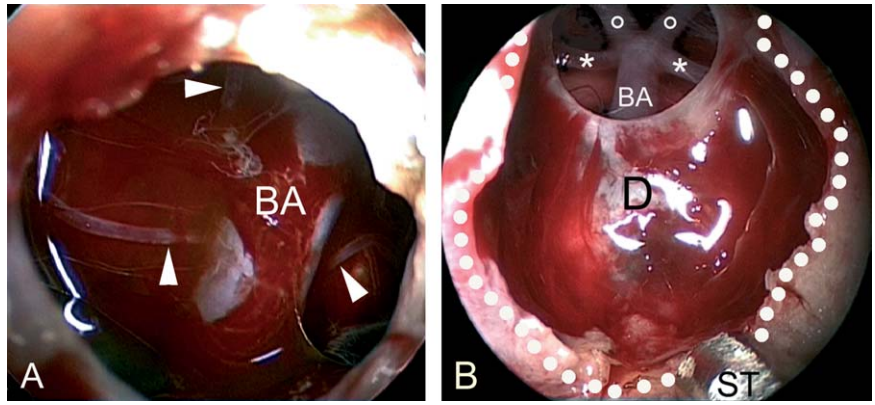

Fig. 3. Endoscopic view by 0° endoscope of case 1 (A) and case 2 (B). A round-shaped medial dural defect is present with exposure of the basilar artery (BA) in both cases. The white dotted line highlights the bony clival defect. white arrowheads = basilar artery perforators; white circles = posterior cerebral arteries; white asterisks = superior cerebellar arteries. D = dura; ST = suction tip. [Color figure can be viewed in the online issue, which is available at wileyonlinelibrary.com.]

1.5% of the MRI[3] and episodically assumes clinical relevance. Only 14 cases had been reported in scientific publications.[4–11] This is why the diagnosis and treatment of EP do not rest upon reliable guidelines. Symptoms typically include headache and diplopia,[11] but the general picture may vary greatly and range from the total absence of symptoms to sudden death due to subarachnoidal bleeding.[6]

The differential diagnosis between EP and clival chordoma (CC) is challenging. In both of these cases, the MRI showed a hypointense signal in the T1-weighed and a hyperintense signal in the T2-weighed images.[12] In CC, however, gadolinium enhancement is variably present, whereas it is always absent in EP.[3] A histology-based differential diagnosis is equally difficult due to their common embryonic notochordal origin. EP typically features a lower Ki67 staining,[13] no mitosis, and a very low MIB count.[11,14]

Surgical treatment is considered the gold standard for symptomatic EP, usually performed by craniotomy.[11] During the last 20 years, the transnasal, endoscopic approach has been safely applied in the treatment of sellar and parasellar lesions.[16,17] The endoscopic transnasal, transclival approach has already proved to be effective in the treatment of clival lesions.[13] The endoscopic "gasket-seal" technique can be performed even if the extracranial attempts of dural repair have already failed.[1] The first case of a retroclival lesion treated with an extended transphenoidal–transclival endoscopic approach was published in 2009 by Ciarpaglini et al.[13]

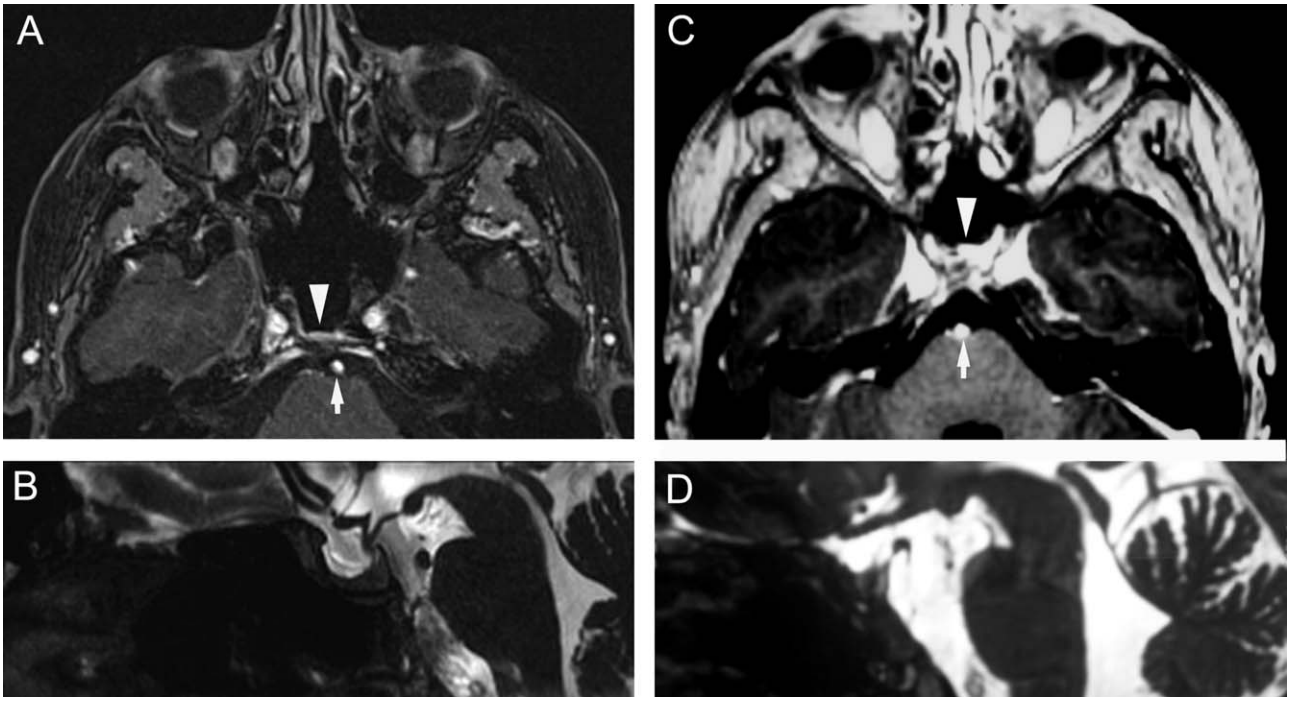

Fig. 4. Postoperative MR examination. Case 1: axial, T1-weighted sequence with fat saturation and gadolinium (A) and sagittal, T2-weighted sequence (B). Case 2: axial, T1-weighted sequence with gadolinium (C) and sagittal, T2-weighted sequence (D). In both cases, the NSF appears well vascularized and properly placed within the infrasellar recess (white arrowhead). The white arrow points at the basilar artery.

In that case, the patient had undergone partial excision of a lesion compatible with EP or intradural chordoma, and duraplasty was performed by a double layer of autologous material and a NSF. There was no need for lumbar drainage and the postoperative course was uneventful; yet the patient kept complaining of memory loss, which had been reported to be the main symptom. Yamamoto et al.[11] reported about an additional case of endoscopy-based EP management in which only a partial excision was achieved and a small dural defect was repaired by synthetic materials and a NSF.

Unlike what reported by Leng et al.,[1] we believe a septal cartilage buttress to be a valid option. In fact, the mild flexibility of the septal cartilage perfectly conjugates an appropriate resistance and safety during the transclival positioning. In addition, the use of a NSF will further stabilize the duraplasty and expedites the healing process.

CONCLUSION

In our experience, the transnasal endoscopic approach is a feasible minimal invasive technique that allows for a complete excision of clival EP with no postoperative complications and limited hospitalization time. High pressure CSF leakages originating from the posterior cranial fossa can be safely managed by multilayer autologous skull-base reconstruction.

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