

High risk of cerebrospinal fluid leakage in surgery of a rare primary intraosseous cavernous hemangioma of the clivus showing meningeal infiltration: A case report and review of the literature

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Abstract

Background: Primary intraosseous cavernous hemangiomas (PICH) of the skull represent an infrequent bone tumor. Although some rare cases of PICHs located in the skull base have been published, to our concern only three cases have been reported in the English literature of PICHs arising within the clivus.

Case Description: We present the case of a patient presenting an isolated abducens paresis due to a rare PICH of the clivus showing also an unusual destruction of the inner table as well as infiltration of the dura mater. Due to this uncommon infiltrative pattern of an otherwise expected intraosseous tumor, a cerebrospinal fluid (CSF)-fistula occurred while performing a transnasal biopsy. The patient recovered successfully without need of lumbar drainage or re-surgery. Additionally, intratumoral decompression was sufficient to relieve the abducens paresis.

Conclusions: Our case provides new and meaningful information about clinical features as well as growth pattern of these rare clival tumors. We also discuss the importance of knowing these peculiarities before surgery in order to plan the optimal operative management as well as to avoid complications while approaching PICHs localized in such a delicate cranial region.

Key Words: Clivus, cavernous hemangioma, cerebrospinal fluid leakage

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INTRODUCTION

Primary intraosseous cavernous hemangiomas (PICH) comprehend only between 0.7 and 1% of all bone tumors.^[19] They can be found in almost all bones, but they tend to affect more frequently the spine and calvarial bones.^[9] Although sporadically cases of PICH affecting the skull base have been described, up to the present

only three cases have been reported of PICH arising from the clivus.^[9,29,30] Few experiences in respect to their clinical evolution, added to the complexity of a surgical approach to the tumors in this location, can constitute a challenge for the surgeon. We present a case of a clival PICH with meningeal involvement. This feature, as well as some aspects of the clinical evolution, make our case

unique with important implications regarding surgical management and post-operative concerns.

CASE REPORT

A 62-year-old female patient consulted because of diplopia, headache, and gait disbalance. From previous clinical antecedents it was remarkable a right carotid-ophthalmic aneurysm, which had been successfully clipped 14 years ago. Despite a hyperthyroid-related osteoporosis, the patient had no other remarkable clinical history. At the presentation in our emergency department, the woman was awake, fully orientated and pupils were isocoric with normal light response. There were no signs of meningeal irritation or sight disturbances. The patient had only a left abducens paresis. Otherwise was cranial nerve and the remaining neurological examination inconspicuous. Initial computed tomography (CT) images showed a large, erosive process infiltrating the sphenoidal body, left petrosal tip, and cavernous sinus, as well as extending into the clivus [Figure 1].

There was no evidence of brainstem or cerebellar infarction or intracranial bleeding. Magnetic resonance imaging (MRI) scans confirmed the presence of a large tumor with the above described characteristics [Figure 2].

The petrocavernosal left internal carotid was completely surrounded by the tumor [Figure 3]. The mass grew within the clivus, infiltrated the ethmoidal holes, epipharynx and extended up to the left petrooccipital synchondrosis. The signal was homogenic hyperintense in T1-weighted images and T2 was homogeneously hyperintense.

There was evidence in both CT and MRI of garland-like calcifications and areas of hyperostosis. Nevertheless the tumor showed a clear erosive growth with an expansive destruction of the described bone structures [Figure 1] and there was evidence of possible dural adhesions to the tumor [Figure 2]. It was decided to perform a biopsy and partial excision through a transnasal transesphenoidal approach in order to clarify the histological nature of the tumor and plan the following therapeutic strategy. The procedure was performed under navigation

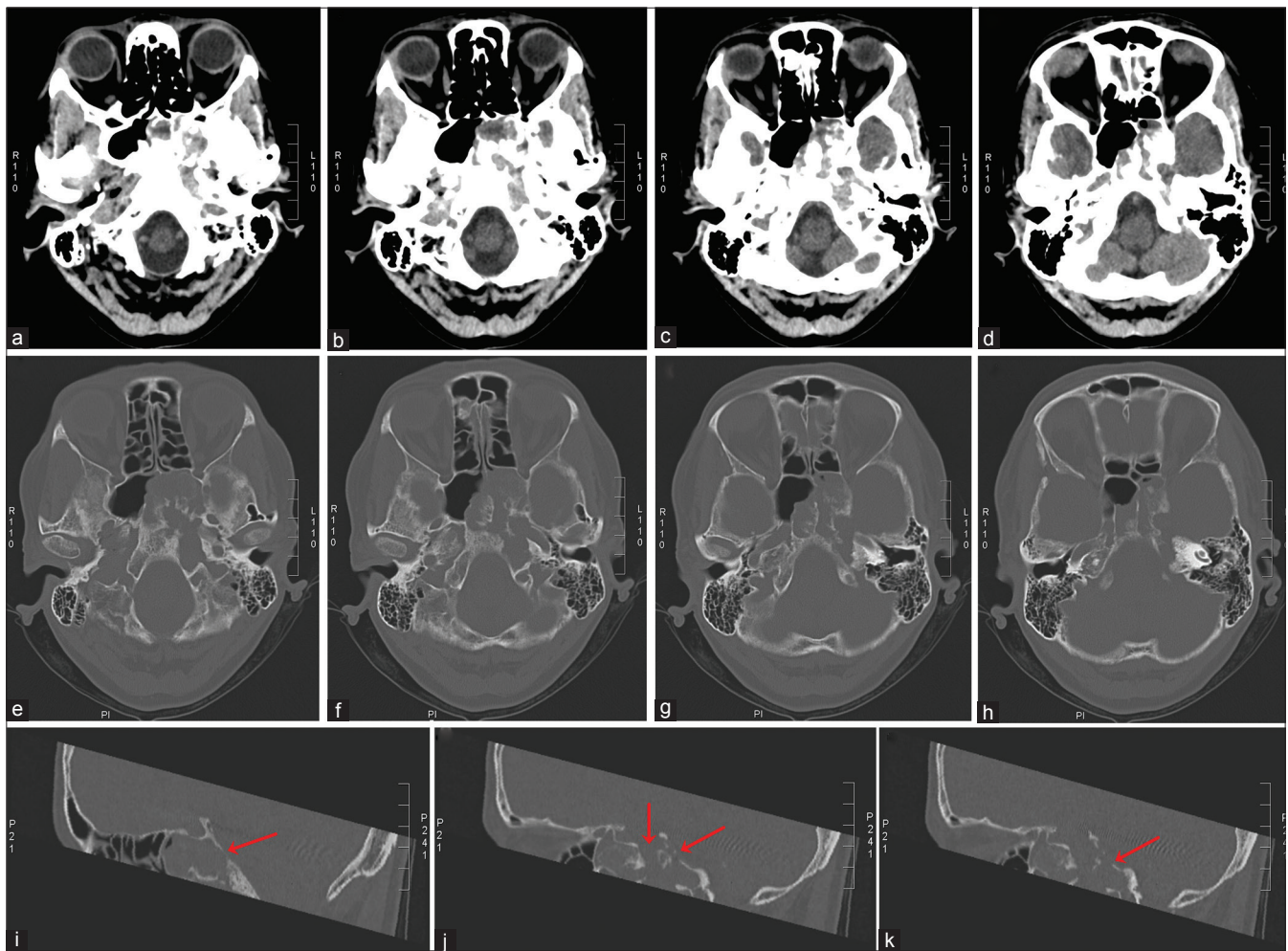


Figure 1: Axial (a-h) and sagittal (i, j, k) reconstructed CT images showing a clear involvement of bone structures within the skull base. It is remarkable that the tumor showed infiltration and destruction of the tabula interna in many sites (arrows), (i, j, k) which corresponds to the extension and adhesion to the dural layer

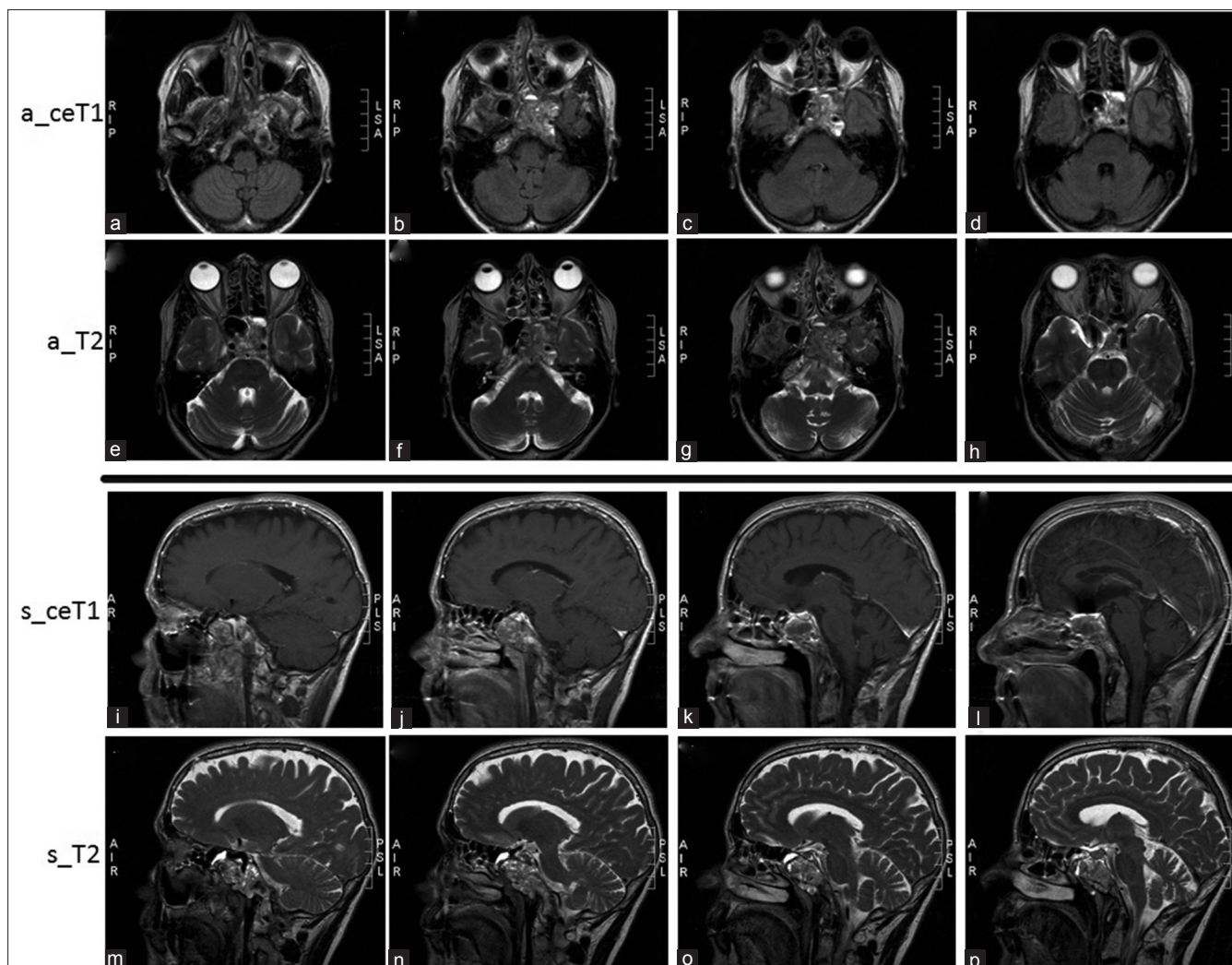


Figure 2: Axial (a_) and sagittal (s_) contrast enhanced T1-weighted (ceT1) and T2-weighted MRI images showing a heterogeneous lesion involving the clivus, (a, b, j, k, n, o) sphenoidal bone, (c, d, g, h, k, o) sphenoidal wing, (b, i) as well as extending to the left petrosal apex and cavernous (b, d, g, h, i, m) sinus

guidance [Figure 4a]. At opening the esphenoidal sinus it was demonstrated a brown-green tissue with multiple hemorrhagic areas and a trabecular aspect [Figure 4b]. Both trabecular and compact bone of the clivus was eroded. The tumor reached the predural space and infiltrated the duramater, preventing a clear recognition of this structure. Despite a careful dissection, following tumor excision exposed the subarachnoidal space and cerebrospinal fluid (CSF) flowed through a small perforation [Figure 4c and d]. The gap was sealed by using muscle graft [Figure 4e], fibrinogen-fibrin-based sponge (TachoSil®) and reabsorbable gelatine (Mabagelan®) applied in a multilevel pattern.

The further histopathological evaluation revealed a cavernous hemangioma. The immediate postoperative evolution was favorable. The patient developed no additional neurological deficits and even the left abducens paresis regressed. On first instance, there was no evidence of CSF fistula and the patient was discharged 5 days later. However, on re-admission 2 days later because of rhinoliqorrhoea, which stopped spontaneously without

need of lumbar drainage or any other invasive measurement. The same phenomenon occurred 4 weeks later. Although the presence of CSF was confirmed by positive traces of beta transferin, the rhinoliqorrhoea ceased without any intervention. An otorhinolaryngologist examination including a meticulous rhinopharyngoscopy revealed no evidence of an active CSF fistula and no more episodes of rhinoliqorrhoea have occurred up to the present.

DISCUSSION

General overview of cavernous hemangiomas of the skull

Among all primary bone tumors, PICH account for only 0.7–1% of all bone tumors.^[9] Although they can occur in almost all bones, the most frequent localization is the spine (30–50%) and skull (20%).^[9] PICH of the cranium account for 0.2% of all benign skull tumors^[16,18,31] and affect mostly frontal and parietal bones.^[10-12,17-20,25,26,31] PICH within the skull base constitutes an extreme

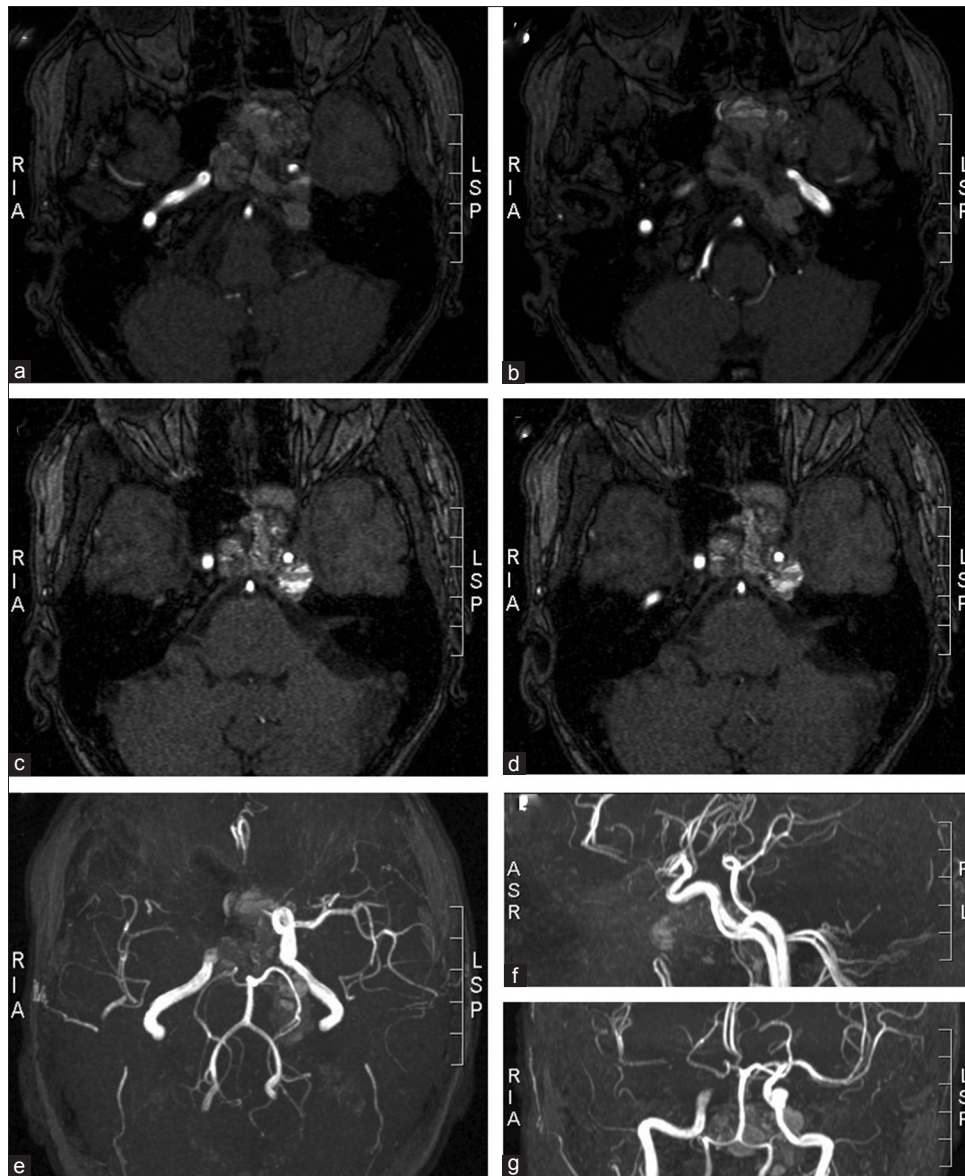


Figure 3: MRI angiographic scans show the relationship between the tumor and major vessels of the skull base. The intracavernous and supraclinoidal segment of the left internal carotid was surrounded and compressed by the tumor (c, d, e, g). However, sufficient blood supply to the left medial and anterior brain arteries provided by the right carotid circulation through the Willis circle (e, f, g), as well as a slow tumor growth enabling collateralization, can explain the lack of neurological deficits besides the ones produced by a direct tumor compression of cranial nerves

uncommon feature and the structure affected in the vast majority of cases is the petrous bone.^[2-4,8,14,15] These tumors have been anecdotally described in other skull base bones, such as the sphenoidal wing,^[5,16] sphenoid body,^[27] and occipital condyle.^[7] To date, only three cases have been reported of PICH of the clivus.^[9,29,30]

Because of their clinical evolution, radiological features, and complexity of a surgical approach the management of skull base PICH differs significantly from those arising in cranial vault bones. Furthermore, lack of cases in the literature on skull base PICH compared with calvarial PICH constitute an additional challenge for surgeons, at the moment, for deciding the proper management of these tumors.

PICHs of the cranial vault manifest in most of the cases as clinically indolent palpable masses. They tend to grow very slowly and, conform they expand, patients complain of headache.^[17,31] However, due to their tendency to grow in an extracranial pattern, neurological deficits are evidenced only in rare cases of calvarial PICH with intracranial expansion.^[21,23,32]

By contrast, PICH comprising orbital or skull base bones can be associated to a higher morbidity due to their tendency to involve novel neurovascular structures during their growth. Orbital PICH have been described to produce progressive proptosis, diplopia, and visual loss in some severe cases.^[1,13,24]

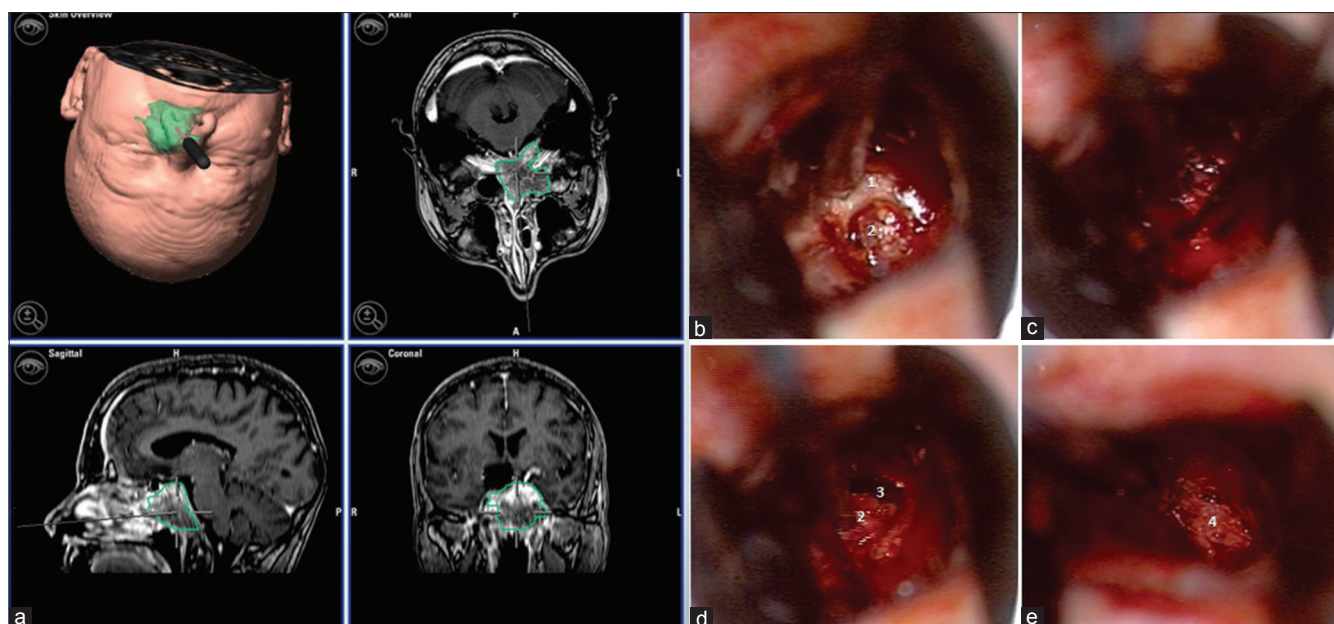


Figure 4: Microsurgical tumor biopsy performed with neuronavigation (a) through a transnasal transesphenoidal approach. After osteotomy of the esphenoidal sinus (b), the tissue showed a strong tendency to bleed (c). The destruction of the tabula interna and adhesions to the dura made the opening of subarachnoid space almost unavoidable (d) and muscle (e) was used to close the dural defect. Esphenoidal bone (1), tumor (2), subarachnoid space (3), and muscle graft (4)

PICH of petrosal bone commonly impair facial nerve function,^[4,6,8] although lower cranial nerve involvement has been also described in a PICH of the petrosal bone compressing the jugular foramen.^[8] A rare case of PICH localized in the petrosal bone has also been reported, which manifested with a spontaneous bleeding and subsequent epidural hematoma.^[15] Moreover, localization within the sphenoid body has been associated to compression-induced optic atrophy.^[27]

Nevertheless, some large PICH of the skull base can grow without disturbing cranial nerve function. In the case reported by Liu *et al.* of a sphenoid wing PICH, the indication for surgical removal was due to the increase in severity of headache along a period of many years of follow up, but the tumor did not produce cranial nerve deficits.^[4] Similarly, the case of occipital condyle PICH published by García-Marín *et al.* was associated with neck pain and torticollis, but not with neurological impairment.^[7]

Regarding the only few cases of clival PICH, these tumors can course with minimal symptomatic manifestation or, in other cases, they may be related to severe progressive cranial nerve deficits including Garcin syndrome, characterized by a dysfunction of cranial nerves from the V to XII.^[9] Interestingly, our case is the first describing an isolated abducens paresis produced by a PICH of the clivus. Additionally, the fact that the paresis remitted during the early postoperative follow up is also remarkable, considering that no mass reduction was attempted during the surgery. We propose that probably opening the tumor cavity relieved the intratumoral pressure and therefore the compressive effect on the abducens nerve.

With respect to the radiological features, it is important to remark that the typical and almost pathognomonic characteristics of vertebral intraosseous hemangiomas such as honeycomb or polka-dotted pattern on CT are often absent in PICH.^[9,16,28-30] MRI images are also unspecific, with heterogeneous signal in both T1- and T2-weighted sequences depending on the amount of venous flow and fatty transformation within the tumor.^[9,29,30] Moreover, PICH cannot be evidenced on angiography, making this study useless to clarify the diagnostic preoperatively.

Additionally, it is accepted that intraosseous cavernous hemangiomas respect in all cases the cortical bone layer and this feature distinguishes these lesions in radiological studies from bone malignancies.^[16] Nevertheless, another unique characteristic in the case is that the internal cortical layer was not respected, presenting the tumor adhesions to the duramater. This feature is unique in our case and suggests that PICH of the clivus could be difficult to distinguish preoperatively from malignant infiltrating neoplasms, such as chordoma.

Assessing a surgical management of cavernous hemangiomas of the clivus and other deep skull base structures: Relevance of being aware about possible dural adhesions and risk of CSF leakage

Due to the fact that PICH have the tendency to grow slowly but they do not show involution, it is suggested that symptomatic PICH should be removed en bloc when possible, because the recurrence after en bloc resection is extreme rare.^[16,22]

Although this issue seems to be clear in cases of PICH localized in the cranial vault, whether en bloc or even a partial resection of a skull base PICH should be performed remains unclear. As mentioned earlier, the low incidence of PICH located deeply in the skull base, such as in clivus, makes it difficult to assess and predict the clinical evolution of these tumors with or without surgery.

The higher tendency to compromise novel structures due to the slow but constant growing as well as the high curability provided by a complete tumor removal constitutes the main argument to attempt a radical tumor excision when possible. In addition, the concept that PICH grows exclusively intraosseous, respecting the cortical bone layer and without adhesions to the meninges, favors the idea of trying to aim a gross tumor removal. Given this condition, a complete tumor resection through an intracranial extradural approach may be feasible, even in tumors located in delicate regions of the skull base.^[16] However, if extensive skull base involvement with neurovascular encasement makes a complete resection unfeasible, a subtotal resection may be indicated to relieve symptoms of mass effect.

We demonstrate that PICH of the clivus may present exceptionally not only in involvement of the internal cortical bone layer, but also in adhesion to the dura. Although we report the first case of dural involvement in clival PICH, this feature has been already reported in three cases of PICHs arising from cranial vault bones.^[21,23,32] This eventuality has very important implications at the moment of deciding a surgical approach to a PICH arising from the clivus or other bones of the skull base. The adhesion to the dura would constitute an obstacle for the surgeon while attempting a radical tumor removal through an anterior approach to the clivus because of the high risk of CSF leakage and concurrent postoperative meningitis. If an en block resection is planned, other surgical approaches should be taken in consideration, in order to provide a better accessibility for placing a dural graft. Additionally, if a complete tumor removal is intended, the surgeon should try to choose an approach that avoids opening the pharyngeal mucosa in the aim to reduce the risk of meningitis if the dura is to be opened. If a transnasal approach is selected and a high flow CSF leak is observed, some colleagues would recommend closing the dura interposing a pedicled vascularized flap, particularly the nasoseptal flap, due to its demonstrated efficacy in reducing the risk of post operative CSF leak. Finally, if the surgeon decides to perform a diagnostic frameless stereotactic biopsy, the considerations in respect to the depth of tissue excision should be also carefully reviewed in order to avoid possible dural adhesions.

CONCLUSIONS

Skull base PICH and specially clival PICH represent a very infrequent tumor entity, which can manifest from almost

no symptoms to multiple cranial nerve impairment. Because of their unspecific radiological characteristics, a frameless stereotactic biopsy may be always considered in lesions involving these structures in order to confirm the diagnosis and exclude other entities. Furthermore, we have shown that some neurological deficits can resolve after opening the tumor cavity, probably due to a reduction of intracavitary pressure and its compressive effect on neural structures.

The need of a radical surgical tumor removal in clival PICH should be always pointed out against the risks and benefits for every individual patient and an attempt to reach a more radical removal should be considered for patients with newly developed symptoms or evidence of tumor progression during the follow up. Moreover, we have evidenced that occasionally dural adhesions may be present and this should not be ignored at the moment of planning a stereotactic biopsy or tumor resection.

Ethical standards and conflict of interest

The manuscript does not contain clinical studies or identity patient data and the authors declare that they have no conflict of interest.

Patient consent

The patient has given consent to the submission of the case report for submission to the journal.

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Commentary

The authors report a cerebrospinal fluid (CSF) leak during transnasal surgery for a rare case of primary intraosseous cavernous hemangioma (PICH) of the clivus. The tumor showed atypical findings of involvement of the internal cortical bone layer and adhesion to the dura mater increasing the risk of CSF leak.

The clinical presentation of PICH is variable ranging from no symptoms to multiple cranial nerve deficits. Classical radiographic findings of sunburst trabeculations, honeycomb configurations, and hypervascularity on angiography are uncommon features in the skull base. They can mimic other more common skull base lesions and the diagnosis is most often made after resection or biopsy.^[1, 2]

Indications for surgical removal include mass effect, neurological compromise, and cosmetic deformity. En bloc resection is recommended with establishment of normal bony margins, but in extensive skull base involvement with neurovascular encasement the complete resection becomes unfeasible. In these cases, a subtotal resection may be performed to relieve symptoms of mass effect.^[1, 2]

Although transnasal approach to the clivus represents an increasingly important surgical corridor for minimally invasive skull base surgery, CSF leak remains a major concern. The adoption of pedicled vascularized flaps over the past decade has significantly reduced the rate of CSF leak. The need for vascularized flap reconstruction is

governed primarily by defect size and location, and by the presence of a high-flow CSF leak.^[3, 4]

As shown by the authors, dural infiltration in PICH of the skull base is uncommon and associated with high risk of CSF leak. If a transnasal approach is selected, it is important to consider the closure during the opening time preparing the pedicled vascularized flap, particularly the nasoseptal flap,^[4] to seal the dura mater defect and prevent postoperative CSF leak.

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