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Case Report

Primary intraosseous cavernous hemangioma of the clivus: A case report and literature review

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ABSTRACT

Background: The radiographic presentation of the primary intraosseous cavernous hemangiomas (PICHs) is nonspecific. We report a case of clival PICH mimicking a chordoma with a literature review.

Case Description: A 57-year-old woman presented with diplopia that started a few days before the presentation. She had transient diplopia at the right lateral gaze and upper gaze with normal eye movement. The symptoms disappeared spontaneously 1 week later. She had no other complaints or neurological deficits. Computed tomography revealed an intraosseous mass lesion and bone erosion of the middle and lower clivus, extending laterally to the right occipital condyle. Magnetic resonance imaging (MRI) showed hyperintense and hypointense components on T2- and T1-weighted images, respectively. The lesion was larger than on MRI performed 10 years earlier. Chordoma or chondroma was considered a possible preoperative diagnosis. An endoscopic transsphenoidal approach removed the tumor. In the operating view, the lesion appeared as "moth-eaten" bony interstices filled with vascular soft tissue. Histologically, an intraosseous cavernous hemangioma was diagnosed.

Conclusion: Diagnosis before surgery is difficult without characteristic radiographic findings. When making a differential diagnosis of malignant skull lesions, PICH should be considered.

Keywords: Case report, Cavernous hemangioma, Clivus, Endoscope, Transsphenoidal surgery

INTRODUCTION

Primary intraosseous cavernous hemangiomas (PICHs) are rare, benign osteolytic tumors that are vascular in origin. PICHs constitute 0.2% of all bone neoplasms [2,9] and hardly ever involve the base of the skull. There have only been four recorded cases of clival PICH thus far.[1,4,7,8] The clinical course differs among cases, ranging from chronic headaches to multiple neurological deficits. Without properly established management protocols, surgeons must create individualized treatment plans for each patient. Herein, we report a case of clival PICH mimicking a chordoma, including a literature review.

CASE REPORT

History

A 57-year-old woman came to our hospital with diplopia, which had started a few days before the presentation. At presentation, she had transient diplopia in the right lateral and upper gaze, with normal eye movement. Her symptoms disappeared spontaneously 1 week later, and she had no other symptoms or neurological deficits. Computed tomography (CT) showed an intraosseous mass lesion and bone erosion of the middle and lower clivus, extending laterally to the right occipital condyle [Figure 1]. Magnetic resonance imaging (MRI) showed components that were hyperintense on T2-weighted and hypointense on T1-weighted images [Figure 2]. The lesion was larger than it had been on the first MRI study done 10 years earlier. Neither 11C-methionine positron emission tomography (PET) nor 18F-fluorodeoxyglucose PET revealed significant accumulation. Thus, a preoperative diagnosis of chordoma or chondrosarcoma was made.

Treatment

Tumor resection using an endoscopic transnasal transsphenoidal trans ethmoidal approach was planned. A surgical navigation system was used to aid in the intraoperative localization of the lesion. Tumor resection was performed through the right nasal cavity. Using a zero-degree endoscope, we opened the ethmoidal bra and posterior ethmoidal sinus and passed it to the sphenoidal sinus. The middle turbinate was translocated laterally, and the anterior wall of the sphenoid bone was drilled through the nasal septum. A protruding tumor was observed through the cortical bone [Figure 3a]. The cortical bone was subsequently drilled out, and the tumor (red-colored and encapsulated) was exposed.

The gross inspection did not indicate the tumor to be either a chondroma or chondrosarcoma, and hemangioma was suspected. We subsequently exfoliated the preserved cortical bone and coagulated the tumor capsule. The tumor capsule was expansive, noninvasive in the bone, and contiguous in the multiple cavities. The cavities were opened with a drill, and the tumor was removed with the capsule preserved, and we did not encounter uncontrollable bleeding in that way [Figures 3b-d]. The zero-degree endoscope was set to 30° one to inspect the lower clivus. Finally, the tumors were grossly resected by intraoperative findings with neuronavigation.

The preserved mucous membrane of the sphenoid sinus was spread into the cavity and fixed with fibrin glue. Pathological examination revealed an intraosseous cavernous hemangioma [Figure 3e].

Postoperative course

The patient was moved from the operating room to the intensive care unit following successful extubation. She remained at the neurological baseline and was transferred to the regular floor on the 1st postoperative day (POD).

She was discharged from the hospital on POD9 with her neurological condition stable. Postoperative MRI [Figure 4] revealed a residual tumor in the central area of the clivus covered with the cortical bone. The residual tumor was unchanged on 6-month and 12-month follow-up MRI. We plan to follow-up MRI once a year.

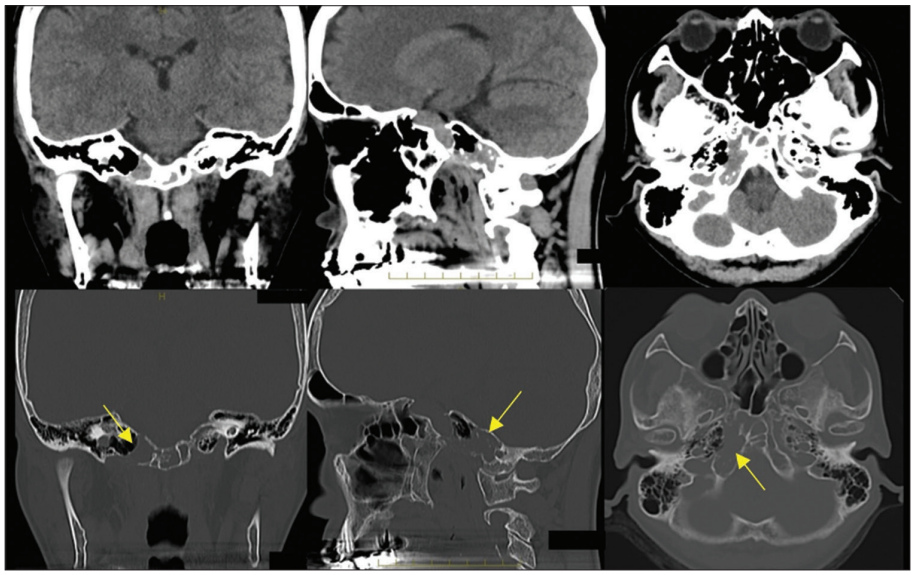

Figure 1: Computed tomography demonstrating the intraosseous mass lesion and bone erosion of the middle and lower clivus, extending laterally to the right occipital condyle (arrows).

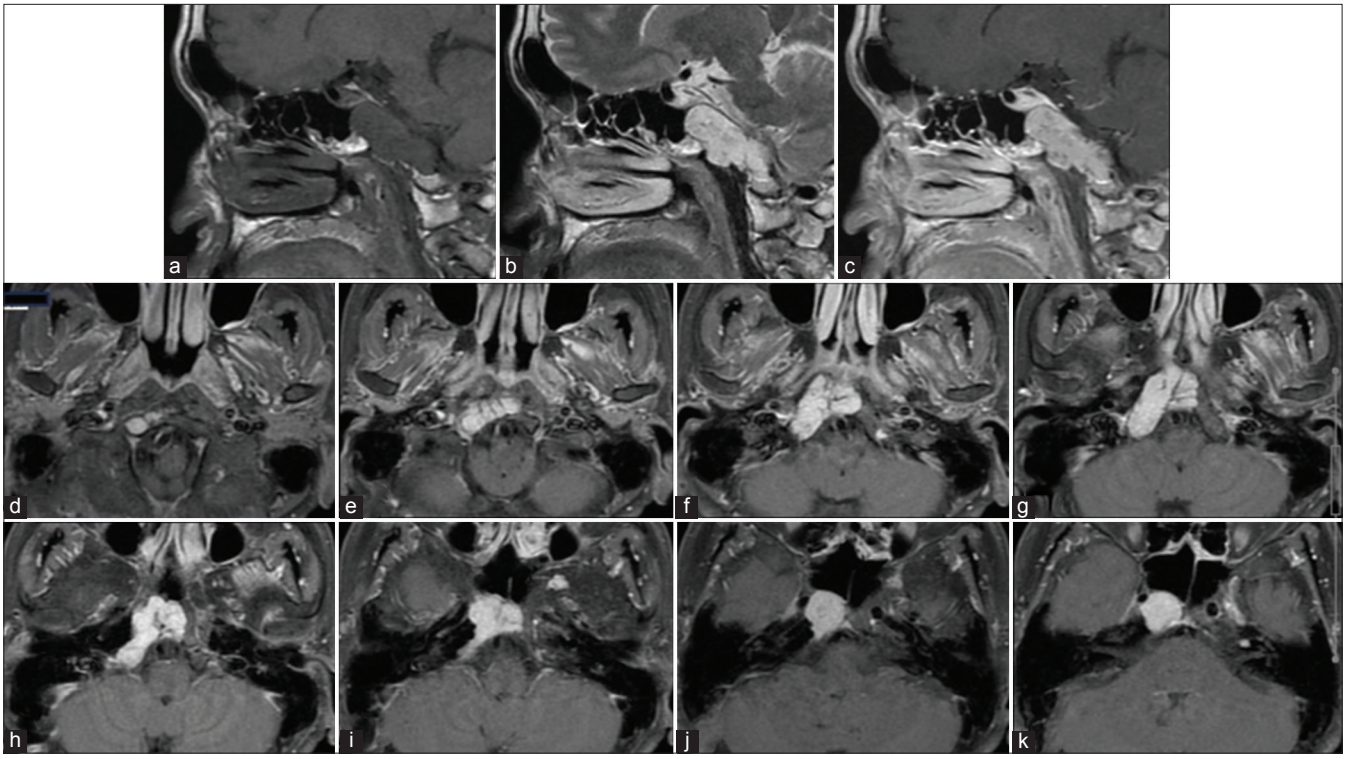

Figure 2: (a) Magnetic resonance imaging showing hypointensity on T1-weighted imaging and (b) hyperintense components on T2-weighted imaging in the lesion. (c) After gadolinium administration, the tumor is markedly enhanced in a homogenous fashion. (d-k) The axial images show the tumor area. On the upper side, the tumor became markedly enhanced in a homogenous fashion. On the upper side, the tumor protrudes in the sphenoidal sinus.

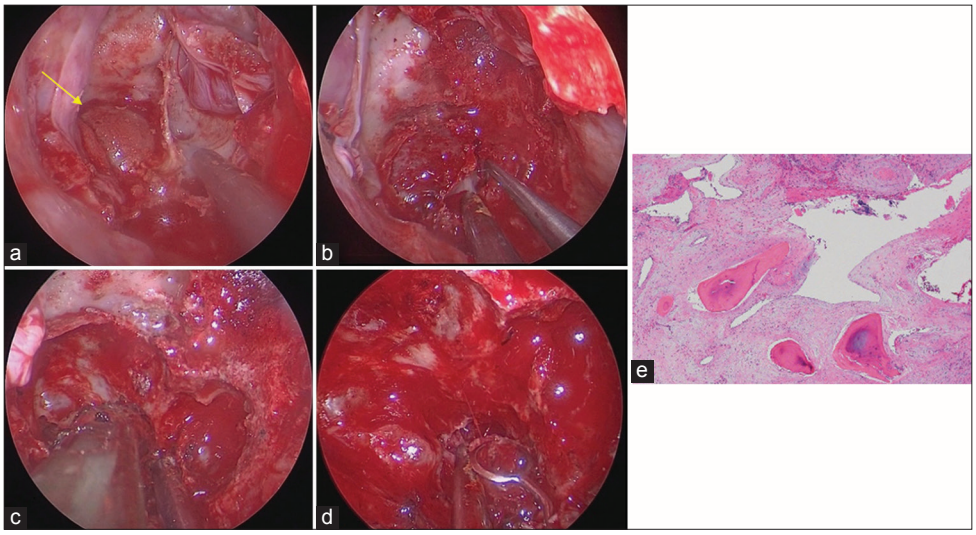

Figure 3: (a) Inside the sphenoid sinus, the protruding tumor (arrow) can be seen extending throughout the cortical bone. (b-d) Along the preserved cortical bone, the tumor was exfoliated and coagulated. (e) A single layer of flattened endothelial cells scattered amid bony trabeculae bordered the thin-walled vascular channels shown by pathological testing, which was consistent with an intraosseous cavernous hemangioma.

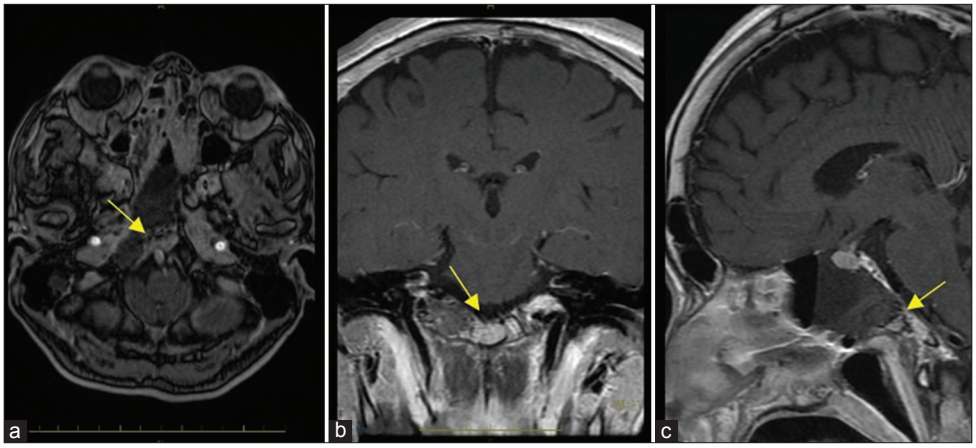

Figure 4: (a) In the spoiled gradient recalled acquisition in the steady state-gradient refocused axial image and (b and c) T1-weighted magnetic resonance imaging using gadolinium-based contrast coronal and sagittal images, the residual tumor (arrows) can be seen in the central area of the clivus covered with the cortical bone.

Table 1: Comparison of the four previously reported cases and our case of primary intraosseous cavernous hemangioma of the clivus.

	Age (years)	M/F	CT	MRI T1WI	MRI T2WI	Angiography	Preoperative diagnosis
No. 1[7]	37	F	Honeycomb pattern	Mottled high-intensity areas in the tumor	Heterogeneously hyperintense	Tumor shows a dense stain	PICH
No. 2[1]	89	F	Only osteolytic area	Mottled high-intensity Areas in the tumor	No data	No significant tumor blush	Metastatic bone tumor
No. 3[4]	62	F	Only osteolytic area	Homogeneously hyperintense	Homogeneously hyperintense	No data	Unknown
No. 4[8]	45	F	Honeycomb pattern	Homogeneously hypointense	Homogeneously hypointense	No significant tumor blush	PICH
Our case	57	F	Only osteolytic area	Homogeneously hypointense	Homogeneously hyperintense	No data	Chordoma/chondroma

M: Male; F: Female; CT: Computed tomography; MRI: Magnetic resonance imaging; T1WI: T1-weighted imaging; T2WI: T2-weighted imaging; PICH: Primary intraosseous cavernous hemangioma

DISCUSSION

PICHs are rare, benign, osteolytic tumors that are vascular in origin. PICHs generally occur in adults, and patients are predominantly women in their fourth–fifth decades of life. PICHs of the skull primarily affect the frontal and parietal bones but rarely involve the base of the skull.[4] They hardly ever involve the base of the skull. PICHs may present as reticulated, honeycomb-like, or sunburst-shaped radiolucent unilocular or multilocular lesions. Some cases can be diagnosed based on characteristic radiographic findings. Nonetheless, in numerous instances, traditional characteristics are lacking, manifesting solely as growing or osteolytic dense bone masses.

A review of the English literature revealed that only four reported cases of clival PICH were reported.[1,4,7,8] We compared the findings in our case with those in the previously reported cases [Table 1]. The patients were between the ages of 37 and 89 (average 58 years), and all were women. On CT, two patients had honeycomb patterns, which are characteristic of PICH, thereby allowing a correct preoperative diagnosis. On T1-weighted imaging, three cases showed hyperintensity. The remaining two cases, including our case, showed hypointensity. On T2-weighted imaging, four cases showed hyperintensity. Angiography was performed in three cases, one of which showed dense tumor staining.

Typical CT signs, such as a honeycomb pattern, are most likely helpful for diagnosis; however, some cases lack these findings.[1,2,7,8] The signals on MRI are erratic and mostly based on the degree of slow venous blood flow and the ratio of red to converted fatty marrow. Lesions with a higher fat content or smaller size generally exhibited stronger signals, especially in T1-weighted sequences. Lesions characterized by slow venous flow or blood pooling showed a higher signal intensity on T2-weighted sequences. Tumor adhesions to the dura mater were visible in one of the five cases where the internal cortical

layer was missing.[4] These tumors are difficult to differentiate from malignant infiltrating neoplasms.

As very few cases of skull base PICH have been reported in the literature, there is currently no consensus on proper management.[4] According to one study, the requirement for a conclusive diagnosis, newly manifested symptoms, or evidence of tumor development during follow-up could all be signs that the tumor should be surgically removed.[4] Preferably, normal bone margins should be preserved after *en bloc* resection. However, subtotal resection is the only feasible option for the skull base. It is currently unclear whether radiotherapy is effective for the treatment of intraosseous hemangiomas of the skull base. For insufficient surgical resection, several authors endorse radiotherapy as an additional or alternative therapy.[2,3,5,6]

CONCLUSION

PICH of the skull base frequently mimics other, more typical tumors of the skull base, including chordoma. Herein, we reported a case of a clival PICH mimicking chordoma and further provided a review of all other previously reported cases. Symptoms at presentation varied in each case. In our case, she had transient diplopia. We believe that the cause of the transient symptom is that repeated bleeding and fluctuated size of PICH made compression to the VIth cranial nerve stronger temporarily. Diagnosis of PICH before surgery can be difficult without characteristic radiographic findings, but the improvement or fluctuations of symptoms may indicate no malignant tumors with a progressive course. When making a differential diagnosis of malignant skull lesions, PICH should be considered.

Ethical approval

Institutional Review Board approval is not required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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