



Intraosseous Hemangioma of Sella: Case Report and Review of Literature

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Key words

- Mixed hemangioma
- Pituitary
- Primary intraosseous hemangioma
- Sella turcica
- Skull base
- Sphenoid

Abbreviations and Acronyms

- ICA:** Internal carotid artery
MRI: Magnetic resonance imaging
PIH: Primary intraosseous hemangioma

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INTRODUCTION

Primary intraosseous hemangiomas (PIH) comprise about ~1% of all primary bone neoplasms.¹ These infrequent tumors are benign vascular neoplasms, most commonly involving the vertebra and only rarely involving the calvarium.² They have been reported to occur in craniofacial and orbital bones.^{3,4} Involvement of the skull base is rare. Lesions have been reported in petrous bone, greater wing, and body of sphenoid and clivus.^{5,6} To the best of our knowledge, there has been no documented report of a PIH involving the sella. We report such a case of intraosseous cavernous hemangioma of the sella presenting as a space-occupying intrasellar mass, and we discuss its management in the context of the pertinent literature.

■ **BACKGROUND:** Primary intraosseous hemangioma (PIH) of the skull base, when localized in the sella, is a rare, benign lesion that can mimic other common sellar tumors. Such tumors may be asymptomatic incidental radiologic findings or present with nonspecific symptoms (e.g., headaches).

■ **CASE DESCRIPTION:** We present a case of a primary intraosseous hemangioma of the body of sphenoid bone extending into the sellar cavity, clinicoradiographically mimicking an atypical pituitary adenoma.

■ **CONCLUSIONS:** PIH should be included as a rare differential diagnosis in cases of space-occupying sellar lesions with atypical features. Radiologic and intraoperative findings may be suited to entertain a probable diagnosis; however, a definite diagnosis can only be obtained via histopathologic analysis. Surgical excision may be chosen under the assumption of dealing with a primary pituitary lesion, but extent of resection depends on the accessibility, extent, involvement of surrounding structures (such as the internal carotid artery/cavernous sinus), and control of intraoperative bleeding. When facing inoperable or residual lesions, radiotherapy can be a viable option.

CASE HISTORY

A 29-year-old gentleman with no known comorbidities presented with complaints of headache for a duration of 1 month, which had been worsening over the past 10 days. He also complained of acute and progressive visual loss in the left eye for 10 days. He sought medical attention, and all regular laboratory and endocrinologic studies returned within normal limits. Ophthalmologic examination revealed a relative afferent pupillary defect of the left eye indicative of compressive optic neuropathy; extraocular movements were full. However, visual evoked potential study of both eyes was normal.

Gadolinium-enhanced magnetic resonance imaging of the brain was obtained. A space-occupying lesion at the skull base involving the sella and sphenoid was noted, measuring 19.17 × 17 × 15.2 mm. It displayed intense homogenous enhancement of contrast, as well as erosion of the sellar floor and general expansion of the sella turcica, more toward the left of midline. Superior extension created mass effect on the prechiasmatic segment of the left optic nerve. There was also inferior extension into the sphenoid sinus by

eroding its roof and mild left parasellar extension. Laterally, the mass was abutting the medial wall of the left cavernous sinus, making contact with the cavernous portion of the left internal carotid artery (ICA). The lesion appeared to compress the pituitary posteriorly, and the infundibulum appeared thickened and displaced superiorly. The intrinsic signal of the optic chiasma appeared normal (Figure 1).

On the basis of clinicoradiologic features, we suspected a possible differential of atypical pituitary adenoma followed by the possibility of craniopharyngioma and hypophysitis.

Assuming that, the patient was planned for excision via the endoscopic transnasal transphenoidal approach. This was set up in a standard fashion. After sphenoidotomy, the roof of the sphenoid appeared unremarkable endoscopically, in contrast to the findings noted on magnetic resonance imaging (MRI). While drilling the anterior wall of sella, unusually profuse bleeding was noted. After the sellar floor was opened, a highly vascular compressible mass was encountered in close proximity to the left ICA and cavernous sinus.

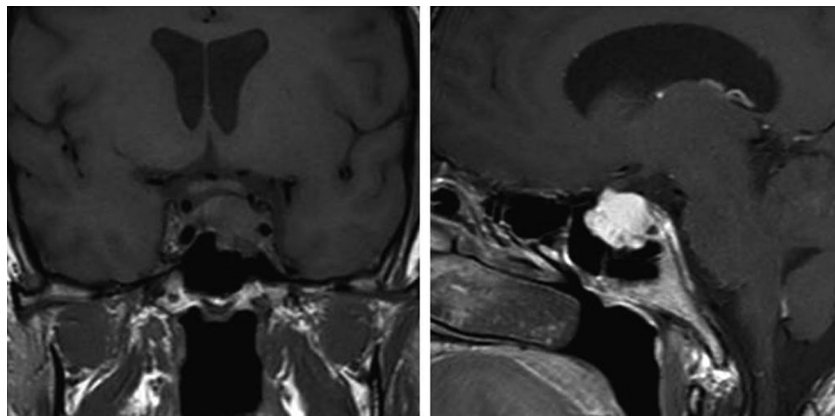


Figure 1. Coronal noncontrast T1-weighted sequence and sagittal T1-weighted sequence following contrast showing a lesion involving the body of sphenoid extending into the sellar cavity that enhances brightly with contrast.

In view of inadvertent profuse bleeding encountered intraoperatively, we were unable to obtain a clear plane of dissection; hence partial resection was done, and the tissue along with bony bits were sent for fresh-frozen analysis. Pathologic assessment of the frozen section of the soft tissue component revealed features suggestive of hemangioma. Due to profuse intraoperative bleeding, complete resection could not be achieved and achievement of complete hemostasis was focused on. At the end of the case, the sella defect was packed with autologous fat, which was reinforced with Surgicel (Ethicon,

Somerville, New Jersey, USA) and fibrin glue. Nasal packings were applied and removed with caution on postoperative day 2. There were no other complications encountered during the case, and the patient had an uneventful recovery.

Complete histopathologic evaluation was consistent with that of a benign intraosseous vascular neoplasm. The final pathologic readout reported a mixed hemangioma (**Figure 2**).

Given that our resection remained subtotal, we discussed the case with our multidisciplinary team and the patient is currently undergoing radiotherapy.

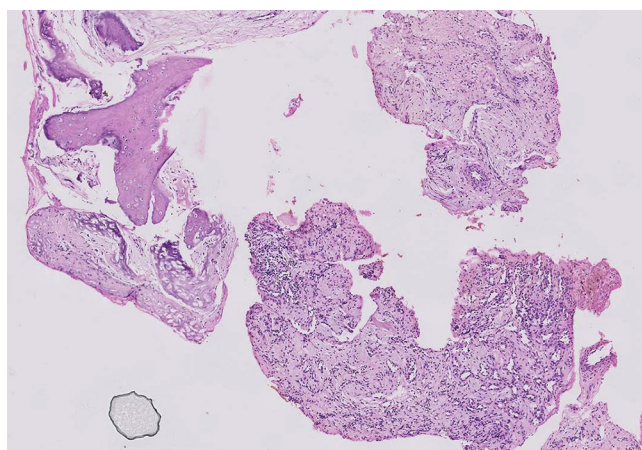


Figure 2. Histopathologic image: intertrabecular fibrotic marrow showing both thin-walled capillary-sized vessels along with few ectatic and thick-walled larger-caliber vessels.

DISCUSSION

PIHs are benign neoplasms of the bone mainly involving the vertebra; those involving the calvarium comprise only 0.2%.¹ The first documented reporting of such a cranial lesion, by Toyne in 1945, described a vascular tumor of the parietal bone.⁷ Calvarial PIHs commonly involve the frontal and parietal bones, but they have also been reported to involve the craniofacial bones.² PIHs located in the skull base are even less frequent, with the majority of reported cases involving the petrous bone.⁶ Other reported cases that involved the skull base bones were 2 cases involving the wings of sphenoid, 1 case involving the body of sphenoid, 4 cases involving the clivus, and 1 case involving the occipital condyle.⁸

We report a case of mixed hemangioma involving the body of sphenoid and occupying the cavity of the sellar proper, thus mimicking an intrasellar mass resulting in bony expansion and localized mass effect.

The pathogenesis of these lesions remains unclear. However, it has been hypothesized that intraosseous hemangiomas can be either congenital or secondary to trauma.⁹ These lesions show a male predominance, with a male-to-female ratio of 3:1. Peak incidence falls into the fourth and fifth decades of life.¹⁰ Our case was hence somewhat atypical as it afflicted a young gentleman 29 years of age.

PIHs are slowly growing expansile lesions, often presenting as an asymptomatic incidental finding on radiographic studies. PIHs of the cranium/calvarium usually do not involve the underlying dura and are slow growing.⁹ However, they may cause localized pain or neurological deficits in lesions with intracranial mass effect, possibly due to stretching or irritation of dura.¹¹ In contrast, the clinical profile of PIHs located at the skull base is less known due to their rare occurrence and scarce reporting in the literature. Our case presented with a short history of headaches with sudden progression and acute unilateral visual loss of only 10 days' duration. Other locations of such lesions will likely cause focal symptoms depending on their specific topography. The risk for spontaneous hemorrhage from any such lesion is unknown.

Radiologically there is marked difference in signal characteristics of vertebral

PIH as opposed to PIHs of the skull. Vertebral PIHs radiographically present as well-circumscribed expansile areas showing bone remodeling in the form of a pathognomic “polka dot” pattern on cross-sectional computed tomography images (thin cuts, bone windows).¹² This characteristic pattern is not seen with PIHs of the skull. MRI features are nonspecific as they greatly depend on the amount of venous flow and fatty content within a lesion, resulting in mottled heterogenous signal noted on T1- and T2-weighted sequences.^{2,13} Larger PIHs of the skull show characteristic features of a delayed contrast blush, which does not occur in smaller lesions and especially those involving the skull base.

Our preoperative diagnosis was that of a nonfunctioning atypical pituitary macroadenoma. Pituitary adenomas on MRI show hypointense signals on T1-weighted sequence with variable signal intensity on T2-weighted sequences. The differentiating features that may present with PIH are marked hyperintense signals on T2-weighted sequences. Both can show a late homogenous contrast enhancement.^{14,15} In our case, MRI features indicated a similar space-occupying lesion of the sella with limited bony expansion but with avid homogenous contrast enhancement.

Classification of vascular bone tumors remains controversial. Due to clinico-radiologic overlapping, histopathology remains the most reliable diagnostic tool. Errani et al¹² reclassified these tumors on the basis of their characteristic clinical, radiologic, histopathologic, and genetic features including atypical variants. PIHs histologically can be of the capillary, cavernous, and mixed type. The cavernous type is most commonly involving the calvarium and skull base.^{2,16} Cavernous PIHs show histologic features of thin-walled blood vessels with intervening collagenous fibers and calcification; in contrast, the capillary type lacks the intervening fibrosis with smaller blood vessels.¹⁷ Our case was of the mixed type, showing features of both cavernous and capillary hemangiomas.

PIHs do not regress spontaneously and hence warrant a definitive treatment. A complete surgical excision, if feasible, provides the patient with a favorable

prognosis because most cases can be cured. Recurrence is rare.² However, in unresectable or residual tumors (as a result of subtotal excision), radiotherapy has been reported as an acceptable option.^{2,3,11} We encountered inadvertent profuse intraoperative bleeding, which limited adequate visibility of the surgical field. Also, the lesion was noted to be in close proximity to the left ICA and cavernous sinus. Hence we did not proceed with complete surgical excision to avoid untoward damage to the aforementioned structures, and achievement of complete hemostasis was focused on. Hence our patient was advised to pursue radiotherapy and is currently undergoing treatment.

CONCLUSION

PIHs of the skull base are benign, rare, and slow-growing tumors and may mimic other common skull base tumors. Hence it should be kept in mind as a differential diagnosis, especially in those with atypical radiologic presentation. They may remain asymptomatic or present with neurological deficits when in close proximity to critical neurological structures. Most often, classical radiologic appearances may not be obvious and definitive diagnosis is often made after histopathologic analysis of surgically resected tissue. Complete surgical excision should be the primary aim of treatment; however, in inoperable or unresectable tumors, radiotherapy can be a viable option.

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