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Case Report

Primary intraosseous cavernous hemangioma of the clivus in a pediatric patient: A case report and review of the literature

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ABSTRACT

Background: Primary intraosseous cavernous hemangiomas (PICH) of the skull are rare, benign, vascular tumors mainly found in adults' calvarium. Affection of the clivus has been reported, with the most frequent clinical presentation being headaches followed by cranial nerve compromise. In the pediatric population, it has only been described once and was treated with surgery and radiation.

Case Description: A 14-year-old previously healthy female presented with episodes of right lingual fasciculations. The investigation of these signs with brain magnetic resonance imaging (MRI) revealed a large clival contrast-enhancing lesion. The patient underwent an endoscopic endonasal biopsy. The histologic findings were consistent with the diagnosis of cavernous hemangioma. She started treatment with propranolol, with serial MRIs showing lesion stability. The last follow-up MRI, 4 years after diagnosis, revealed a mild decrease in the volume of the lesion, and she remains asymptomatic.

Conclusion: We report the first clival PICH in a pediatric patient, managed without surgery and radiotherapy and treated with propranolol. We present neuro-imaging findings at diagnosis and during follow-up and a brief review of the literature on the topic.

Keywords: Clivus, Endoscopic endonasal biopsy, Intraosseous hemangioma, Propranolol

INTRODUCTION

Primary intraosseous cavernous hemangioma (PICH) of the skull is a rare, benign, vascular tumors mainly found in the cranial vault of adult patients. The affection of the skull base is exceedingly rare, with only seven reported cases of PICH affecting the clivus and only one in a pediatric patient. The management of this type of skull base tumor is not well established and differs significantly from those arising in the calvarium. We report a case of a clival PICH in a female adolescent confirmed by biopsy and treated with propranolol. It is the first reported case showing tumor control without surgery or radiotherapy.

CASE DESCRIPTION

History and imaging

A 14-year-old otherwise healthy female patient presented with 3 weeks of recurrent right lingual fasciculations. She described transient episodes triggered by head rotation movements and progressively worsening in duration and frequency.

Magnetic resonance imaging (MRI) of the brain revealed a large lesion of the clivus extending to the prepontine space and further to the right [Figure 1]. The lesion was slightly hypointense with mottled high-intensity areas in T1-weighted imaging, heterogeneously hyperintense in T2-weighted imaging, and showed intense gadolinium enhancement. Computerized tomography (CT) showed an erosive lesion of the clivus, preserving the cortical bone.

Based on the radiographic studies, the differential diagnosis included chordoma, chondrosarcoma, osteosarcoma, intraosseous meningioma, and hemangioma.

Operative details

After discussion in the pediatric oncology multidisciplinary tumor board, it was decided to perform a biopsy to obtain a histological diagnosis and plan the following treatment.

The biopsy was performed through an endoscopic endonasal approach with intraoperative neuronavigation. A thirty-degree endoscope was employed to access the nasal cavity and to reach the sphenoid sinus through the left nostril. Multiple fragments of the clival lesion were collected. The mass was extremely hemorrhagic, leading to significant blood loss during the procedure. Bleeding was controlled with a hemostatic matrix and compression with cotonoides. Closure was performed with posterior absorbable nasal packing.

Surgery and the postoperative period underwent without complications. She was discharged on the 2nd postoperative day, after performing a CT scan that ruled out complications.

During a reassessment in the Pediatric Neurosurgery and Ear, Nose, and Throat outpatient clinic, she did not present any surgical complications or neurological deficits.

Histopathology and postoperative details

Histological analysis showed multiple dilated thin-walled vessels lined by single-layer endothelial cells. The histopathological diagnosis was cavernous hemangioma [Figure 2].

The patient was rediscussed in the pediatric oncology multidisciplinary tumor board, and since the child was asymptomatic, treatment with propranolol was decided. She started treatment with propranolol 10 mg twice a day

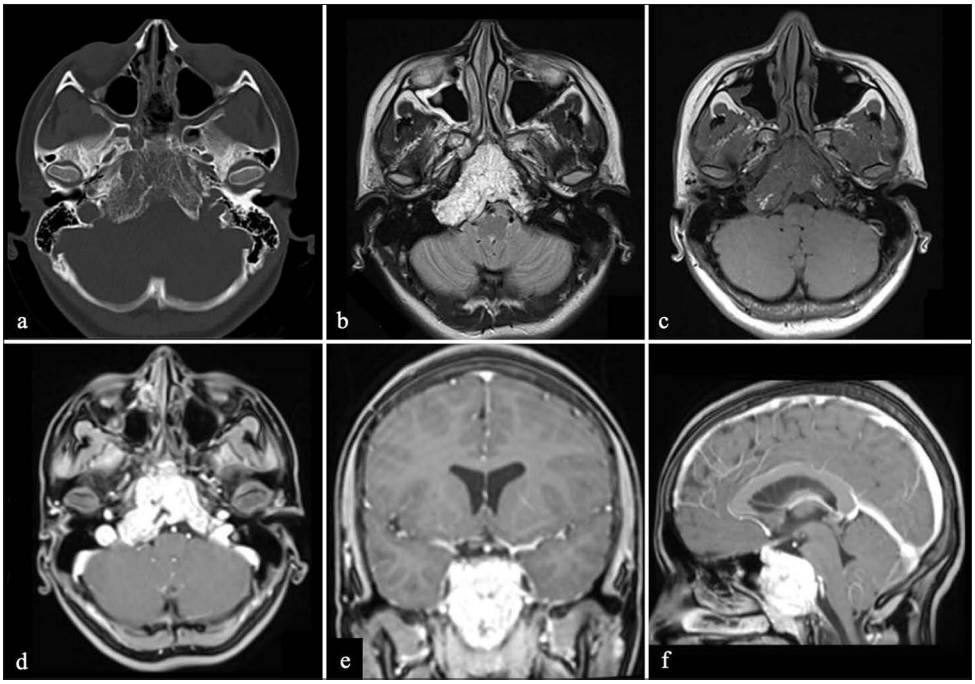

Figure 1: Neuro-imaging findings at diagnosis. (a) Axial bone Computerized Tomography (b) Brain magnetic resonance imaging (MRI): Axial T2 (c) Brain MRI: Axial T1 without contrast (d) Brain MRI: Axial T1 with Gadolinium (e) Brain MRI: Coronal T1 with Gadolinium (f) Brain MRI: Sagittal T1 with Gadolinium.

for a month, with good tolerance. After a month, the dose increased to 20 mg twice a day. She has maintained this therapeutic for 4 years without any adverse effects. The radiological surveillance was done initially with a brain MRI every 3 months. Once lesion stability was observed, follow-up MRIs were performed twice a year and then annually. The last MRI showed a small reduction in the tumor size with less compression of the brainstem [Figure 3].

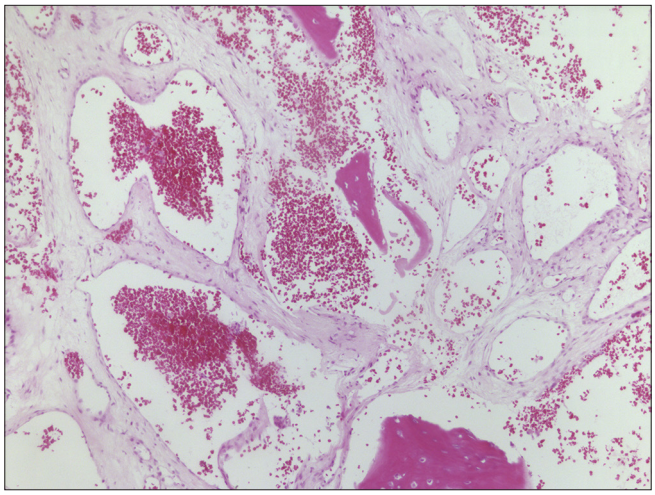

Figure 2: Histopathology: Microphotographs with hematoxylin-eosin staining showing multiple dilated thin-walled vessels lined by single-layer endothelial cells (×100 augmentation).

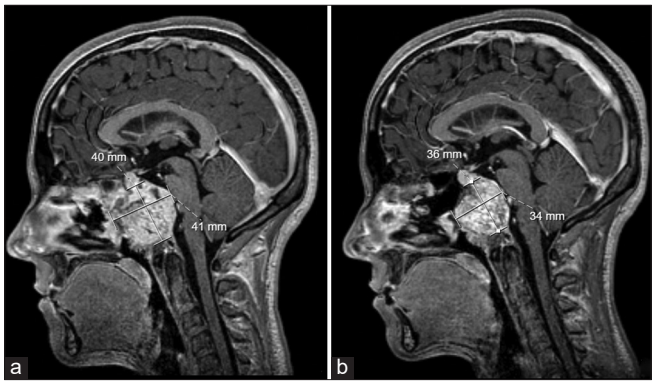

Figure 3: Comparison of the brain magnetic resonance imaging at diagnosis and 4 years later (a) Brain magnetic resonance sagittal T1 with Gadolinium, May 2020 (b) Brain magnetic resonance sagittal T1 with Gadolinium, May 2024.

DISCUSSION

PICHs are rare, benign bone tumors most commonly found in the vertebral column. They represent 0.7% of all osseous neoplasms.[7,20] Less frequently, they are found in the skull, accounting for ~0.2% of all bone tumors and 10% of benign skull tumors,[26,31] and affect predominantly frontal and parietal bones.[13]

PICHs of the skull base are extremely rare tumors, and their management differs as, due to their deep location, they are associated with greater risks of neurovascular deficits, and total resection is difficult to achieve. To our knowledge, only seven clival PICH have been reported in the literature [Table 1].[2,5,10,18,24,26,29] PICHs typically occur in women in their fourth and fifth decades of life.[13] Only one pediatric case of clival PICH has been reported in the literature, in an 11-year-old male treated surgically twice and with radiation after a second relapse.[2]

Clinical presentation

Cranial PICHs can be asymptomatic and diagnosed incidentally. When symptomatic, the clinical presentation depends on the location of the lesion. In the calvarium, they tend to manifest as a growing lump, sometimes associated with throbbing headaches. Less commonly, they can be found in craniomaxillofacial bones and present as painless facial swelling. When involving the orbital bones, patients can present local swelling, neuralgia, nasolacrimal obstruction, proptosis, blepharoptosis, diplopia, or even visual loss.[5,13]

Table 1: Clival PICH reported in the literature.

Author, year	Age	Sex	Diagnosis	Surgical treatment	Complementary treatment
Tashiro *et al.*, 1991	37	Female	Cavernous hemangioma	Surgery	
Vanhoenacker *et al.*, 2011	45	Female	Cavernous hemangioma	Biopsy	
Moravan *et al.*, 2012	62	Male	Capillary hemangioma	Biopsy	
Gologorsky *et al.*, 2013	89	Female	Cavernous hemangioma	Partial resection	
Serrano *et al.*, 2015	62	Female	Cavernous hemangioma	Partial resection	
Campbell *et al.*, 2019	11	Male	Capillary hemangioma	Total resection 1st recurrence: near total resection	2nd recurrence: radiation
Kobayashi *et al.*, 2024	57	Female	Cavernous hemangioma	Partial resection	
Branco *et al.*, 2025 (Present Case)	14	Female	Cavernous hemangioma	Biopsy	Propranolol

PICH: Primary intraosseous cavernous hemangioma

Skull-based affection is very unusual. In 1921, Brandt reported the first case of a hemangioma arising from the petrous bone.[1] Beyond headaches, cranial neuropathies have been associated with skull base lesions [5,24], and there is one report of spontaneous epidural hemorrhage of a petrosal lesion.[8]

Neuro-imaging findings

The earliest description of radiographic characteristics of intraosseous hemangiomas was in 1930.[1] They are typically expansive lesions with a well-circumscribed area of bone rarefaction. The CT is the most helpful imaging technique, showing a sunburst pattern of trabeculations radiating from a common center or a honeycomb appearance. Hemangiomas usually respect the cortical bone layer, as opposed to radiological studies from bone malignancies that are more invasive.[13] MRI imaging is not specific, with heterogeneous signals in both T1- and T2-weighted sequences depending on the amount of venous flow and fatty transformation within the tumor.[5,21,26]

In our case, all these characteristics were present; they were not enough to make the diagnosis, that was why it was essential to confirm it with a biopsy.

Histology

PICHs have a vascular origin and can be divided into three types: cavernous, capillary, or mixed. In contrast to vertebral hemangiomas that are more frequently of the capillary type, most of the calvarial hemangiomas are cavernous. They arise from vessels of the diploic space and are supplied by the branches of the external carotid artery.[9]

Treatment

Indications for PICH surgical treatment include neurological compromise, recurrent headaches, mass effect, cosmetic deformity, or need for definitive diagnosis.[5] Most authors recommend "*en bloc*" resection when possible,[13,20] with a 1 cm wide margin resection of uninvolved bone to prevent recurrence.[7] Because of its vascular origin, hemorrhagic risk is substantial, so embolization before surgery has been described as helpful in preventing excessive bleeding.[14]

Nonetheless, surgical resection of a clival hemangioma can be particularly challenging and associated with higher morbidity due to their tendency to involve neurovascular structures during their growth.[24] In addition, the low incidence of PICH located deeply in the skull base, such as the clivus, makes it difficult to access and predict the clinical evolution of these tumors with or without surgery.[24]

Taking into consideration the risk of a surgical procedure, particularly bleeding, in an asymptomatic child, treatment with propranolol was decided. The patient has been on propranolol for 4 years following the same treatment protocol as for infantile hemangioma (IH). Propranolol has been used to treat IH for more than 15 years,[12] and it has been widely accepted as the first-line treatment with satisfactory results and fewer adverse effects.[22] Its regulatory roles in adipogenesis, angiogenesis, and endothelial cell apoptosis prevent growth and promote the reduction of hemangiomas.[3,6,11,23,30,33] Considering its safety profile,[15] it has been used for other indications such as hepatic hemangiomas and airway IHs.[16,17,34] Its use in PICH has been previously reported to manage back pain in children with vertebral hemangioma.[28] However, to our knowledge, this is the first report of a clival PICH treated with propranolol and exhibiting tumor reduction after 4 years of therapy. More studies are necessary to determine the role of propranolol in the treatment of PICH.

Despite sparse evidence in the literature, radiation has been described as an alternative or adjunct treatment to incompletely resected PICH to prevent tumor growth, but with no clear effect on tumor reduction.[20,25,32] However, there have been some concerns about the long-term side effects of radiation, including malignant transformation, secondary malignancy generation,[4] hypopituitarism, and cranial nerve deficit.[2,5] Considering that these tumors do not spontaneously involute[2] but usually grow slowly, some authors defend that it may be preferable to follow such lesions conservatively rather than subject a patient to radiation. In case of progression, fractionated stereotactic radiation therapy or radiosurgery may be considered.[13,19,27]

CONCLUSION

We report a case of a clival PICH in a pediatric patient. Even though the first treatment option regarding skull PICH is surgical resection, this case illustrates that in deep-sited lesions associated with high surgical risks, medical treatment with propranolol might be an option to stabilize the disease. It also opens the discussion about the use of propranolol for intraosseous hemangiomas, although further data will be needed.

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REFERENCES

1. Bucy PC, Capp CS. Primary hemangioma of bone with special reference to roentgenologic diagnosis. AJR Am JRoentgenol

1930;23:1-33.
2. Campbell JI, Mural M, Rubino F, Lopez ES, Cervio A, Olvi L. Clivus hemangioma in a pediatric patient: Case report. World Neurosurg 2019;130:512-5.
3. England RW, Hardy KL, Kitajewski AM, Wong A, Kitajewski JK, Shawber CJ, *et al*. Propranolol promotes accelerated and dysregulated adipogenesis in hemangioma stem cells. Ann Plast Surg 2014;73 Suppl 1:S119-24.
4. Fredrickson JM, Haight JS, Noyek AM. Radiation-induced carcinoma in a haemangioma. Otolaryngol HeadNeck Surg 1979;87:584-6.
5. Gologorsky Y, Shrivastava RK, Panov F, Mascitelli J, Signore AD, Govindaraj S, *et al*. Primary intraosseous cavernous hemangioma of the clivus: Case report and review of the literature. J Neurol Surg Rep 2013;74:17-22.
6. Hansen T, Kunkel M, Katenkamp D, Eletr S, Wagner W. Hemangioma of the mandible: Case report with special emphasis on bone degradation. Oral Maxillofac Surg 2009;13:239-42.
7. Hook SR, Font RL, McCrary JA, Harper RL. Intraosseous capillary hemangioma of the frontal bone. Am J Ophthalmol 1987;103:824-7.
8. Kessler LA, Lubic LG, Koskoff YD. Epidural hemorrhage secondary to cavernous hemangioma of the petrous portion of the temporal bone. J Neurosurg 1957;14:329-31.
9. Khanam H, Lipper MH, Wolff CL, Lopes MB. Calvarial hemangiomas: Report of two cases and review of the literature. Surg Neurol 2001;55:63-7.
10. Kobayashi Y, Satoh S, Kishida Y, Goto H, Fujimori D, Onuki A, *et al*. Primary intraosseous cavernous hemangioma of the clivus: A case report and literature review. Surg Neurol Int 2024;15:177.
11. Kotrashetti SM, Urolagin SB, Kale TP, Baliga SD. Central hemangioma - A case report and review of literature. Asian J Oral Maxillofac Surg 2011;23:46-9.
12. Léauté-Labrèze C, Dumas de la Roque E, Hubiche T, Boralevi F, Thambo JB, Taïeb A. Propranolol for severe hemangiomas of infancy. N Engl J Med 2008;358:2649-51.
13. Liu JK, Burger PC, Harnsberger HR, Couldwell WT. Primary intraosseous skull base cavernous hemangioma: Case report. Skull Base 2003;13:219-28.
14. Lobato RD, Lamas E, Amor T, Rivas JJ. Primary calvarial hemangioma: Angiographic study. Surg Neurol 1978;10:389-94.
15. Love JN, Sikka N. Are 1-2 tablets dangerous? Beta-blocker exposure in toddlers. J Emerg Med 2004;26:309-14.
16. Maturo S, Hartnick C. Initial experience using propranolol as the sole treatment for infantile airway hemangiomas. Int J Pediatr Otorhinolaryngol 2010;74:323-5.
17. Mhanna A, Franklin WH, Mancini AJ. Hepatic infantile hemangiomas treated with oral propranolol--a case series. Pediatr Dermatol 2011;28:39-45.
18. Moravan MJ, Petraglia AL, Almast J, Yeaney GA, Miller MC, Edward Vates G. Intraosseous hemangioma of the clivus: A case report and review of the literature. J Neurosurg Sci 2012;56:255-9.
19. Nakamura N, Shin M, Tago M, Terahara A, Kurita H, Nakagawa K, *et al*. Gamma knife radiosurgery for cavernous hemangiomas in the cavernous sinus. Report of three cases. J Neurosurg 2002;97:477-80.
20. Peterson DL, Murk SE, Story JL. Multifocal cavernous hemangioma of the skull: Report of a case and review of the literature. Neurosurgery 1992;30:778-81.
21. Ross JS, Masaryk TJ, Modic MT, Carter JR, Mapstone T, Dengel FH. Vertebral hemangioma: MR imaging. Radiology 1987;165:165-9.
22. Sánchez-Carpintero I, Ruiz-Rodriguez R, López-Gutiérrez JC. Propranolol in the treatment of infantile hemangioma: Clinical effectiveness, risks, and recommendations. Actas Dermosifiliogr 2011;102:766-79.
23. Sepulveda I, Spencer ML, Platin E, Trujillo I, Novoa S, Ulloa D. Intraosseous hemangioma of the mandible: Case report and review of the literature. Int J Odontostomatol 2013;7:395-400.
24. Serrano L, Archavlis E, Januschek E, Ulrich PT. 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. Surg Neurol Int 2015;6:S117-23.
25. Sweet C, Silbergleit R, Mehta B. Primary intraosseous hemangioma of the orbit: CT and MR appearance. AJNR Am J Neuroradiol 1997;18:379-81.
26. Tashiro T, Inoue Y, Nemoto Y, Shakudo M, Mochizuki K, Katsuyama J, *et al*. Cavernous hemangioma of the clivus: Case report and review of the literature. Am J Neuroradiol 1991;12:1193-4.
27. Tsao MN, Schwartz ML, Bernstein M, Halliday WC, Lightstone AW, Hamilton MG, *et al*. Capillary hemangioma of the cavernous sinus. Report of two cases. J Neurosurg 2003;98:169-74.
28. Uzunaslan D, Saygin C, Gungor S, Hasiloglu Z, Ozdemir N, Celkan T. Novel use of propranolol for management of pain in children with vertebral hemangioma: Report of two cases. Childs Nerv Syst 2013;29:855-60.
29. Vanhoenacker FM, Praeter GD, Kools D, Voormolen M, Parizel PM. Unusual lesion of the clivus. Skelet Radiol 2011;40:223-4.
30. Wong A, Hardy KL, Kitajewski AM, Shawber CJ, Kitajewski JK, Wu JK. Propranolol accelerates adipogenesis in hemangioma stem cells and causes apoptosis of hemangioma endothelial cells. Plast Reconstr Surg 2012;130:1012-21.
31. Wyke BD. Primary hemangioma of the skull; a rare cranial tumor; review of the literature and report of a case, with special reference to the roentgenographic appearances. Am J Roentgenol Radium Ther 1949;61:302-16.
32. Yang Y, Guan J, Ma W, Li Y, Xing B, Ren Z, *et al*. Primary intraosseous cavernous hemangioma in the skull. Medicine 2016;95:e3069.
33. Yao TH, Pataer P, Regmi KP, Gu XW, Li QY, Du JT, *et al*. Propranolol induces hemangioma endothelial cell apoptosis via a p53BAX mediated pathway. Mol Med Rep 2018;18:684-94.
34. Zegpi-Trueba MS, Abarzúa-Araya A, Silva-Valenzuela S, Navarrete-Dechent C, Uribe-González P, Nicklas-Díaz C. Oral propranolol for treating infantile hemangiomas: A case series of 57 patients. Actas Dermosifiliogr 2012;103:708-17.

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