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Case Report

Unusual case of hemangioblastoma of the cerebellopontine angle

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Abstract

Background: Hemangioblastomas are the most common primary tumor of the posterior fossa. There are few cases of hemangioblastoma of the cerebellopontine angle (CPA). When present in this location, hemangioblastoma presents a diagnostic challenge as its imaging findings closely resemble those of vestibular schwannoma (VS), which is much more common in the CPA.

Case Description: We report the case of a 42-year-old man presenting with vertigo and diplopia found to have a CPA tumor with imaging resembling VS. He underwent retrosigmoidal resection of his tumor, which was found to be a hemangioblastoma.

Conclusion: Hemangioblastoma, though rare in the CPA, should be considered in the differential diagnosis of CPA tumors.

Key Words: Cerebellopontine angle, CPA, hemangioblastoma

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BACKGROUND

Hemangioblastomas are well-differentiated, vascular, benign tumors primarily located in the posterior cranial fossa. They are the most common primary intra-axial tumor located in the posterior fossa in adults comprising ~2.5% of all intracranial tumors.[17] Very rarely, hemangioblastoma may occur in the cerebellopontine angle (CPA).[1,5-7,9,10,12,14-18,20,21] Hemangioblastomas of the CPA represents a diagnostic conundrum due to the extra-axial appearance of these lesions as well as the imaging similarity with vestibular schwannoma (VS), which is much more common in this region. Distinction between these diagnoses is important due to the significant blood loss that may result from internal debulking of hemangioblastoma.

CASE DESCRIPTION

A 42-year-old man presented with vertigo and progressive diplopia on extreme lateral right gaze. He denied tinnitus, hearing loss, headaches, and was found to be grossly neurologically intact. Magnetic resonance (MR) of the brain revealed a lesion demonstrating T1 hypointensity and T2 hyperintensity with peripheral and septal enhancement in the right CPA [Figure 1]. Whether this lesion was intra-axial or extra-axial was not clear. After discussion with the patient regarding options for management, the decision was made to proceed with surgical resection.

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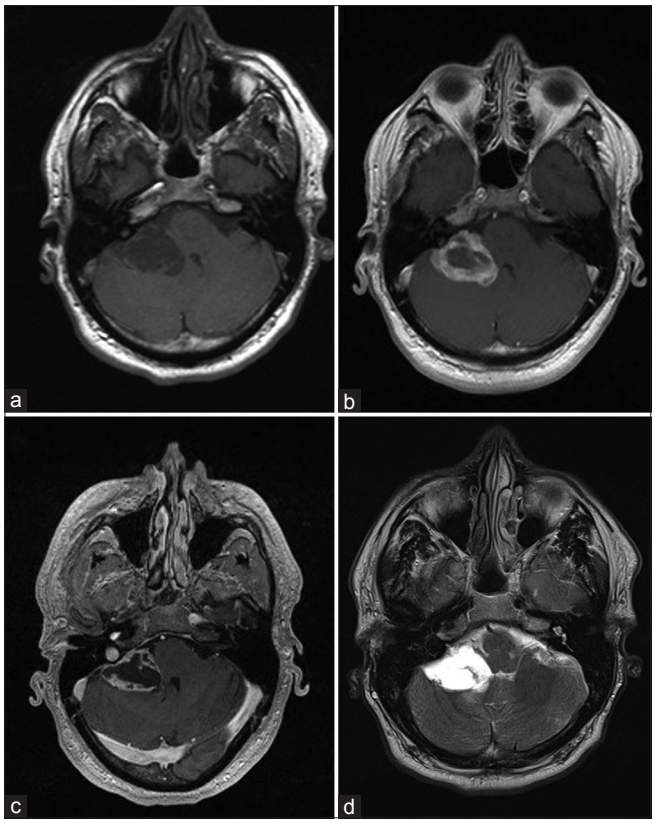

Figure 1: (a) axial T1 weighted, (b) axial FLAIR, (c) axial post-gadolinium and (d) axial T2 weighted MR images of the CP angle tumor. The lesion is hypointense on T1WI, hyperintense on FLAIR and T2WI, and display enhancement along its periphery and septations. Mass effect on the 4th ventricle is appreciated

The patient was booked for a retrosigmoid craniotomy for removal of the CPA tumor. A right frontal external ventricular drain (EVD) was placed at the beginning of the case. During craniotomy, retraction of the cerebellum revealed the tumor to be extra-axial [Figure 2]. The gross appearance of the tumor was suggestive of a highly vascular lesion, and so intraoperatively, a decision was made to attempt an *en-bloc* resection. The tumor was circumferentially dissected from the cranial nerve VII/VIII complex as well as from the choroid plexus at the foramen of Luschka. Following this, vascular feeders of the tumor were coagulated and the tumor was excised. Frozen sections returned consistent with low-grade tumor. Final pathology showed epithelioid foamy stromal cells with abundant thin-walled capillary-like vessels. The stromal cells were positive for inhibin, vimentin, and S-100. Reticulin and CD34 staining outlined the dense reticular capillary network of the tumor, but the typical dense pericellular network found in schwannoma was not present. No expression of epithelial membrane antigen (EMA) or progesterone receptor was observed. MIB1 index was <5%. The final diagnosis was reticular variant hemangioblastoma [Figure 3].

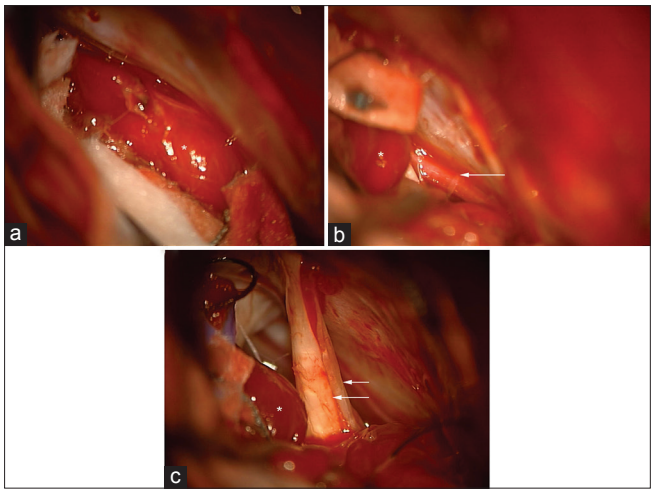

Figure 2: Intraoperative photographs of the CPA tumor, marked with asterisk (*). (a) Microscopic appearance of the tumor. (b) Microscopic view of the tumor with adjacent anterior inferior cerebellar artery (AICA) (arrow). (c) Microscopic view of CPA tumor with adjacent cranial nerve VII and VIII complex (arrows)

The patient developed a cerebrospinal leak (CSF) leak in his postoperative course, which required re-operation for revision duraplasty. Otherwise his postoperative course was uneventful and he was discharged home with no neurological impairment.

DISCUSSION

Hemangioblastoma is most commonly diagnosed between the third and fifth decades of life and exhibit a male preponderance. They are formed of the remnant mesoderm of the brain. Approximately 25% of cases are associated with von Hippel Lindau (vHL) disease, with 60–84% of vHL patients having hemangioblastoma.[21] vHL is an autosomal dominant syndrome resulting from mutations to the *VHL* gene on chromosome 3 causing hemangioblastomas in a variety of locations such as pheochromocytoma, epididymal tumors, and renal cell carcinoma. Due to the high incidence of heamngioblastoma in vHL, it has been suggested that even patients presenting with a single central nervous system hemangioblastoma should be considered for vHL testing.[20]

Typically, hemangioblastomas are found within the cerebellum. Hemangioblastoma involving the CPA is rare.[1,5-7,9,10,12,15-18,20,21] Usually, hemangioblastomas originate in the subcortical cerebellar parenchyma, with intra-axial extension leading to the typical presentation of cerebellar dysfunction. With larger lesions, obstructive hydrocephalus may ensue from fourth ventricular obstruction. Hemangioblastoma often remain asymptomatic for long periods due to their slow growth. The symptomatology of hemangioblastoma depends on the size and location of the lesion. As with any other

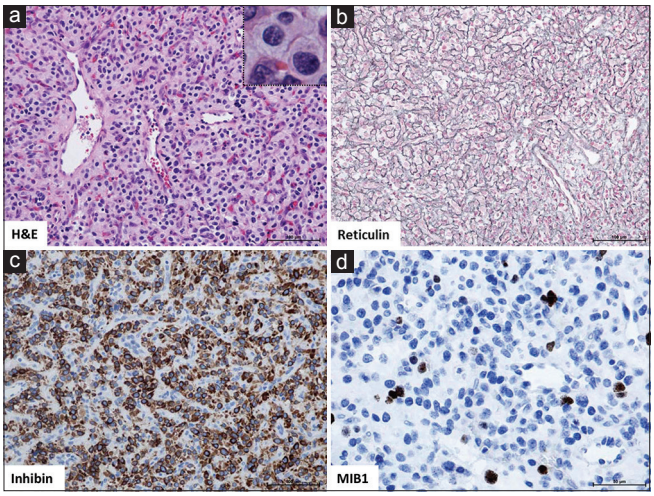

Figure 3: (a) H and E: Tumor composed of epithelioid, slightly foamy stromal cells with mild degree of degenerative atypia and predominant capillary proliferation (reticular variant). No evidence of increased mitotic activity or extramedullary hematopoiesis. ×20 magnification with ×40 inset; (b) Reticulin, highlights reticular fibres of the dense capillary network of this reticular hemangioblastoma, ×20; (c) Inhibin, demonstrates cytoplasmic expression in stromal tumor cells, ×20; (d) MIB1, Proliferation index <5%, ×40

lesion arising from this location, hemangioblastomas of the CPA may present with symptoms secondary to compression of the cranial nerves. In this case, the patient presented with vertigo, either from compression of cranial nerve VIII or direct cerebellar mass effect. The coincident diplopia may have been from compression of cranial nerve VI.

Macroscopically, hemangioblastomas can be divided into solid and cystic subtypes. The vast majority, 60–90% of hemangioblastomas are cystic. The typical gross appearance of hemangioblastoma is a reddish or yellow mural nodule with a cystic component. Histologically, hemangioblastoma can de divided into cellular and reticular subtypes. The cellular variant consists of foamy stromal cells clustered in sheets about tangles of capillaries, while in the reticular subtype the stromal cells are evenly distributed around a dense capillary network. Immunohistochemical staining for inhibin, vimentin, and S100 is typically positive in hemangioblastomas.

The differential diagnosis of tumors within the CPA is broad, but the majority of these tumors are VS and meningioma.[3,4,8,13] VSs comprise 70–90% of CPA lesions and meningioma another 5–15%. Another 6% of CPA lesions are epidermoid cysts, and around 1% are lipomas. Other lesions within the CPA are rare but have been observed. Of these, hemangioblastoma presents a particular diagnostic challenge due to the similarities between hemangioblastoma and VS on MRI. Both appear as hypointense on T1-weighted images (WI) and hyperintense on T2WI, and both have strong enhancement with gadolinium. Cystic appearance, involvement of the internal auditory canal, presence of dilated feeding arteries, or flow void due to the hypervascularity of the lesion may help as differentiating factors.

Management of hemangioblastoma is guided by presentation. Serial MRIs can be used to follow asymptomatic lesions. The hypervascularity of hemangioblastoma makes it a surgically challenging lesion. These lesions are typically well circumscribed, and if possible, an *en-bloc* resection is preferred. Internal debulking of the lesion as in VS resection could lead to disastrous results due to its vascularity. While radiotherapy has been investigated as a treatment,[11,12] surgical resection remains the standard of care due to lower recurrence rates. Small tumors less than 3 cm and cystic features are associated with better outcomes.[6] Preoperative strategies such as embolization[18,19] and radiosurgery[10] have been reported in order to minimize bleeding. We obtained good intraoperative exposure for complete *en-bloc* resection using the retrosigmoidal approach with no postoperative neurological deficit. Follow-up imaging at 6 months and 12–24 months is recommended due to the high risk of recurrence in these patients,[2] especially where there is concern of vHL.[6]

CONCLUSION

Hemangioblastoma should be included in the differential diagnosis of CPA tumors. While uncommon, the potential for misdiagnosis and attempt at inappropriate surgical options could be potentially disastrous due to the potential for catastrophic bleeding from these tumors. The retrosigmoidal approach is a safe and effective hearing-preserving approach for resection of these tumors.

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Conflicts of interest
There are no conflicts of interest.

REFERENCES

1. Amano T, Tokunaga S, Shono T, Mizoguchi M, Matsumoto K, Yoshida F, *et al*. Cerebellar hemangioblastoma manifesting as hearing disturbance. Neurol Med Chir (Tokyo) 2009;49:418-20.
2. Bamps S, Calenbergh FV, Vleeschouwer SD, Loon JV, Sciot R, Legius E, *et al*. What the neurosurgeon should know about hemangioblastoma, both sporadic and in Von Hippel-Lindau disease: A literature review. Surg Neurol Int 2013;4:145.
3. Bonneville F, Sarrazin JL, Marsot-Dupuch K, Iffenecker C, Cordoliani YS, Doyon D, *et al*. Unusual Lesions of the Cerebellopontine Angle: A Segmental Approach. Radiographics 2001;21:419-38.
4. Brackmann DE, Bartels LJ. Rare tumors of the cerebellopontine angle. Otolaryngol Head Neck Surg 1980;88:555-9.
5. Bush ML, Pritchett C, Packer M, Ray-Chaudhury A, Jacob A. Hemangioblastoma of the cerebellopontine angle. Arch Otolaryngol Head Neck Surg 2010;136:734-8.
6. Cheng J, Liu W, Zhang S, Lei D, Hui X. Clinical features and surgical outcomes

in patients with cerebellopontine angle hemangioblastomas: A retrospective series of 23 cases. World Neurosurg 2017;103:248-56.
7. Dow GR, Sim DW, O'Sullivan MG. Excision of large solid haemangioblastomas of the cerebellopontine angle by a skull base approach. Br J Neurosurg 2002;16:168-71.
8. Friedmann DR, Grobelny B, Golfinos JG, Roland JT Jr. Nonschwannoma tumors of the cerebellopontine angle. Otolaryngol Clin North Am 2015;48:461-75.
9. Grahovac G. Solid hemangioblastoma of vestibular nerve mimicking vestibular schwannoma. Neurol Sci 2015;36:1537-9.
10. Kamitani H, Hirano N, Takigawa H, Yokota M, Miyata H, Ohama E, *et al*. Attenuation of vascularity by preoperative radiosurgery facilitates total removal of a hypervascular hemangioblastoma at the cerebellopontine angle: Case report. Surg Neurol 2004;62:238-44.
11. Kano H, Niranjan A, Mongia S, Kondziolka D, Flickinger JC, Lunsford LD. The role of stereotactic radiosurgery for intracranial hemangioblastomas. Neurosurgery 2008;63:443-51.
12. Laviv Y, Thomas A, Kasper EM. Hypervascular Lesions of the Cerebellopontine Angle: The Relevance of Angiography as a Diagnostic and Therapeutic Tool and the Role of Stereotactic Radiosurgery in Management. A Comprehensive Review. World Neurosurg 2017;100:100-17.
13. Moffat DA, Ballagh RH. Rare tumours of the cerebellopontine angle. Clin Oncol (R Coll Radiol) 1995;7:28-41.
14. Moon BH, Park SK, Han YM. Large solid hemangioblastoma in the cerebellopontine angle: Complete resection using the transcondylar fossa approach. Brain Tumor Res Treat 2014;2:128-31.
15. Nair BR, Joseph V, Chacko G, Keshava SN. Giant solid hemangioblastoma of the cerebellopontine angle: A technically challenging case. Neurol India 2014;62:228-9.
16. Qiao PF, Niu GM, Han XD. Hemangioblastoma originating from the right cerebellopontine angle. Neurosciences (Riyadh) 2011;16:372-4.
17. Rachinger J, Buslei R, Prell J, Strauss C. Solid haemangioblastomas of the CNS: A review of 17 consecutive cases. Neurosurg Rev 2009;32:37-48.
18. Sakamoto N, Ishikawa E, Nakai Y, Akutsu H, Yamamoto T, Nakai K, *et al*. Preoperative Endovascular Embolization for Hemangioblastoma in the Posterior Fossa. Neurol Med Chir (Tokyo) 2012;52:878-84.
19. Takeuchi S, Tanaka R, Fujii Y, Abe H, Ito Y. Surgical treatment of hemangioblastomas with presurgical endovascular embolization. Neurol Med Chir (Tokyo) 2001;41:246-51.
20. Wanebo JE, Lonser RR, Glenn GM, Oldfield EH. The natural history of hemangioblastomas of the central nervous system in patients with von Hippel- Lindau disease. J Neurosurg 2003;98:82-94.
21. Young S, Richardson AE. Solid haemangioblastomas of the posterior fossa: Radiological features and results of surgery. J Neurol Neurosurg Psychiatry 1987;50:155-8.