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Noncystic cerebellopontine angle hemangioblastoma: A case of an atypical location

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ABSTRACT

INTRODUCTION: Extra-axial cerebellopontine angle (CPA) hemangioblastoma is a rare condition in which the correct differential diagnosis from other CPA lesions can affect the best treatment choice. These are benign tumors that are highly vascularized and mostly present in the cystic form. About twenty-six cases have been reported in the literature with this same location and with a noncystic aspect.

PRESENTATION OF CASE: We report a case of a 63-year-old male with a complaint of progressive headache associated with imbalance and difficulty walking. Neurological examination showed discreet facial paresis, left dysmetria and mild gait ataxia. Magnetic resonance imaging (MRI) showed a solid mass with isointensity on T1-weighted sequences; hypointensity and a heterogeneous appearance on T2-weighted sequences; and intense homogeneous contrast enhancement located in the left CPA region extending superiorly to the tentorial notch. The first diagnosis was meningioma, but during the microsurgical suboccipital retrosigmoid approach, it was observed that the lesion was extremely bloody with several vessels on its surface. We achieved gross total resection, and the pathology confirmed hemangioblastoma.

DISCUSSION: Although it is rare, hemangioblastoma should be one of the differential diagnoses when dealing with CPA solid lesions with high contrast enhancement and heterogeneity on T2-weighted MRI. Analysis of the radiological characteristics allows a greater chance of confirmation and is one of the main tools for surgical planning.

CONCLUSION: Correct preoperative evaluation and the possibility that hemangioblastoma may arise from the CPA can avoid trans-operative risks mainly related to bleeding and can improve results.

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1. Introduction

Hemangioblastomas are benign and highly vascularized, are mostly present in the cystic form, and correspond histologically to grade 1 tumors according to the WHO's latest classification [1]. Macroscopically, they appear well delimited and are formed by reddish nodules that are quite vascularized or yellowish due to the high lipid content of tumor cells. Histologically, they are formed by stromal cells that have variable-sized nuclei, in addition to highly

vascularized capillaries [1]. They are more common in children and young adults, with a peak incidence between 35 and 45 years as well as a male predominance (M:F= 2:1). They may be solitary and sporadic, but in nearly 25% of cases, they are associated with von Hippel-Lindau disease (VHLd), an autosomal dominant syndrome with variable gene expression and complete penetrance [2]. The most affected sites in descending order are the cerebellum, brainstem, spinal cord and, rarely, the supratentorial space [1].

The differential diagnosis of lesions located in the cerebellopontine angle is important since it can affect the treatment choice. Although 80–95% of cerebellopontine angle (CPA) lesions are represented by schwannomas and meningiomas, other differential diagnoses should be suspected, such as hemangioblastomas [3]. Thus, we report a rare case of a noncystic hemangioblastoma located in the CPA operated on in a public hospital, and we reviewed other differential diagnoses that can occur in the same region. This case is reported in line with the SCARE 2018 criteria [4].

Abbreviations: CPA, cerebellopontine angle; CT, computed tomography; MRI, magnetic resonance imaging; VHLd, von Hippel-Lindau disease.

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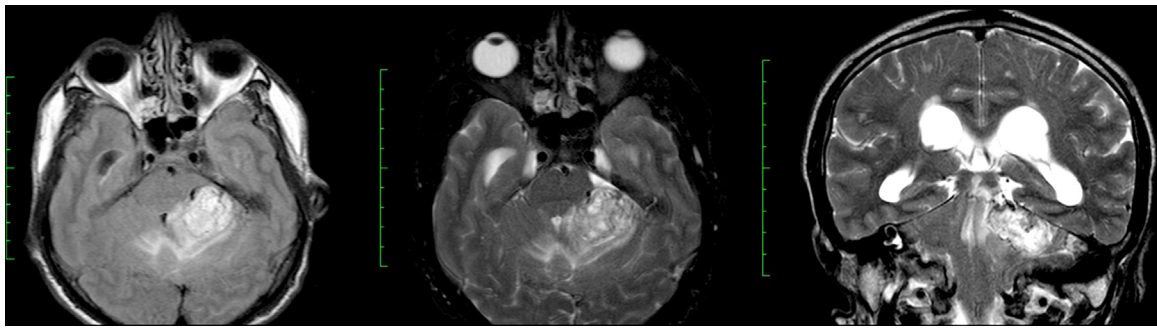


Fig. 1. Brain MRI showing a lesion located in the left CPA. The first image is an axial Flair image that shows a lesion with a hypointense signal in relation to brain tissue and an edema signal in the cerebellar parenchyma. The second and third images, axial and coronal T2-weighted images, show heterogeneous signal intensity.

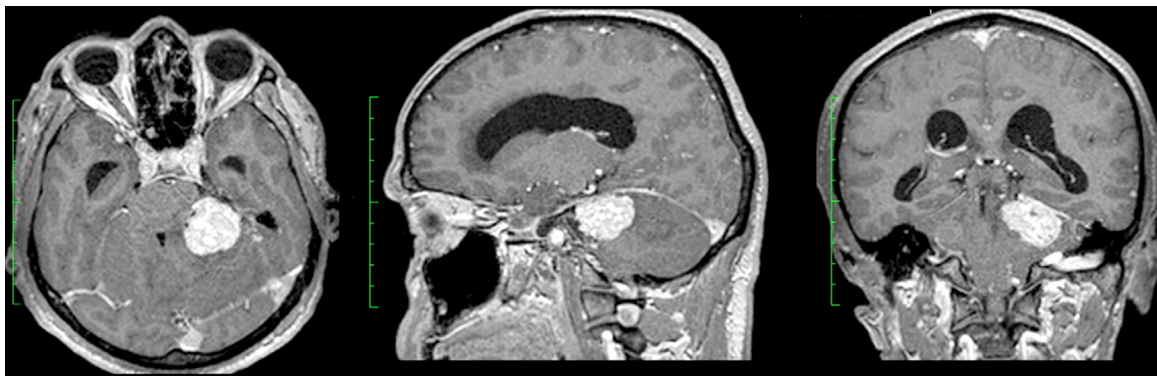


Fig. 2. Axial, sagittal and coronal gadolinium-enhanced T1-weighted MRI showing a CPA lesion with homogeneous contrast enhancement very close to the tentorial notch.

2. Presentation of case

We report a case of a white 63-year-old male, farmworker, who sought the emergency unit of the Emergency Hospital of Sergipe with a complaint of headache that had progressed for 2 months and was associated with imbalance and difficulty walking. The past medical history was negative, without relevant information and with no family history of neurological disorders. Upon admission, a neurological examination was performed, and the patient was lucid and oriented with discreet facial paresis (House-Brackman grade 1), no changes in muscle strength, left dysmetria (finger-to-nose test) and mild gait ataxia. Brain computed tomography (CT) showed a small isodense CPA lesion without calcification. The patient underwent contrast-enhanced magnetic resonance imaging (MRI), which showed isointensity on T1-weighted sequences; hypointensity and heterogeneity on T2-weighted sequences; and intense homogeneous contrast enhancement located in the left CPA region and extending superiorly to the tentorial notch as well as medially displacing the superior cerebellar peduncle and dorsal portion of the midbrain. The larger dimensions were $2.6 \times 2.7 \times 2.4$ cm (anterior $\times$ lateral $\times$ superior) (Figs. 1 and 2). Due to the presentation of a much higher frequency, the first diagnosis for this case was a meningotheelial lesion or a schwannoma, but brain CT did not show any sign of internal acoustic remodeling, reducing the chances of this latter hypothesis. Because of his neurological complaints, surgical treatment was indicated.

Five days after hospital admission and preoperative clinical and anesthetic risk evaluation, the patient was operated on. Under general anesthesia, the patient was placed in a supine position; his head was fixed with Mayfield 3-point fixation; and his head was lateralized to the right, keeping the mastoid region elevated. The senior neurosurgeon, A.M.P.O. with more than 8 years of experience in brain tumor surgery, made a retro-auricular linear incision of 10 cm followed by a classic suboccipital retrosigmoid craniotomy.

During exposure and dissection of the CPA, it was observed that the lesion was extremely bloody with several vessels on its surface. We started the procedure by coagulating as much of the tumor as possible to reduce arterial irrigation. Then, we performed debulking followed by circumferential dissection and gross total resection. All the cranial nerves were isolated, identified and kept intact after complete tumor resection, which practically ruled out the possibility of schwannoma.

The patient was referred to the intensive care unit in the immediate postoperative period, and the patient evolved with worsening of the left peripheral facial palsy and previous cerebellar syndrome. After five days, the patient was discharged from the hospital and was referred for rehabilitation with physiotherapy. In outpatient follow-up after 4 weeks, the patient showed improvement in terms of the cerebellar syndrome but maintained slight facial palsy. The pathology report revealed a hemangioblastoma (Fig. 3). Because there were no clinical or radiological findings of VHLd, we classified this patient as having the sporadic type. In the long-term follow-up, MRI examinations 6 months and 18 months after surgery (Fig. 4) showed gross total resection and no signs of recurrence. Despite mild neurological deficits, the patient believes that his surgery was necessary and that he is currently able to perform most of his daily activities.

3. Discussion

The cerebellopontine cistern contains nerves, arteries and some embryological remnants, and each of these structures has the potential to be the source of tumor growth [5]. A total of 75% of hemangioblastomas are classified as sporadic, while the remaining cases are associated with VHLd; moreover, when located in the CPA, they tend to be well delimited in the cystic form or to have a solid appearance. When cystic, the exudative liquid portion appears hypointense on T1-weighted sequences and hyperintense

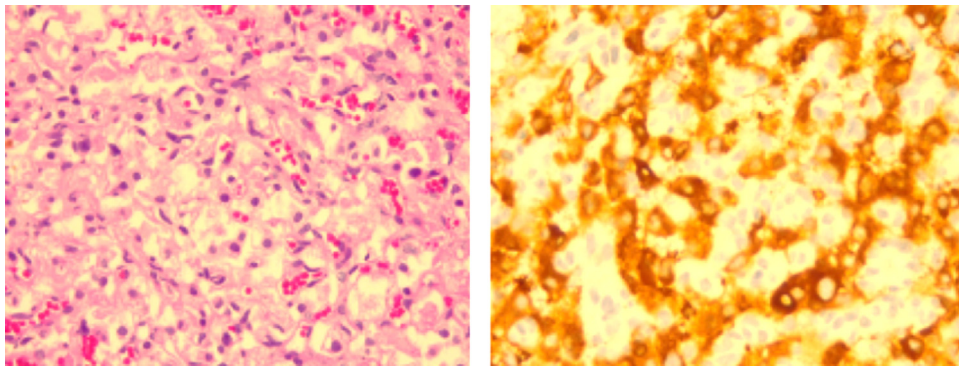


Fig. 3. A morphological finding of proliferation of variable sized, closely packed, thin walled vessels and large neoplastic stromal cells with pink to clear foamy cytoplasm with fine vacuoles and strong inhibin expression, indicates the diagnosis of hemangioblastoma. In addition, the negativity to anti epithelial membrane antigen (EMA) and progesterone receptors excluded the possibility of meningioma.

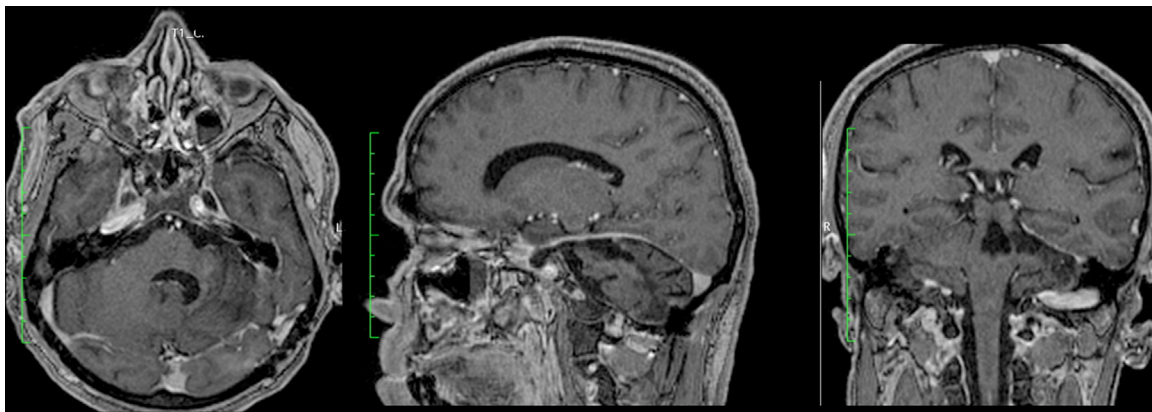


Fig. 4. Axial, sagittal and coronal gadolinium-enhanced T1-weighted MRI 18 months after surgical treatment that do not show evidence of residual lesion or recurrence.

on T2-weighted MRI, with an intense, contrast-enhancing solid component after gadolinium injection [6]. Due to the high vascularity of hemangioblastomas, high-flow vessels as well as flow voids are commonly observed at the edge of the tumor. Preoperative embolization when available and when anatomically favorable can be performed to reduce perioperative bleeding and morbidity [6,7].

As the CPA is an unusual location for hemangioblastoma, reports in the scientific literature are scarce. Until 2014, there were only 11 cases reported in the literature, the majority (eight) with exclusively solid radiological findings. One of them was diagnosed in a patient with VHLd, 6 cases were classified as sporadic, and in 4 reports, it was not established whether the case was sporadic or related to VHLd [8–15]. Cheng et al. [2], in 2017, reported a case series of 23 patients with hemangioblastoma in the CPA in which 8 (35%) patients had VHLd. All patients underwent surgical treatment, with the retrosigmoid approach chosen in 17 patients. Postoperative morbidity was much higher in cystic tumors [2], although most neurological deficits improved during the follow-up (78.3%). Four (17.4%) patients had local recurrence during the follow-up, which was correlated with the younger age and the presence of VHLd, reassuring that radiological follow-up is necessary for the early detection of recurrence mainly in VHLd patients [2]. Similarly, our case showed the most common form of this pathology in the CPA with sporadic, noncystic hemangioblastoma and neurological improvement during the follow-up.

Considering the range of lesions that can emerge in the CPA, neuroimaging knowledge is crucial since lesions located in this region have clinically nonspecific characteristics and since the resulting symptoms do not match the nature of the tumor itself, originating

from the nerves and structures involved in the lesion [2]. The first eligible imaging test is brain CT to analyze the density of soft tissue structures and to analyze bone and signs of calcification, while MRI is fundamental for the visualization of soft tissue structures, including cranial nerves and arterial vessel individualization.

The most frequent tumors located in the CPA region are acoustic schwannomas and meningiomas, but other types of lesions, such as arachnoid cysts, aneurysms, metastases, inflammatory/infectious meningeal lesions, epidermoid cysts, dermoid cysts and lipomas, are possible [5]. Other tumors also have the ability to reach the CPA from the skull base or petrous bone, such as cholesterol granuloma, paraganglioma, chondromatosis, chordoma, endolymphatic sac tumors and pituitary adenoma [5]. A CPA mass may arise secondary to an exophytic brainstem tumor or ventricular tumors, such as lymphoma, ependymoma, medulloblastoma and dysembryoplastic neuroepithelial tumor, in addition to the hemangioblastoma itself [5].

4. Conclusion

We report a case of a sporadic and noncystic hemangioblastoma of the CPA. Although it is rare, it is important for neurosurgeons know that it should be one of the differential diagnoses when dealing with expansive lesions of the CPA. Analysis of the radiological characteristics allows a greater chance of confirmation and is one of the main tools for surgical planning.

Conflicts of interest

No conflicts of interests have been declared by the authors.

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Ethical approval

This article does not include research involving patients, so it does not require ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

Conception and design: Arthur M.P. Oliveira; Acquisition of data: Jose M.B. Santana, Barbara P.F. Neto; Analysis and interpretation of the data: all authors; Drafting the article: Arthur M.P. Oliveira, Jorge D.S. Lapa, Thais C.S. Melo; Critically revising the article: Arthur M.P. Oliveira; Approved the final version of the manuscript on behalf of all authors: Arthur M.P. Oliveira; Study supervision: Arthur M.P. Oliveira.

Registration of research studies

As our case report isn't first in man (i.e. the first time a new device or surgical technique is performed) we didn't submit our work to Research Registry UIN.

Guarantor

Arthur Maynart Pereira Oliveira.

Provenance and peer review

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