



## Case Report

# Extra-axial cerebellopontine angle cavernoma: A case report and review of literature

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## ABSTRACT

**Background:** Extra-axial cavernomas at the cerebellopontine angle (CPA) are rare clinical entity that can radiologically mimic several lesions encountered at this location.

**Case Description:** A 36-year-old female patient referred to our emergency service with acute decreased level of consciousness and vomiting. Neurological examination showed Glasgow Coma Scale of 12 with downbeat nystagmus of the right eye. Brain computed tomography scan and magnetic resonance imaging showed multilobulated extra-axial mass lesion located in the right CPA. The lesion was with various signal intensities in T1- and T2-weighted images suggestive of hemorrhages of different ages. T2 gradient echo sequences showed multiple sinusoid-like channels and diffuse hemosiderin deposition. These figures were compatible with cavernous malformation. The patient was operated by retrosigmoid approach. Dissection of the mass from the trigeminal, facial, vestibulocochlear, and lower cranial nerves was performed and total resection of the tumor was achieved. Histopathological examination confirmed the diagnosis of cavernoma.

**Conclusion:** Although CPA cavernomas are very rare, they should be considered for differential diagnosis when evaluating CPA lesions preoperatively for better intraoperative management and postoperative outcomes.

**Keywords:** Cavernoma, Cavernous hemangioma, Cerebellopontine angle, Extra-axial lesion

## INTRODUCTION

Cavernoma, also known as cavernous hemangioma and cavernous angioma, accounts for 10–20% of vascular malformations.<sup>[7]</sup> Most of them are intra-axial lesions and commonly occur supratentorially. Extra-axial cavernomas in the cerebellopontine angle (CPA) are extremely rare lesions, and intensive literature review by the authors showed only 19 cases that have been reported so far.<sup>[1-17]</sup>

This paper reports the clinical case of a patient with extra-axial cavernoma of CPA. Clinical, radiologic, and pathologic data are discussed, and literature review of similar cases was performed and summarized.

## CASE REPORT

A 36-year-old female patient referred to our emergency service with acute decreased level of consciousness and vomiting. Neurological examination showed Glasgow Coma Scale of 12 with

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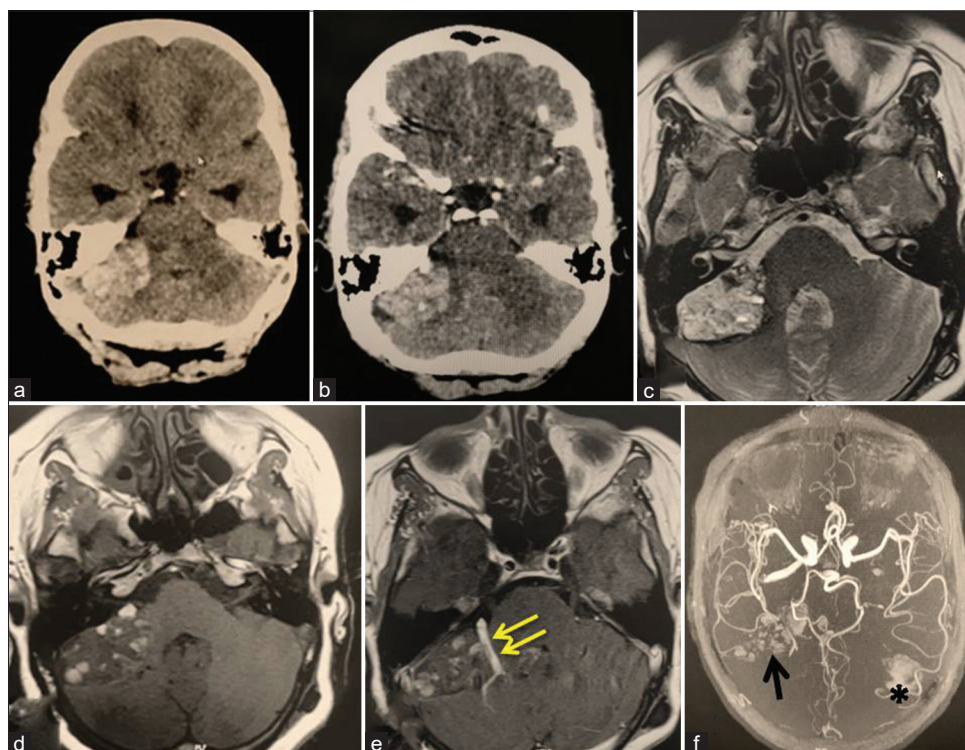
downbeat nystagmus of the right eye. No pupils abnormalities nor motor deficit were documented. Urgent brain computed tomography (CT) scan showed hyperdense lesion of the right CPA, with an associated mass effect compressing the fourth ventricle and secondary acute hydrocephalus [Figure 1a and b]. The patient was operated urgently with the left occipital ventriculoperitoneal shunt. Postoperative period was uneventful and the patient showed marked improvement and regained normal consciousness and activity in 1 week. Later examination showed persistence of the right eye nystagmus with documented right dysmetria and dysdiadochokinesia. Brain magnetic resonance imaging (MRI) showed a multilobulated extra-axial mass lesion located in the right CPA. The lesion was with various signal intensities in T1- and T2-weighted images suggestive of hemorrhages of different ages [Figure 1c and d]. T2 gradient echo sequences showed multiple sinusoid-like channels and diffuse hemosiderin deposition. Minimal heterogeneous enhancement was seen after the administration of contrast material. MRI sequences also showed signs of associated venous anomaly in the right cerebellar hemisphere [Figure 1e]. MR angiography showed sinusoid pools in the site of the lesion [Figure 1f], and digital subtraction angiography revealed occult lesion and

confirming the presence of associated developmental venous anomaly. The patient was operated by retrosigmoid approach. A gray and soft vascular extra-axial mass was exposed, with old hemorrhagic components and minimally adherent to the cerebellum and brain stem. Dissection of the mass from the trigeminal, facial, vestibulocochlear, and lower cranial nerves was performed and total resection of the tumor was achieved [Figure 2a-c]. Early postoperative period was uneventful, however, the patient showed no change in the cerebellar examination. Postoperative brain MRI showed total resection of the mass [Figure 2d].

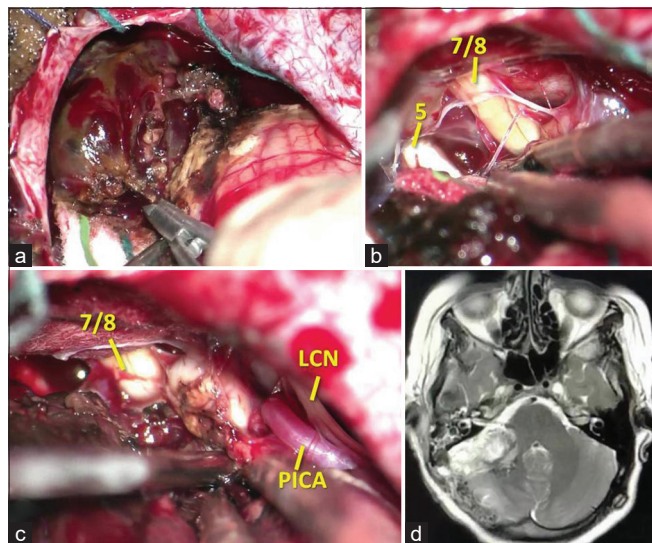
Histopathological examination revealed a lobular arrangement of dilated and congested vascular spaces lined by a single layer of endothelial cells. Intervening stroma was fibrocollagenous and devoid of nervous tissue, compatible with the diagnosis of a cavernoma [Figure 3].

### Methods

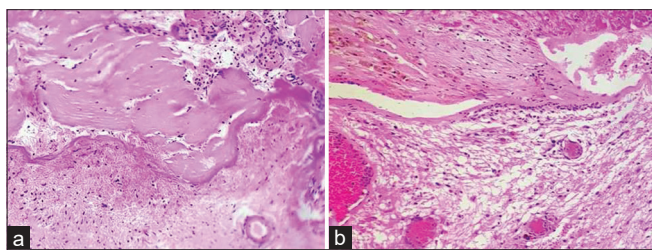
The authors performed a review of the English literature considering extra-axial CPA cavernomas published in PubMed, reported up to June 2020. Searching key words included “cavernoma,” “cavernous malformation,” “cavernous



**Figure 1:** Preoperative brain neuroimaging showing an extra-axial vascular mass lesion involving the right cerebellopontine angle (CPA) with associated venous anomaly. (a) Brain computed tomography (CT) scan without contrast, (b) CT scan with contrast, (c) axial T2 weighted, (d) axial T1 weighted without contrast, (e) postcontrast axial T1 weighted, and (f) MR angiography. Double arrow: associated venous anomaly, arrow: sinusoid pools in the site of the lesion in the right CPA, asterisk: small intraparenchymal hematoma in the site of VP shunt insertion.



**Figure 2:** Intraoperative views showing: (a) the gross appearance of the extra-axial lesion in the cerebellopontine angle, (b) dissection of the trigeminal nerve and the facial/vestibulo-cochlear complex, (c) dissection of the lower cranial nerves and posterior inferior cerebellar artery. (d) Early postoperative brain magnetic resonance imaging showing total resection of the lesion. 5: Trigeminal nerve, 7/8: Facial/vestibulocochlear complex, LCN: Lower cranial nerves, PICA: Posterior inferior cerebellar artery



**Figure 3:** Histological examination showed a multilobulated lesion composed of sinusoidal vascular spaces of various sizes lined with a single layer of endothelial cells. These vascular spaces are separated by fibrous connective tissue. (a and b) H and E  $\times 200$ .

hemangioma,” “cavernous angioma,” and “cerebellopontine angle.” Case of intra-axial cavernomas of the cranial nerves in the CPA was identified and excluded. The results of the search showed a total of 19 cases of extra-axial CPA cavernoma. These cases, in addition of our case, are summarized in [Table 1] and analyzed.

## RESULTS

In the review, there were 14 males (70 %) and 6 females (30 %) (ratio M:F = 2.3:1). The age of presentation ranged between 16 and 76 years (mean =  $41.6 \pm 6$  years), with mean age of 44.4 years for males and 35 years for females. The most common presenting symptom was vestibulocochlear which was seen in 16 patients (80%), followed by cerebellar

in 10 patients (50%), trigeminal in 8 patients (40%), facial in 7 patients (35%), and lower cranial nerves’ palsy in 3 patients (15%). There was a variable overlap and combination of the symptoms and signs in the patients.

All cases documented their CT scan findings reported iso- to hyperdense lesions with variable calcifications and enhancement. On MRI, most cases showed typical multilobulated reticular core like that of intra-axial cavernomas (70%) and presented a hypointense surrounding rim on T2 or T2 gradient and hyperintense on T1. Eleven cases (55%) showed postcontrast enhancement, most of them enhanced heterogeneously. The majority of cases were sporadic, except two cases reported by Albanese *et al.*<sup>[1]</sup> and Wang *et al.*<sup>[16]</sup> were associated with cerebral cavernomatosis. In cases evaluated with angiography, all lesions were found avascular, with three cases found to be associated with developmental venous anomaly (Ferrante *et al.*,<sup>[6]</sup> Wu *et al.*,<sup>[17]</sup> and our case).

The surgical approach for these lesions in all cases was the retrosigmoid approach. Intraoperatively, all lesions were hemorrhagic with xanthochromic fluid-filled cavities, and they appeared as red-purple-gray-bluish lesions, adherent to the brainstem, adherent or encasing the trigeminal, VII-VIII complex, or lower cranial nerves. Outcome after surgery was documented in 19 cases. Thirteen cases showed postoperative improvement of the neurologic symptoms/signs; four cases were stable without significant change; one case presented a worsening of facial paresis; and one patient died from massive bleeding at day 3 postoperatively.

Histologically, all cases showed typical pattern of lobular arrangement of dilated and congested vascular spaces lined by a single layer of endothelial cells. Intervening stroma was fibrocollagenous and devoid of nervous tissue.

## DISCUSSION

Cavernomas are considered as cerebral angiographically occult vascular malformations. They consist of dilated vascular sinusoid channels lined by endothelium, without an associated media or adventitia and without intervening brain parenchyma, with the absence of associated feeding arteries or draining veins.<sup>[1,14]</sup>

They are benign and slow-growing lesions. Recurrent hemorrhage from the sinusoids of the vascular malformation or from the neocapillary of the cyst wall and the osmotic transport of water into the cyst are thought to induce the growth of the cyst.<sup>[1]</sup> Most of them are intra-axial lesions and commonly occur supratentorially.<sup>[2,7,14]</sup> Extra-axial cavernous angiomas are rare. Moreover, extra-axial cavernomas in the CPA are extremely rare lesions.

According to Deshmukh *et al.*,<sup>[4]</sup> the CPA cavernous malformations are radiologically subdivided into two

**Table 1:** Summary of the reported cases of extra-axial cavernomas of the cerebellopontine angle.

Age (years)/sex	Clinical presentation	Postoperative follow-up	Reference
30/M	hearing loss, bilateral papilledema, V1-V2 sensory loss	No improvement, postoperative facial palsy	Iplikcioglu et al., 1986 <sup>[9]</sup>
60/M	Facial numbness, tinnitus, hearing loss, vertigo	Postoperative bleeding and death on day 3	Brunoni et al., 1996 <sup>[3]</sup>
32/M	Facial hypesthesia, facial paresis, sensorineural hearing loss, facial sensory loss	Improved facial palsy	Kim et al., 1997 <sup>[10]</sup>
24/F	Nausea, vomiting, hearing loss, positive Romberg test, vestibular impairment	Facial palsy improved on postop day 10, persistent hearing loss	Ferrante et al., 1998 <sup>[6]</sup>
46/M	Left-sided hearing loss, tinnitus, vertigo, horizontal nystagmus, cerebellar signs	Permanent hearing loss	Vajramani et al., 1998 <sup>[15]</sup>
19/M	Dizziness, nausea, vomiting, right decreased hearing, facial numbness and palsy, right hypesthesia V1/V2, truncal ataxia	Residual facial hypoesthesia	Beskonakli et al., 2002 <sup>[2]</sup>
25/M	Facial numbness, right hearing loss, hypesthesia V1/V2	Good recovery	Beskonakli et al., 2002 <sup>[2]</sup>
76/M	Hearing loss, dysphagia, facial droop	NA	Deshmukh et al., 2003 <sup>[4]</sup>
53/M	Hearing loss, facial palsy	Improved facial palsy, stable hearing	Deshmukh et al., 2003 <sup>[4]</sup>
57/M	Hearing loss, tinnitus, facial numbness, gait imbalance	Good recovery	Stevenson et al., 2005 <sup>[13]</sup>
48/M	Dizziness, vomiting, unstable gait, hoarseness, multiple cavernomas on MRI	Significant improvement at 3 months	Albanese et al., 2009 <sup>[1]</sup>
16/F	Headaches	No complications	Sasani et al., 2010 <sup>[12]</sup>
16/F	Hearing loss, gait instability, facial weakness	Permanent hearing loss, facial palsy recovery after 10 days	Engh et al., 2010 <sup>[5]</sup>
50/M	Hearing loss, facial numbness, unsteady gait	Complete recovery	Huang et al., 2011 <sup>[8]</sup>
36/M	Facial numbness and palsy, hearing loss	Mild improvement of hearing and facial palsy	Wu et al., 2012 <sup>[17]</sup>
74/F	Left-sided hearing loss, tinnitus, vertigo, horizontal nystagmus, and cerebellar signs	Permanent hearing loss	Otani et al., 2012 <sup>[11]</sup>
50/M	Left lower CN palsies, ataxia, cerebellar signs	Lower cranial nerve palsy recovery after 3 months	Ghanta et al., 2013 <sup>[7]</sup>
40/M	Right hemifacial spasm, multiple lesions on MRI	Recovery from hemifacial spasm after 12 months	Wang et al., 2018 <sup>[16]</sup>
44/F	Tinnitus, vertigo, nausea and vomiting, nystagmus	Recovery of symptoms	Tarabay et al., 2019 <sup>[14]</sup>
36/F	Decreased level of consciousness, vomiting, right downbeating nystagmus, hydrocephalus	Recovery of consciousness after VP shunt, persistent nystagmus and cerebellar signs	Current case

F: Female, M: Male, NA: None available information

subsets, solid or cystic. Solid lesions are usually smaller, hyperintense on T1, without any significant uptake of contrast and are very similar in morphology and shape to schwannomas, especially with the tendency to arise near the VII/VIII nerve complex, extending into the IAC.<sup>[4]</sup> The cystic lesions, on the other hand, are larger and multilobulated, with heterogeneous signal resulting from the presence of various blood degradation products, variable calcifications, and contrast enhancement, mimicking large atypical schwannomas or meningiomas. Other differential diagnoses include epidermoid cyst, dermoid cyst, lipoma, and thrombosed aneurysm. In this review of CPA

cavernomas, most of the cases (14 patients) showed typical radiologic signs of cavernoma, with multilobular appearance and reticulated core, surrounded by a hypointense rim of hemosiderin deposits. All cases documented their CT scan finding reported iso- to hyperdense lesions with variable calcifications and enhancement. In cases evaluated with angiography, all lesions were found avascular, with three cases found to be associated with developmental venous anomaly (Ferrante et al.,<sup>[6]</sup> Wu et al.,<sup>[17]</sup> and our case). The majority of cases were sporadic, except two cases report by Albanese et al.<sup>[1]</sup> and Wang et al.<sup>[16]</sup> were associated with cerebral cavernomatosis.

Histologically extra- and intra-axial cavernomas share the same pattern of lobular arrangement of dilated and congested vascular spaces lined by a single layer of endothelial cells, with fibrocollagenous intervening stroma devoid of nervous tissue. Extra-axial cavernomas typically arise from the dural surfaces or cranial nerves. Therefore, the clinical presentation is more likely to be that of cranial nerve involvement, especially the vestibulocochlear nerve (80%).

The surgical approach for these lesions in all cases was the retrosigmoid approach. This approach is proper for wide field exposure with sufficient visualization of the three CPA complexes. Intraoperatively, these lesions were described as hemorrhagic, which can cause massive bleeding.<sup>[3]</sup> For this reason, preoperative proper studying of the imaging is essential to avoid surgical procedures which are considered standard for most CPA tumors but dangerous for cavernomas.

For CPA cavernomas, lesion debulking (with suction, curetting, or ultrasonic aspiration) must be avoided to prevent entering the vascular sinusoids. Emptying the cystic part, coagulation of the capsule, shrinking of the cavernoma, and an “*en bloc*” resection as far as possible with careful dissection of the cranial nerves are the main steps for resection of CPA cavernomas.<sup>[14]</sup>

Outcome after surgery was documented in 19 cases. Thirteen cases showed postoperative improvement of the neurologic symptoms/signs; four cases were stable without significant change; one case presented a worsening of facial paresis; and one patient died from massive bleeding at day 3 postoperatively. As an important finding of the outcome review, patients with preoperative hearing impairment remained permanent, whereas all patients (except one patient) with facial palsy had significant improvement.

## CONCLUSION

Cavernomas are benign lesions and usually have a good prognosis after total resection. Although very rare, cavernomas should be considered for differential diagnosis when evaluating CPA lesions preoperatively for better intraoperative management and postoperative outcomes.

## Declaration of patient consent

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

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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