A rare case of an extra-axial cavernous angioma in the cerebellopontine angle

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ABSTRACT

Intracranial extra-axial cavernous angiomas are rare lesions. We report a rare case of extra-axial cavernous angioma in the cerebellopontine angle (CPA) in a 50-year-old male, who presented with lower cranial nerve palsy and gait ataxia. Computed tomography (CT) scan of the brain showed a hyperdense lesion in the left cerebellopontine angle. The lesion was totally excised by the retrosigmoid approach and a pathological examination confirmed the lesion to be a cavernous angioma. Following surgery, the lower cranial nerve palsy recovered significantly.

Key words: Cavernous angioma, cerebellopontine angle, cavernous hemangioma

Introduction

Cavernous angiomas in the brain account for 10-20% of vascular malformations.[1] Most of them are intra-axial lesions and commonly occur supratentorially. Extra-axial cavernous angiomas are rare. Moreover, cavernous angiomas in the cerebellopontine angle (CPA) are extremely rare lesions with few cases reported in the literature.[2] Cavernous angiomas are benign lesions and present with cranial nerve palsies, hemorrhage, or hydrocephalus due to compression of the fourth ventricle. Radiological appearance may mimic that of other CPA tumors (meningiomas and schwannomas). We report the radiologic and pathologic features of a left cerebellopontine lesion in a 50-year-old male, who presented with gait ataxia and lower cranial nerve palsy.

Case Report

A 50-year-old male presented with a four-month history of dysarthria, dysphagia, and unsteady gait. Clinical examination revealed left lower cranial nerve palsy and cerebellar signs were positive on the left side; there was no facial palsy or decreased hearing in the left ear. Contrast enhanced computed tomography (CECT) images of the brain revealed an enhancing hyperdense mass lesion in the left CPA [Figure 1]. Widening of the CPA was noted with minimal perilesional edema. The lesion was extra-axial in location with a lobulated surface. No calcification was evident within the lesion.

The retrosigmoid suboccipital approach was used to operate on the patient in the supine position with head

Figure 1: Preoperative contrast enhanced computed tomography of the brain showing a hyperdense lesion in the left CPA; axial plane (a and b), coronal plane (c), and sagittal plane (d)
turned to the right side and fixed to the Sugita head frame. Intraoperatively, there was a lesion of 3 × 3.5 cm size in the left CPA, which was vascular and bled when attempts were made to separate it from the surrounding structures. It was adherent to the petrous dura and a thin tail of the lesion extended into the internal auditory canal. Small vessels connecting the cavernous angioma and surrounding tissues were coagulated and divided. Total excision of the lesion was done.

Histopathological examination of the specimen showed a lobular arrangement of dilated and congested vascular spaces lined by a single layer of endothelial cells [Figures 2a and b]. Intervening stroma was fibrocollagenous and devoid of nervous tissue. Postoperatively, there were no complications and the patient did not have further neurological deficits. He was discharged on the seventh day after the operation. The lower cranial nerve palsy recovered significantly in three months.

Discussion

Cavernous angiomas in the brain are usually intra-axial lesions. Extra-axial cavernous angiomas are rare and generally occur in the sinuses, Meckel’s cavity, parasellar cavernous, and posterior fossa including the CPA and internal auditory meatus. 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. Cavernous angiomas confined only to the internal auditory canal (IAC) are also reported. Extra-axial cavernous angiomas in the CPA region are extremely rare tumors. Common and uncommon lesions of the CPA are listed in Table 1. In an analysis of unusual tumors in the CPA and IAC, Kohan et al., found only one case of a cavernous angioma in a series of 426 patients with CPA and IAC lesions (0.0023%).

The clinical and radiological appearance of extra-axial CPA cavernous angioma may mimic that of other CPA tumors (meningiomas and schwannomas). Cranial nerve palsies, due to long-term compression, is one mode of presentation. Compression of the cerebellum and fourth ventricle can result in hydrocephalus. Cavernous angiomas have a tendency to bleed and can present with intrallesional bleed or as subarachnoid hemorrhage. Cavernous angiomas with cystic degeneration have been reported. It may be difficult to diagnose the lesion preoperatively on radiology. They can be misdiagnosed as meningiomas or schwannomas on a CT scan. Magnetic resonance imaging (MRI) is most helpful in distinguishing between extra-axial cavernous angioma and meningioma. The MRI features specific for cavernous angiomas include hypointense rim containing hemosiderin deposits on T2-weighted images (T2WI) or diffusion weighted imaging (DWI) and marked hyperintensity on T2WI. Despite their extreme vascularity, cavernous angiomas are not visible on conventional angiography. Diagnosing the lesion before surgery can help in avoiding complications as cavernous angiomas have a tendency to bleed.

Table 1: Lesions in cerebellopontine angle

<table>
<thead>
<tr>
<th>Common lesions</th>
<th>Uncommon lesions</th>
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<tbody>
<tr>
<td>Vestibular schwannoma, trigeminal schwannoma</td>
<td>Arachnoid cyst</td>
</tr>
<tr>
<td>Meningioma</td>
<td>Lipomas</td>
</tr>
<tr>
<td>Epidermoid cyst</td>
<td>Choroid plexus papillomas</td>
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<tr>
<td></td>
<td>Metastatic tumors</td>
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<td></td>
<td>Ependymomas, lymphoma, exophytic pontine gliomas</td>
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</tbody>
</table>

Figure 2: (a) Photomicrograph of cavernous hemangiomas with closely packed blood vessels of varying wall thickness without intervening central nervous system parenchyma (H and E, ×200), (b) photomicrograph showing dilated vascular spaces lined by endothelium and separated by thin fibrous septa (H and E, ×200)
Our case was unique as the patient presented with lower cranial nerve palsy. As chronic compression by cavernous angiomas on cranial nerves or cerebellum causes symptoms, surgery is indicated for excision of these lesions. Surgery is usually done by the retrosigmoid suboccipital approach. Cavernous angiomas usually adhere to the cranial nerves. The lesion should be carefully dissected from the cranial nerves. In the present case, the lesion was well circumscribed and could be separated from the surrounding structures, including the lower cranial nerves, and was totally excised. Cavernous angiomas are benign lesions and usually have a good prognosis after total resection. Though cavernous angiomas in the CPA are very rare, they should be considered for differential diagnosis when evaluating CPA lesions preoperatively for better intraoperative management and postoperative outcomes.

References


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