

Vascular Hamartoma of Cerebellopontine Angle: A Rare Case Report

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Abstract

Cerebellopontine angle (CPA) hamartoma is rare. Roughly, 90% of CPA tumors are acoustic neuromas. Other prevalent lesions include meningiomas and epidermoid tumors, and additional lesions are rare. We present a case of 12-year-old girl who presented with left-sided hearing loss of more than 1-year duration with vertigo since childhood and headache. Magnetic resonance imaging revealed a brilliantly enhancing lesion in the left CPA extending through tent in the supratentorial region. Lesion was eroding the posterior petrous and occipital bone, extending into the subgaleal planes, causing hydrocephalus. Computed tomography imaging shown hyperdense lesion in the left CPA. She opted for surgery, lesion excised and pathology confirmed a diagnosis of left-sided CPA hamartoma (vascular). Neurosurgeons and otolaryngologists should be familiar with this uncommon tumor and include it in the differential diagnosis of CPA lesions.

Key words: Cerebellopontine angle, Hamartoma, Hydrocephalus, Meningioma, Ventriculoperitoneal shunt

INTRODUCTION

Cerebellopontine angle (CPA) tumors are a relatively rare entity and typically are either idiopathic or due to spontaneous mutations of several genes. About 90% of CPA tumor are neuromas.¹ Intracranial hamartomas do not occur frequently. Most of the hamartomas are usually asymptomatic and generally found at autopsy, computed tomography (CT) or magnetic resonance imaging (MRI) scan. The most frequently hamartomas located in CPA are symptomatic therefore most often encountered in surgical cases.

CASE REPORT

We reported a case of 12-year-old girl, brought to the pediatric Neurosurgery Department with complaints of headache and vomiting since 1-month duration. The

symptoms were aggravated since the since 10 days. The patient also presents with gait ataxia for the past 10 days duration. On examination, it was found that patient was conscious, oriented with Glasgow coma scale of 15/15 and no focal neurological deficits. There were dysmetria and dysdiadochokinesia present on left side of body. On radiological evaluation, MRI brain shown brilliantly enhancing lesion in the left CPA, extending into the supratentorial compartment. The lesion was eroding the posterior petrous and the occipital bone, extending into the subgaleal planes. It is severely compressing the cerebellum and fourth ventricle causing obstructive hydrocephalus. On CT brain shown hyperdense lesion in the left CPA, broad based toward tentorium, extending into the supratentorial compartment. With above complaints and investigations, the patient was diagnosed as a case "left CPA space occupying lesion with obstructive hydrocephalus." Following that, the patient underwent surgery of left combined pre- and retro-sigmoid approach and excision of tumor along with right ventriculoperitoneal shunt and Chhabra shunt medium pressure. Intraoperatively, there was gray-brown mass of tissue occupying left CPA with extension to supratentorial area and toward 4th ventricle. Histopathology of specimen shown with gross features consists of multiple irregular gray-brown masses of tissue together measuring 4 cm × 3 cm. Microscopically,

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specimen section shows fibrocollagenous tissue and dilated vascular channels with large areas of hemorrhage and hemosiderin laden macrophages, with impression of vascular hamartoma with hematoma.

DISCUSSION

Most common CPA tumor is schwannoma and over 90% of them arise from superior vestibular nerve.² Others are meningiomas and cysts of posterior fossa. Rare tumors are cranial nerve neuromas and vascular tumors. The patient may present with hearing loss for long duration and vertigo since childhood.³ After excluding other differential diagnosis, the more plausible explanation would be non-traumatic hamartomatous origin. In fact finely distributed, prominent vascular component represents a strong clue to the hamartomatous nature of the lesion. Hamartomatous tissue containing neuroectodermal elements may have become separated from the developing neuraxis during neural migration. Hamatomas should be considered when masses are discovered originating from eighth nerve other than superior vestibular nerve and when magnetic resonance signal characteristics vary from the T1 enhancement typically seen with schwannomas and meningiomas.⁴ Hamartomas are rare lesions and most were described as case reports or small series. Rare lesions are described as lipomas of the internal auditory canal

are lipomatous hamartomas.⁵ However, given the finely dispersed vascular components and absence of other tissue elements, we suggest the term vascular hamartoma for this peculiar lesion to alert to high vascularity that might suggest a more serious pathology on imaging procedures.

CONCLUSION

We described an unusual case that we believe to represent the description of a vascular hamartoma of CPA. Pathogenesis of this rare lesion at this location remains unknown. Hamartomas should be included in the pre-operative differential diagnosis of CPA tumors.

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