A rare case of cavernoma presenting as fourth nerve palsy

Abstract

This is a rare case of cavernoma which presented with left fourth nerve palsy. A 36-year-old male came with sudden onset binocular vertical diplopia for 1 month. On examination, there was a restriction in intorsion on depression in left eye. Magnetic resonance imaging (MRI) of brain and orbits was suggestive of cavernoma in dorsal aspect of midbrain in the right paramedian periaqueductal region. Usually cavernomas present with seizures, headache, decrease in function, hypothalamic/hypophyseal disorders. Presentation of cavernoma in a neuro-ophthalmic clinic is rare.

Anjali Raju[1]
Virna M. Shah[1]
Karthik Kumar M.[1]
Avani B. Chandran[1]

1 Aravind Eye Hospital, Coimbatore, India

Introduction

Intracranial cavernomas or cavernous hemangiomas are uncommon vascular hamartomas of the central nervous system. Estimated incidence and prevalence of intracranial cavernomas in the general population vary between 0.4 and 0.9% of the population [1]. 40% of the cases are found incidentally on neuroimaging. The most common neurological symptom in symptomatic patients is seizures [2]. The main neuro-ophthalmic manifestation of intracranial cavernomas is diplopia [3].

Case description

A 36-year-old male presented to our outpatient department with complaints of sudden onset, painless double vision with vertical separation of images for one month. There was no history of headache or vomiting or seizures. Patient was not a known case of any systemic disorder nor did he suffer from any trauma. On examination, he had a head tilt to right side. His best corrected visual acuity was 6/6 in both eyes. The fundi of both eyes were within normal limits. Oculomotor examination showed torsional restriction in the right eye suggestive of right fourth nerve palsy. It was confirmed by diplopia charting, Hess charting and Park three step test. The other cranial nerves were normal. His routine blood investigations were within normal limits.

The patient was advised neuroimaging. Magnetic resonance imaging (MRI) of brain and orbits and magnetic resonance angiography (MRA) were suggestive of exophytic cavernoma 1x0.9x0.8 cm in the dorsal aspect of the midbrain in the right median periaqueductal region close to right trochlear nerve nucleus and along the course of the right trochlear nerve (Figure 1).

The patient was advised occlusion therapy and was referred to a neurologist for further management.

The neurologist advised conservative management and started him on oral steroids in tapering dose and the patient was advised to repeat MRI after 3 months.

Discussion

Cavernous malformation consists of endothelium lined dilated spaces deficient in smooth muscle layer and tight junction. They show evidence of thrombosis and calcification [1]. The most common intracranial occurrence is in the subcortical white matter of the cerebral hemispheres. Cavernomas are low flow, low pressure lesions without arterialized veins or large feeding arteries which make them angiographically occult. They usually enhance very poorly or not at all with contrast [4]. CCM1/Krit1, CCM2/malcavernin, CCM3/PDCD10 genes have been identified with autosomal dominant pattern of inheritance. Commonly, sporadic forms with single lesion and familial forms with multiple lesions are seen [5].

Cavernomas are classified as either intra-axial or extra-axial. Intra-axial cavernomas include cavernomas in the brainstem, cerebellum or cerebral hemisphere. Extra-axial cavernomas include cavernomas primarily in the floor of middle cranial fossa, particularly cavernous sinus. 80% of the cavernomas occur in the supratentorial compartment, 15% in the infratentorial compartment and 5% of the cavernomas within the spinal cord [6].

Although cavernomas causing cranial nerve palsies have been reported in literature earlier [3], our case presented with an isolated fourth nerve palsy due to an exophytic cavernoma in the right median periaqueductal region probably causing compression of the right trochlear nerve, as there were no signs of any haemorrhage.

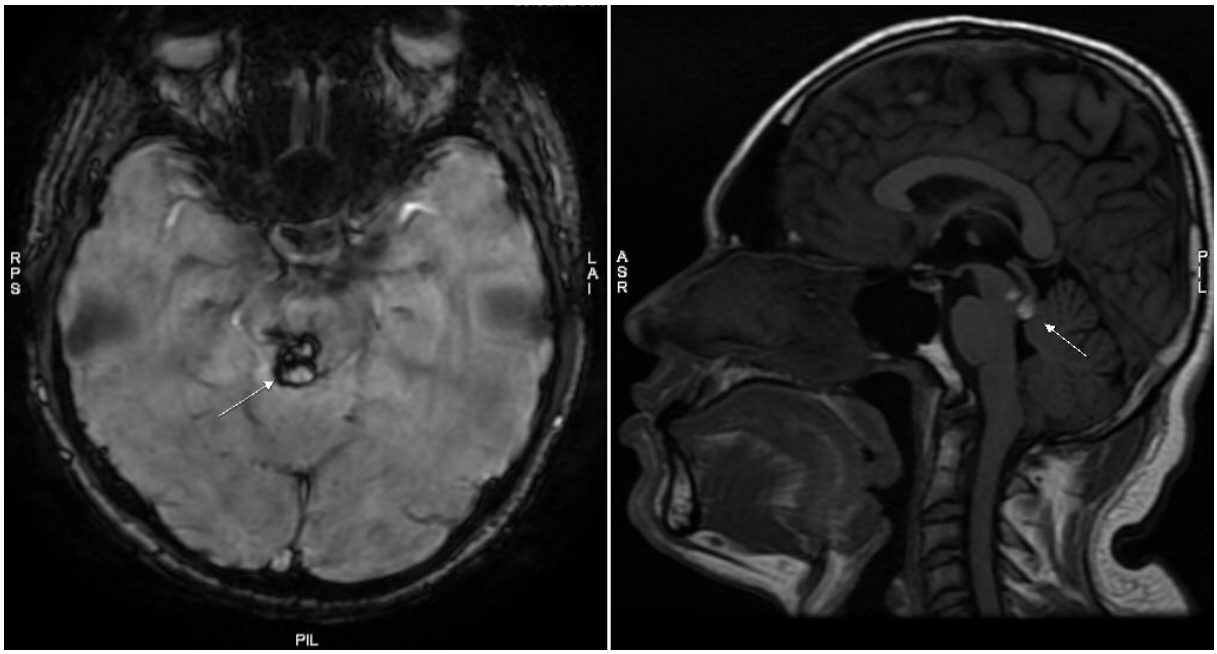


Figure 1: Axial and sagittal T1-weighted MRI images of the brain demonstrating an exophytic cavernoma in the right dorsal midbrain involving the periaqueductal region and closely related to the right trochlear nerve nucleus and its course

Cavernomas which are purely incidental in finding should be managed conservatively and followed up with annual MRI. Diplopia should be treated symptomatically with occlusion or prisms [3]. Patients with neurological symptoms like intractable seizures or progressive significant neurological deficit should be treated with surgical resection [7].

Conclusion

Cavernomas should be considered as a possible differential diagnosis of tumors causing fourth nerve palsy.

Notes

Competing interests

The authors declare that they have no competing interests.

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Corresponding author:
Dr Anjali Raju
Aravind Eye Hospital, Avinashi Road, Coimbatore 641 014, Tamil Nadu, India
anjalirajuab@gmail.com

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