Painful Mono-Ocular Vision Loss by Apical Orbital Cavernous Hemangioma-A Rare Cause

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Abstract: We present a 41-year-old male with features of painful optic neuropathy in the form of side-locked headache and ipsilateral marked diminution of vision due to an apically located cavernous hemangioma. The lesion was excised and was found to be a Cavernous hemangioma on histopathology. The patient had improvement in vision post-operatively. Orbital cavernous hemangiomas one of the common benign intracranial orbital tumors, it usually presents with proptosis and its location at orbital apex is very rare. The present case describes a rare location of orbital hemangioma and the need to include the entity in the list of differential diagnoses for lesions presenting with optic neuropathy. Besides, the case reiterates the typical imaging features of orbital hemangioma.

Key words: Cavernous hemangioma, Optic neuropathy, visual acuity

1. Introduction

Orbital cavernous hemangioma (OCH) is one of the common benign orbital tumors seen in adults [1]. It is an extremely slow growing lesion that is mostly detected incidentally [2, 3]. When present at the orbital apex, an uncommon location and a space where important anatomic structures lie, it poses a significant management difficulty [3]. The present article discusses about the detection and management of OCH in a 41-year-old male who presented with right-sided headache and ipsilateral loss of vision attributed to apically situated OCH.

2. Case

A 41-year-old non-diabetic, non-hypertensive male presented to our hospital with a history of right-sided headache since 4 months. The pain was predominantly in right frontotemporal region. It was continuous, dull and boring in character with a severity of 4/10 on VAS scale. The pain was relieved by over-the-counter pain medications. There was no history of photophobia/phonophobia, nausea or vomiting, postural or diurnal variation. The pain did not affect quality of sleep.

The patient additionally complained of progressive pain in the right eye since 3 months. The pain was moderate in intensity but did not increase in severity with eye movements. There was no polypia, redness, itching or lacrimation. In another month, the patient also developed progressive diminution of vision which began as blurring of vision and progressed to complete loss of vision over a months’ time. There was no jaw pain, scalp tenderness or lassitude. The left eye was normal. There was no history of fever, rash, joint pain, photosensitivity, oral ulcer, dryness of eyes or mouth or any high risk behaviour.

On examination, the intra-ocular pressure on palpation and measurement was normal. There was no tenderness over the right temporal area. The right temporal artery was not thickened or cord-like. There was no perception of light in the right eye. The right pupil was mid-dilated and non-reactive to light. There was no associated proptosis or bruit nor was proptosis precipitated on Valsalva manoeuvre. Fundoscopy revealed optic atrophy (figure 1). The left eye examination was normal and so was remainder of the neurological examination. Laboratory evaluation revealed normal ESR and CRP and a normal temporal artery Doppler.

MRI brain with orbits depicted a well-defined, lobulated lesion at the apex of right orbit. The lesion was T1 isointense, T2 hyperintense with progressive filling following contrast administration suggestive of slow flow vascular anomaly (Figure 1 and 2). There was no increase in size on prone images.

The patient subsequently underwent Digital subtraction angiography which was normal. The lesion was explored surgically by drilling the right anterior clinoid process and the orbital roof. It was found to be of vascular pathology and was seen to compress the right optic nerve. The lesion was completely excised. The histopathology revealed a fibrocollagenous-enveloped lesion comprising of anastomosing thick-walled vessels with myxoid degeneration. The lumen of the vessels was filled with haemorrhage and fibrin. These findings were consistent with cavernous hemangioma (figure 3). At a follow-up of one month, the patient had improvement in the vision in right eye to the extent of perception of hand movements.

3. Discussion

Orbital cavernous hemangiomas comprise about 4% of the orbital tumors in adults [1]. Epidemiology unfolds a female
preponderance [4]. The mean range of affliction is 43 to 48 years [4, 5]. OCH is usually an incidental finding in asymptomatic patients.

The OCH is most frequently seen in the intraconal compartment [1, 6, 7] with lesions in conal and extracanal compartments being uncommon. Within the intraconal space, the lesion inhibits the middle-thirds of lateral retrobulbar region [6, 7]. Its location at orbital apex is extremely rare [3, 6]. When clinically evident, its presentation is that of painless progressive proptosis [7]. Our patient did not have any proptosis rather he had headache and pain in eye without any signs of a space occupying lesion in the orbit. The orbital apical location leads to compression on the optic nerve and its vascular supply with consequent visual deficit (as was seen in our patient).

CT reveals OCH to be a well-circumscribed lesion with homogenous hyperattenuating appearance [4, 6]. The lesion on account of slow growth has a tendency to cause displacement of adjacent structures and bony remodelling. OCH seldom shows calcification [4, 6]. The lesions of intraconal space are apple-shaped while the lesions at the apex mould as per the available space to the shape of a pear [7]. The lesions take up contrast on delayed phases of imaging due to low flow arterial input [3]. This feature helps distinguish it from hemangiopericytoma, capillary hemangioma, high flow arteriovenous malformation and carotid-cavernous fistula [4].

On MRI, OCH is isointense to muscle on T1 and hyperintense on T2 weighted sequences. There is no flow void and the lesion shows progressive filling following contrast administration [8].

OCH does not show involution [6]. The management of asymptomatic ones include a follow-up while the symptomatic lesions merit excision. The degree of fusion of its capsule with the surrounding structures is the single most important determinant of surgical outcome [3]. In our case the entire lesion with its capsule could be excised.

OCH is a congenital lesion and [3, 4, 6] comprises anastomosing vascular channels with intervening fibrous stroma. The channels are lined by a flattened endothelium. At the interface with the adjacent tissues, a fibrous capsule is laid [3]. The histopathology in our case was classical of a cavernous hemangioma.

4. Conclusion

OCH though congenital usually presents in mid-adulthood. The clinical manifestations may range from painless proptosis to painful lid swelling, headache, palpable lump, progressive visual loss. The lesion is most often discovered incidentally. Location in orbital apex is extremely rare. Timely excision of the lesion has a favourable prognosis.

References


Figure 1: Fundoscopy reveals optic atrophy

Figure 2 (a): Oblique sagittal section reveals compression of intracanalicular and intracranial segments of the right optic nerve (yellow arrow) by a lobulated, T2 hyperintense lesion (red arrow)
Figure 2(b): Oblique sagittal section reveals compression of intracanalicular and intracranial segments of the right optic nerve (yellow arrow) by a lobulated, T2 hyperintense lesion (red arrow).

Figure 3(a): The lesion at orbital apex is hyperintense on FLAIR and isointense on T1WI. There is gradual filling in of contrast on delayed phase (red arrows).

Figure 3(b): The lesion at orbital apex is hyperintense on FLAIR and isointense on T1WI. There is gradual filling in of contrast on delayed phase (red arrows).

Figure 3(c): The lesion at orbital apex is hyperintense on FLAIR and isointense on T1WI. There is gradual filling in of contrast on delayed phase (red arrows).

Figure 4: Cavernous haemangioma.