A rare case of Cavernous hemangioma presenting as gaze-evoked amaurosis fugax.

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Abstract

Cavernous haemangiomas are the most common vascular lesions of the orbit in adults and accounts for 5 - 7% of all orbital tumours. They usually present in 30 - 50 years of age and there appears to be a female predilection. Cavernous haemangiomas are well circumscribed masses bounded by a fibrous pseudocapsule, without prominent arterial supply (accounting for the relatively slow enhancement). They are composed of dilated large vascular spaces lined by flattened and attenuated endothelial cells. As blood flow is slow, and vascular spaces large, areas of thrombosis are common. The usual clinical presentation is a slowly growing orbital mass resulting in proptosis, diplopia and visual field defects (from optic nerve compression). Rarely, a gaze-evoked amaurosis fugax or headache may result. Amaurosis fugax is a clinical term, describing the occurrence of transient monocular or binocular visual loss due to retinal artery ischemia.

Keywords: *Amaurosis fugax, cavernous hemangioma, transient loss of vision, gaze evoked*

Case report

Hemangiomas, which are benign vascular neoplasms, are classified as capillary and cavernous. A capillary hemangioma usually presents in the first year of life and often increases in size for 6–10 months before slowly involuting[1]. Cavernous hemangiomas are the most common benign noninfiltrative neoplasms of the orbit and have a slowly progressive mass effect[1]. They are usually present in the third or fourth decades of life and are more frequent in females[2,3].

Clinical summary

A 60 year old male presented with chief complaint of transient loss of vision in the right eye on looking towards the right side. He also complained of a gradually increasing swelling over the superonasal quadrant of the orbit for the past 10 years. He also had diplopia of 7 to 8 years duration and increasing protrusion of the right eye for the last 7-8 years. (Fig 1.)

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On examination of the right eye, a non axial proptosis with apex of the cornea 23 mm from the lateral orbital margin was noted, along with restriction of elevation in both abducted and adducted position. The swelling on the superonasal quadrant was 3cmx3cm, lobulated and cystic . The anterior segment of the eye was normal. Both direct and consensual pupillary reactions were normal. Fundus showed no evidence of retinal striae or choroidal folds.

The left eye was within normal limits in all respect. Corrected visual acuity in both eyes were 6/9 and the intraocular pressure in both eyes was 17. 3 mmHg.

MRI was done. (Fig 2a) Axial T1-weighted MR image showed an extraconal, lobulated, irregularly marginated lesion with hypointense signal and small serpentine flow voids (arrow). (Fig 2b) Axial T2-weighted fat-suppressed image showed the same lesion with slight signal hyperintensity, characteristic fine internal septa, and flow voids (arrow). (Fig 2c) Axial contrast-enhanced T1-weighted fat-suppressed image demonstrates homogeneous intense enhancement of the lesion and provides improved delineation of the flow voids.

The patient was subjected to anterior orbitotomy and mass excision. A multilobulated cystic, globular encapsulated mass 3cmx3cmx1.5cm was excised. Histopathological examination revealed a cavernous hemangioma. (Fig. 3a, 3b)

Fig 1

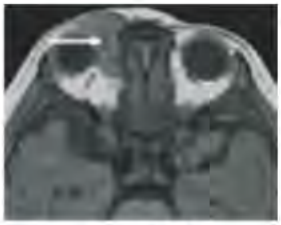

Fig 2a

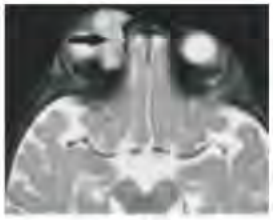

Fig 2b

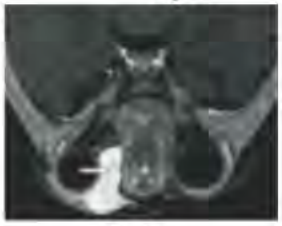

Fig 2c

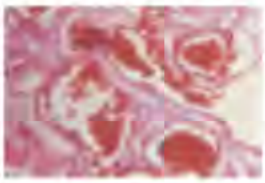

Fig 3a

Fig 3b

Discussion

Cavernous hemangioma is a congenital abnormality that presents after sufficient growth, causes cosmetic or visual disturbance. Growth of the tumor is a result of budding of the vascular channels into the surrounding soft tissue. It has been speculated that a localized, low-grade change in hemodynamics causes opening of new channels allowing for extension of the tumor into the surrounding interstitium[3]. A fibrous capsule forms at the interface of the advancing tumor and the normal neighboring tissue. Surrounding soft tissue is displaced, compressed or occasionally incorporated into the tumor. Symptomatic visual impairment occurs as a result of involvement of the optic nerve, extraocular muscles or surrounding vasculature. Although cavernous haemangiomas can be located anywhere within the orbit over 80% are located within the intraconal compartment, most commonly in the lateral aspect [5-7]. They are usually round or oval in cross section and although frequently around the globe, they do not deform it, but rather are deformed by the globe, on account of their soft consistency [5,7]. Large lesions may be associated with expansion of the bony confines of the orbit [6].

Patients with cavernous hemangiomas usually present with painless, progressive proptosis. As the tumor grows and involves the extraocular muscles, optic nerve and globe, patients will report double vision and decreased vision. Gaze-evoked amaurosis has been reported very rarely.

Depending on the size and location of the cavernous hemangioma, signs may range from normal to severe axial proptosis with poor vision, elevated intraocular pressure (IOP), motility defects and a relative afferent pupillary defect. The most common signs of cavernous hemangioma include axial proptosis, motility defects and optic nerve swelling. It does not usually produce inflammatory signs.

The mechanism of gaze-evoked amaurosis remains unclear. Suggested causes include ischaemia of either the optic nerve or retina,[5] compression of the optic nerve causing interference with propagation of axonal impulses,[10] a rise in intraocular pressure,[5] or mechanical compression of the globe[6]. In the case we have described, the most likely cause of the amaurosis is that, movement of the eye into an eccentric position of gaze results in the enlarged optic nerve compressing either nerve fibres directly or blood vessels, resulting in ischaemia.

The diagnosis of cavernous hemangioma is suspected clinically and confirmed with orbital imaging, most commonly MRI. They most commonly present as solitary, unilateral lesions.

The differential diagnosis for a well circumscribed, round to oval, solid orbital mass includes: peripheral nerve sheath tumors (schwannoma and neurofibroma), hemangiopericytoma, fibrous histiocytoma, solitary fibrous tumor and melanoma.

Management of cavernous hemangioma is dependent on the presence or absence of symptoms. For small, asymptomatic lesions, periodic observation with once or twice yearly pupil exams, visual acuity, color vision, exophthalmometry, dilated fundoscopy, visual fields and MRI studies is appropriate. For larger lesions causing diplopia or visual disturbances, surgical excision is the treatment of choice. There is no role for radiotherapy.

The choice of approach to surgical excision of a cavernous hemangioma depends on the location and size of the tumor. Cavernous hemangiomas involving the anterior two-thirds

of the orbit can be resected via an anterior eyelid, transconjunctival or transcaruncular approach. More posteriorly located tumors may require a lateral orbitotomy. A transcranial approach may be required for lesions involving the orbital apex. A cryoprobe is often used to aid in removal with minimal blood loss. Even large tumors are usually removed easily and completely. With complete excision of the cavernous hemangioma, the visual prognosis is excellent. If the tumor is incompletely excised, recurrences may occur and in some cases are relentless. Hyperopia as a result of mass effect against the posterior globe may persist even after tumor excision.

In our case, the patient recovered from the complaint of gaze evoked amarousis fugax after the surgery.

Conclusion

Though very rare, an anteriorly located cavernous hemangioma of the orbit can present with gaze-evoked amarousis fugax. Surgical excision can relieve the patient of this complaint.

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