ORBITAL VENOUS VARIX- AN UNCOMMON VASCULAR MALFORMATION

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PRESENTATION OF CASE

A 23-year-old male presented to eye OPD with complain of mass in left eye since the last 3 years. He developed mild pain and redness in the same eye since the last 10 days. There was no history of trauma, foreign body or any other relevant history. No history of any systemic illness. Vision was 6/6 in right eye and 6/36 in left eye recorded on Snellen's chart. Ocular movements were normal. On slit lamp examination of left eye, irregular growth on nasal side extending from upper fornix to lower fornix upto medial one third under the conjunctiva with overlying dilated vessels and subconjunctival haemorrhage was seen. Other anterior segment examination was normal. Right eye was normal. Fundus examination showed normal disc with dilated and torturous blood vessels in left eye more than right eye. On Valsalva manoeuvre, eye proptosed 2-3 mm. We then went for MRI orbit with contrast which revealed multiple abnormal segmentally dilated club like vascular channels at the orbital apex at both intra and extra conal retrobulbar space and median canthus of the left orbit. These vascular channels appeared hypointense on T1-weighted and hyperintense on T2-weighted sequence. The retrobulbar lesion showed post contrast enhancement however medial canthal lesions showed minimal post contrast enhancement suggesting thrombosis. This left intra orbital vascular malformation was suggestive of orbital venous varix.

DIFFERENTIAL DIAGNOSIS

There has a limited differential diagnosis of orbital varix without thrombosis. Other orbital vascular lesions to be considered in the differential include: Orbital venous malformation, Orbital Lymphangioma, Orbital haemangioma, Intracranial arteriovenous malformations, Caroticocavernous fistula, Dural arteriovenous fistula.

When the orbital varix is with thrombosis, the differential diagnosis is broader and is essentially that of an orbital mass: Orbital metastases, Orbital Rhabdomyosarcoma, Orbital lymphoma, Orbital neurofibroma / schwannoma, Lacrimal gland tumors.[1]

PATHOLOGICAL DISCUSSION

Orbital varices are rare, abnormally thin walled and distensible veins with low flow velocities. These orbital varicosities are of unknown aetiology. Equally affecting the young and the elderly.[2] Orbital varicosities are mostly asymptomatic, but may present with intermittent periorbital pain, unilateral Proptosis or acute visual loss. As it communicates with the extra-orbital circulation, engorgement may occur through a Valsalva manoeuvre or similar action (e.g., bending, coughing and straining)[3] resulting in intermittent periorbital pain, varying degrees of proptosis, thrombosis and haemorrhage.[4] Vascular complications such as variceal thrombosis or haemorrhage usually result in acute unilateral anopia due to optic nerve compression. Orbital varices are easily identifiable on either CT or duplex ultrasound imaging. CT scan is preferred when there is a suspected possibility of extra-orbital pathology. But, it is less reliable in demonstrating flow across fistulae. Fine cut (3 mm) contrast-enhanced-CT scans usually show ill-defined, heterogeneous multiloculated enhancing soft tissue mass with or without connections to the orbital and extra-orbital circulation. Thrombosed orbital varices may or may not show patchy enhancement, thus clinicians should be cautious for evidence of thrombus extension to the cavernous sinus. MR imaging, varices have hypo- to hyperintense signal on T1-weighted images, have hyperintense signal on T2-weighted MR images, and usually enhance intensely after the administration of contrast material.[5]

On other hand, duplex ultrasound is a primary imaging modality for the orbit and reliably demonstrates flow across arteriovenous fistulae of varying velocities.[6] Orbital ultrasound may be performed using a high frequency (5–7.5 MHz) probe on B-mode with colour Doppler confirming flow: to better identify varices, the patient should be examined lying down while performing a Valsalva manoeuvre in order to demonstrate flow variations within such vessels. However, it is operator dependent and cannot adequately image beyond the orbit.

Orbital varices are usually compliant to conservative management with orthoptics, to correct diplopia if present. Due to the complexity of surgical excision and the risk of visual loss related to it, surgery should be considered only in the presence of recurrent painful thrombosis, disfiguring proptosis or optic nerve compression. For patients with

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distensible varices, curative surgical methods include percutaneous variceal embolisation, alcohol injection sclerotherapy, endovascular CO2 laser ablation or direct surgical excision.

Figure 1. Clinical Picture Showing Conjunctival Mass with Subconjunctival Haemorrhage

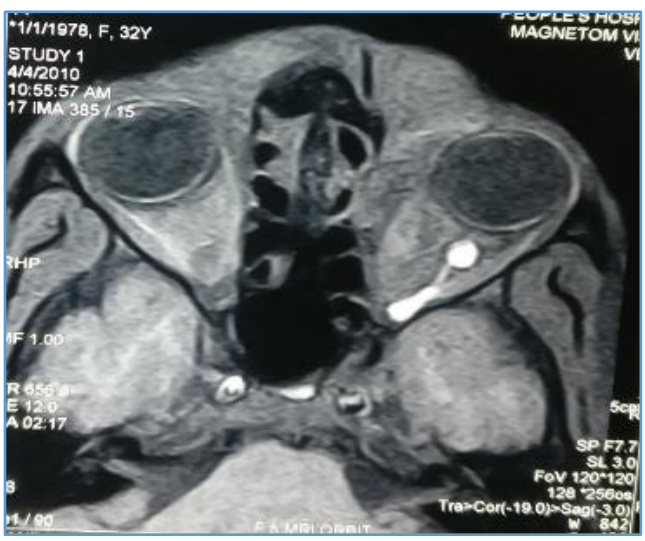

Figure 2. MRI Contrast Showing Enhancement of the Vascular Lesion

Figure 3. Showing Enhancement of Lesion in the Sagittal View

DISCUSSION OF MANAGEMENT

As most of the patient with orbital varices are asymptomatic or have very few symptoms thus requires no treatment.[7] Occasionally, periorbital pain, eye bulging, compression of the optic nerve, vision loss or disfigurement develops and require prompt treatment.[7,8,9] There are no well established guidelines for treatment. Treatment usually depends on the individual patients. Treatment with electrothrombosis (the use of an electrical current to block blood flow to the varix)[9] stereotactic gamma knife radiosurgery[7] sclerotherapy, surgical resection, and embolisation (blocking of blood flow through the varix) with cyanoacrylate glue followed by excision have been described in the medical literature.[9] In our case conservative management was opted by prescribing spectacles correction for best corrected visual acuity and patient was kept on close follow-up to look for any progression.

FINAL DIAGNOSIS

On the basis of clinical examination and as shown in MRI contrast and Color Doppler studies we had concluded with the diagnosis of left eye orbital varix.

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