TOPIC - MANAGEMENT OF ORBITAL VARIX BY SCLEROTHERAPY FOLLOWED BY SURGERY – A CASE REPORT

Ratandeep Kumar Agrawalla, Sharmistha Behera Jayashree Dora, Kanhei Charan Tudu

ABSTRACT

A 32 year old married female with complaints of intermittently appearing bluish mass in her left upper lid since last 2 years causing drooping of eyelid and cosmetic blemish. It was not associated with trauma, pain or visual loss. On clinical examination the mass was soft and reducible which reappeared on any amount of strain. The excision was planned after reducing it by sclerosant injection intralesionally. Sclerosant was also injected intraoperatively. On monthly follow up for 6 months no recurrence noted. Histopathological diagnosis confirmed it as a varix which is rare cause of orbital mass.

KEY WORDS

Orbital Varix, Sclerosant, Arteriovenous Malfomation

INTRODUCTION

Orbital mass due to any etiology is unacceptable to patients because of its symptoms like proptosis, ptosis, diminished vision and impaired cosmesis. Orbital mass may be present in central muscular cone producing axial proptosis with visual impairment, in peripheral cone may produce eccentric proptosis. Orbital mass have various etiology like inflammatory, infective, neoplastic, foreign body granuloma, arteriovenous malformation, venous malformation.

Orbital varix i.e venous malformation of orbit is low flow vascular lesions resulting from vascular dysgenesis1. Varices consist of weakened segments of orbital venous system of variable length and complexity2. According to Llyod 3 orbital varices are of two types, the first are the varices not associated with an arterio-venous malformation within the orbit or in the cranium. It consists of primary congenital venous malformations or mural weaknesses that are not due to arterialisation of the venous system.

The second are the type of orbital varices which are secondary to an intraorbital or intracranial arterio-venous communication. These angiomas are usually present in the middle cranial fossa having venous communication through the superior orbital fissure. Classification of initial symptoms of orbital varices into five groups by Wright:

1. Variable proptosis, associated with dilated veins in the lids and episcleral tissue
2. Dilated veins in the lid and anterior orbit
3. Variable proptosis without any visible lesions
4. Acute orbital haemorrhage
5. Acute thrombophlebitis.

In addition to these, venous varices may be associated with recurrent orbital pain, motility disturbances and diminished vision due to optic nerve involvement.

Walsh & Dandy5 observed that although the anomaly is congenital in origin, the clinical signs may become apparent later in life, most cases presenting themselves in the first three decades.

Orbital venous flow malformations are classified as superficial if they involve the periorbital skin, conjunctiva, or eyelid without any extension posterior to the equator of the globe. Deep orbital lesions are located posterior to the equator of the globe without extra orbital involvement. Lesions with both superficial and deep orbital components are described as combined lesions6.

Orbital varix are treated for symptomatic patients like painful eye, visual impairment and for cosmetic reasons. Treatment of orbital varix is conservative like observation, sclerotherapy, embolization, surgical exposure and resection.
CASE REPORT

32 year old female housewife presented with chief complaint of a bluish pigmented mass on left orbit which appears intermittently after coughing or strainous activity but becomes invisible when she lies down. The swelling was noted 2 years back and it gradually increased in size and was not associated with pain, tenderness or bleeding. Due to the mass the patient complained of drooping of left upper eye lid. On examination the skin over the swelling was bluish in colour and the swelling appears only after coughing or valsalva manuover. On palpation the swelling is soft reducible and cannot be traced on its upper margin. The swelling measured 1 cm in width. There was mild ptosis in the left upper lid on auscultation no bruit was heard. Other ophthalmological findings are within normal limits. Best corrected visual acuity for both eyes was 20/20.

CT scan of brain and orbit does not show any abnormality in resting state. On valsalva manuover CT showed smooth lesion which is contrast enhanced, suspected venous malformation. Other routine investigations were done and are within normal limits.

The patient was willing for treatment. Written consent was taken and patient was explained about sclerosant and excision procedure.

0.2 ml Sclerosant (sodium tetradecyl sulphate1%) was injected in the lesion after clamping it and the site was compressed adequately for 24 hours. The size of the swelling was measured. The swelling shranked a little but still bothering the patient. So surgical exision of the swelling was planned.

A curvilinear incision parallel to the brow 1 cm below, the length of incision being 1 cm. The soft tissues were dissected in anatomical layers. The venous malformation was noted as a deeply pigmented lobulated mass with radiating tributaries. All the tributaries were ligated with 6’0 vicryl. 0.2 ml of 1% Sodium tetradecyl sulphate was injected. The mass was then separated from its connections. Hemostasis was secured. Wound was closed in layers with 6’0 vicryl. Skin was sutured with 5’0 silk. The excised specimen was sent for histopathological study and a diagnosis of varix was confirmed.

Surgical dressing was done. Patient was followed up after 7 days wound was healthy and the swelling was not present even after valsalva manuover. Follow up period constituted 6 months at monthly interval.
DISCUSSION
Orbital varix is a very rare cause of orbital mass. It comprises of only 1.3% of orbital masses. Other orbital vascular lesions to be considered in the differential include:

- orbital venous malformation
- orbital lymphangioma
- orbital haemangioma
- secondary orbital venous varix
  - intracranial arteriovenous malformations
  - carotidocavernous fistula
  - dural arteriovenous fistula

It should be noted that there is confusion and controversy in the literature about the precise definitions of orbital vascular malformation with a venous component (e.g. varix, lymphangioma, venous malformation). When thrombosis has occurred the differential is broader and is essentially that of an orbital mass.

- orbital metastases
- orbital rhabdomyosarcoma
- orbital lymphoma
- orbital neurofibroma / schwannoma
- lacrimal gland tumours

Treatment of orbital varix differs from patient to patient and should be individualized. Treatment is essentially observational if asymptomatic. If patient is symptomatic treatment may be conservative like sclerosant therapy with different sclerosants, embolization, laser treatments. Surgical treatment like excision of the varix is done but for posteriorly located varices it is very tough to completely resect it out. The anterior part of the varix can be easily excised surgically if thrombosed, but it gets difficult to identify it in a supine patient if thrombosis is absent. Subtotal excision may result in recurrence, and repeat treatment is often more complicated. Ideally the vein should be resected or clipped as far back as possible towards the orbital apex.

The goal of sclerotherapy is to occlude the vessel either through direct embolization or by inciting an inflammatory response. For sclerotherapy various sclerosants like ethanol, bleomycin, acetic acid, sodium tetradecyl sulphate, sodium morrhuate, OK 432 etc are used. The success rate of sclerosant therapy is moderate around 60-70%.

Sodium tetradecyl sulfate is an anionic surfactant which occurs as a white, waxy solid. Sodium tetradecyl sulfate is a sclerosing agent. Intravenous injection causes intima inflammation and thrombus formation. This usually occludes the injected vein. Subsequent formation of fibrous tissue results in partial or complete obstruction.

Surgical excision is warranted in case of large symptomatic patients and for cosmetic purpose but recurrence after surgery is due to incomplete resection.

So prior sclerosant treatment and then surgical excision of anterior superficial orbital varix in this case prevented recurrence.

Future treatment modalities like anti VEGF and pigment epithelium derived growth factor is under trial as an alternative to sclerosant therapy.

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