

An Orbital Swelling - Venolymphatic Malformation: A Case Report

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Abstract

Patients presenting with orbital vascular malformations are unusual in ENT outpatient Department. Lymphangioma of an eyelid is recherché though it can occur in the orbit and conjunctiva. Lymphangioma can be traumatic or atraumatic and are the product of sequestered lymphatic sacs. A comprehensive examination is a must in such cases because unless detected with adequate diagnostic tools its diagnosis could be misleading. CT-Scan imaging is helpful in diagnosing but histopathological examination provides the end result. Vision maybe compromised due to surgical treatment of deep intraorbital vascular malformations intraoperatively and postoperatively. Here is a case of 16 yr old male with a swelling over right eyelid which grew over one and half month, clinically diagnosed as dermoid cyst which was subsequently diagnosed as venolymphatic malformation (lymphangioma).

Keywords: Dermoid Cyst, Ectatic Blood Vessel, Orbital Swelling, Unilateral proptosis, Venolymphatic Malformation

INTRODUCTION

Sequestered lymphatic sacs not communicating with peripheral draining channels could be its (lymphangioma) point of provenance, is considered. Histologically they are classified as: cystic hygroma, cavernous hemangiomas, capillary hemangiomas and vasculolymphatic malformations. Since they are multi-locular and predominantly cystic masses with both septa and solid components differentiating them with ultrasonography is arduous.¹⁻²

The incidence of venous malformation is approximately 1:5,000-10,000.³

CASE REPORT

A 16 year old boy came to Otorhinolaryngology OPD with a sudden swelling over right eyelid which was painless, with proptosis and sluggish globe, since one and half month. Eye movement was not restricted. Diplopia on upward gaze

was absent. The intraocular pressure of the eye measured by applanation tonometry was within the normal limits. Visual acuity, visual field and pupillary function were normal. Fundoscopic examination results were normal and no bruit or pulsation were noted or felt. On postural changes no variations were detected on proptosis of the eye. On Intra-venous contrast CT-Scan of paranasal sinuses a 3.1 cm × 2.1 cm sized well defined, low intensity mildly enhancing lesion (25hu) (Figures 1 and 2) in the extraconal compartment of right orbit along the supero-medial portion suggestive of lymphangioma. Based upon the clinico-radiological and ct- scan findings, a tentative diagnosis of venolymphatic malformation was made with a differential diagnosis of Dermoid cyst. A surgical excision was done of the lesion under general anaesthesia and the tissue was sent for histopathological examination (Figures 3 and 4). Histopathological microscopy of the lesion revealed cyst lined by atrophic, cuboidal epithelial cells and cyst containing red blood cells with surrounding tissue showing ectatic blood vessels and lymphatic channels.

DISCUSSION

Neoplasms and inflammations are the most common differential diagnosis of unilateral proptosis of eye in children which have acute onset and are not traumatic. Not only are they disfiguring but are also usually associated with complications, such as pain, ulcers, bleeding, and the compression or invasion of adjacent structures. Lymphangiomas can be superficial or deep, and they can involve single or multiple anatomical sites. The cheek, neck, eyelids, lips, tongue, soft palate, parapharyngeal space, and floor of the mouth are most commonly affected sites. The colour of the skin or mucous membrane may be normal or appear blue or dark purple when the entire dermis is involved. The boundary is not clearly defined, and the lesion is soft, compressible and occasionally phlebolith can be palpated.⁴

Pain, swelling, and even bleeding following trauma, secondary infection, abrupt haemorrhage of the lesions, or changes in hormonal level may eventuate. Venous malformations may develop within muscles (such as the temporal muscle, masseter muscle and tongue muscle), which are known as intramuscular venous malformations.

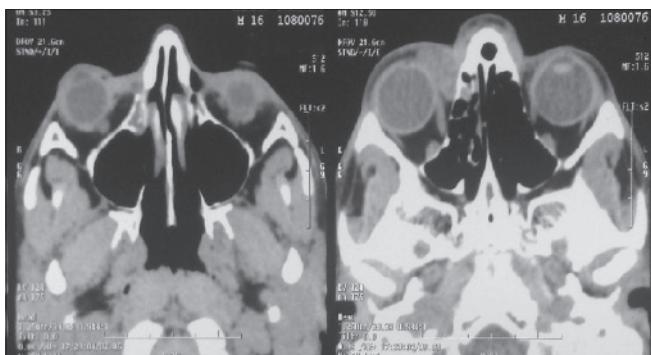


Figure 1: CT-Scan axial

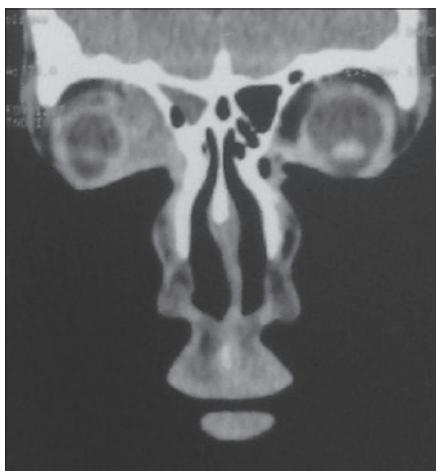


Figure 2: CT-Scan coronal

There is a reasonable debate over the classification of orbital venous malformations, with some authors asserting that the distinction between venous and lymphatic vascular malformation is artificial and prefer to treat according to clinical findings.⁵ Lymphangiomas are considered as malformations without flow, but combined venous – lymphatic malformations also exist.^{6,7}

Histologically, venous malformations (VMs) may be ectatic or micro-venular. They can be malformed in many varying sizes. Ectasia surges up with advancing age, but the rate at which this takes place is variable. Dystrophic calcification of organizing thrombi can cause formation of phleboliths, as a result of stasis in these low-flow lesions. The thrombus may become infected and cause pain and tenderness.⁴

Diverse treatment modalities are effectual for venous malformation, including surgery, sclerotherapy, laser therapy, cryotherapy, electrocoagulation treatment, and treatment with copper needles.



Figure 3: Intra operative



Figure 4: Post operative

Treatment of signs and symptoms following venous malformation is analogous to the site involved and the extent of the venous malformation. Cure may only be obtained in case of small, focal lesions.⁸ Multifocal or extensive venous malformations are rarely cured but the symptom and signs can be controlled.

Conservative treatment is primarily suitable for small, isolated, asymptomatic venous malformations. Local compression, anti-infection therapy, pain control can be adopted to suppress symptoms.

In most cases, surgical treatment is considered primarily improving function and appearance.⁸ Localized or limited venous malformations can be removed surgically.⁹

CONCLUSION

A diagnosis with thorough clinical examination and imaging technique should be carried out with histopathological correlation of the specimen. An orbital vascular

malformation should be kept in mind before making final diagnosis.

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