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Clinical challenge

The glue that holds the situation together

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1. Case report

A 48-year-old Hispanic man presented with a fluctuating mass in his left upper eyelid for several years. He noticed the lesion increased in size when he leaned forward, and he experienced pain and pressure around his left eye when bending down or lying flat. He denied visual changes or pain with eye movement. His past medical history was remarkable only for hypertension.

Visual acuity was 20/20 in each eye. Motility and confrontational visual fields were full. Margin reflex distances 1 and 2 were 4.5 mm and 6 mm in each eye, respectively. Exophthalmometry demonstrated 3 mm of relative enophthalmos in the left eye. Slight hollowing of the left lateral superior sulcus suggested left enophthalmos rather than right exophthalmos. There was some fullness of the medial aspect of the left upper eyelid that increased with Valsalva maneuver (Fig. 1). The increased fullness produced inferior and lateral globe displacement. Slit lamp and dilated fundus examinations were unremarkable. What are the considerations in an adult with a distensible mass in the orbit? What is the significance of the Valsalva maneuver? What initial workup would you recommend?

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2. Comments by Andrew R. Harrison, MD

A chronic distensible mass in an adult typically represents a venous malformation of the orbit. Other differential considerations for an orbital mass includes venolymphatic malformation, arteriovenous malformation, cavernous hemangioma, schwannoma, neurofibroma, lymphoma, and orbital metastasis. It is important to determine the flow characteristics of these lesions and their connection to the deeper orbital venous system. A lesion that shows increased size with Valsalva maneuver suggests connection with the venous drainage of the orbit. At this point, orbital imaging is indicated, and I would obtain magnetic resonance imaging of the orbit with gadolinium. Having the

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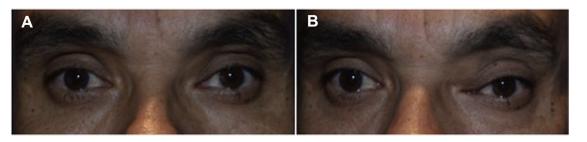


Fig. 1 - B: Frontal photograph demonstrating fullness of the left superomedial orbit (A) that increases in size with Valsalva maneuver (B).

patient Valsalva during the scan will increase the size of the lesion. This also can be accomplished by having the scan performed with the patient's head in a dependent position.

3. Case report (continued)

Contrast enhanced magnetic resonance imaging demonstrated a $2.1 \times 2.5 \times 1.3$ cm extraconal lesion in the left superomedial orbit, interposed between the superior oblique and medial rectus muscles and above the medial canthal tendon. No intraconal extension was noted. The mass was T2 hyperintense with contrast enhancement and had a layering fluid-fluid level (Fig. 2), possibly representing prior hemorrhage. The orbital contents were otherwise unremarkable. Magnetic resonance angiography demonstrated normal intracranial arterial vasculature and did not demonstrate any flow-related enhancement within the lesion to suggest a significant arterial component.

Does this alter your differential diagnosis? How do you distinguish this from a venolymphatic malformation? How would you proceed?

4. Comments by Dr. Harrison (continued)

The magnetic resonance imaging results are consistent with a low-flow venous malformation of the orbit. Orbital venolymphatic malformations are isolated from the normal orbital vascular system and are not affected by postural changes. The venolymphatic malformation would show absence of

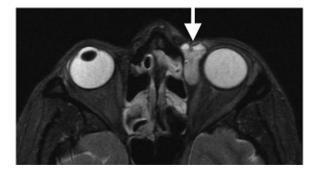


Fig. 2 – Axial T2-weighted magnetic resonance imaging demonstrating a T2-hyperintense, contrast-enhancing mass of the left orbit with fluid-fluid level (*arrow*), possibly representing prior hemorrhage.

enhancement in the lymphatic component. If the patient is asymptomatic, these lesions may be observed. For patients with pain or visual compromise, the lesion may be excised surgically. The use of preoperative embolization with glue has greatly improved the safety of surgical resection owing to improved hemostasis.

5. Case report (continued)

The clinical and radiographic findings were consistent with a low-flow, venous malformation of the orbit-or orbital varix. After discussing options with the patient, including observation, he elected surgical excision because of discomfort, orbital pressure, and disfigurement. A multidisciplinary approach was performed in collaboration with interventional radiology in a 3-step approach: fluoroscopic mapping, gluing, and surgical excision. The lesion was approached surgically through an eyelid crease incision. Once exposed, the lesion was directly cannulated, and the outflow channels were defined under direct fluoroscopy. Real-time mapping showed no high-flow vessels or abnormal communication with the superior ophthalmic vein or cavernous sinus. The lesion was then gently flushed with nonionic 50% dextrose and water (D50W) then filled with a 3:1 mixture of n-butylcyanoacrylate (NBCA) and iodinated contrast, occluding the lesion. Once solidified, the vascular malformation was directly excised.

Histopathologic evaluation of the lesion confirmed a vascular malformation with thrombosis and papillary endothelial hyperplasia (Fig. 3). The patient's recovery was uneventful, and his symptoms were relieved. At 1-year follow-up, he remains well without recurrence.

6. Discussion

Vascular malformations of the orbit comprise a spectrum of entities that are primarily classified by their flow dynamics.⁴ To differentiate these clinically, examination techniques include palpation, ocular auscultation, and observation during Valsalva maneuver. Useful imaging modalities include Doppler ultrasound, computed tomography, and magnetic resonance imaging, including magnetic resonance angiography.^{7,19}

Orbital varix is a historical term for a low-flow vascular malformation of the venous system with dysplastic vessels, but normal endothelial turnover. They are often distensible Download English Version:

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