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CORRESPONDENCE

Sir,

Late ocular manifestation of a childhood venous-lymphatic malformation

Orbital venous-lymphatic malformations are congenital vascular malformations that infiltrate into adjacent tissues comprising of arterial, venous, and lymphatic elements. These elements have led to controversy regarding nomenclature, with 'lymphangioma' and 'orbital venous anomalies' being used by different authorities. We describe a late ocular presentation of a childhood venous-lymphatic malformation.

Case report

A 43-year-old woman was referred with a 6-month history of a left-sided subconjunctival mass (Figure 1). Relevant history included left amblyopia and a left-sided vascular malformation of the tongue, diagnosed during childhood and under observation by the maxillofacial surgeons (Figure 2). There was no history of strabismus or diplopia or bleeding into the lesion.

Best-corrected visual acuities were 6/6 and 6/36 in the right and left eyes, respectively. The right eye was normal. Examination of the left vascular mass revealed a fluctuation in size with the Valsalva manoeuvre. The only other abnormalities were a 1 mm proptosis, and retinal vascular dilatation and tortuosity (Figure 3).

Magnetic resonance imaging (MRI) showed characteristics of a widespread vascular mass (Figures 4 and 5). An orbital venous-lymphatic malformation was diagnosed based on these findings. Histological confirmation was not possible. Comparison with an MRI performed 6-years earlier showed an increase in size, with new extension inferiorly into the superficial soft tissues of the neck.

Comment

Orbital venous-lymphatic malformations (OVLMs) are abnormal congenital vascular malformations composed of arterial, venous, and lymphatic vessels that infiltrate surrounding tissues. They are thought to represent a spectrum of malformations influenced by their occurrence during embryonic vasculogenesis.[1] The Orbital Society[2] has proposed the use of the term 'orbital vascular malformations' with a functional classification, which emphasises haemodynamic relationships, more pertinent to management.

Wright *et al*[3] studied the natural history of these malformations by analysing a series of 158 patients using clinical, radiological, and histological evidence. They

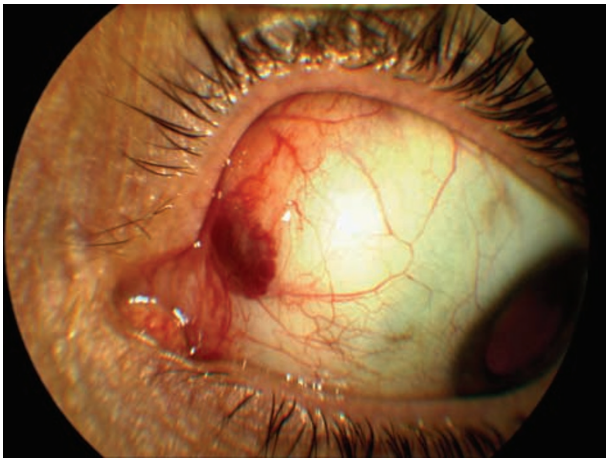

Figure 1 An asymptomatic left subconjunctival vascular mass comprising of dilated blood vessels, located just lateral to the caruncle.

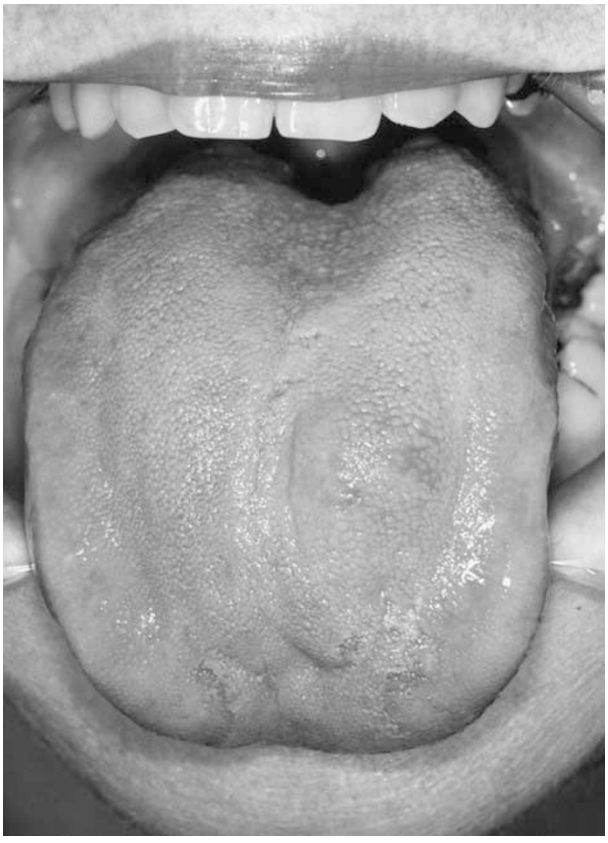

Figure 2 Vascular malformation, which presented as a lump in the tongue during childhood.

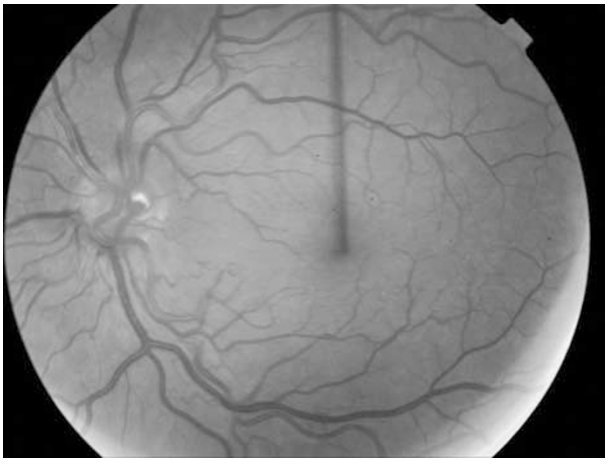

Figure 3 Mild retinal vascular dilatation and tortuosity.

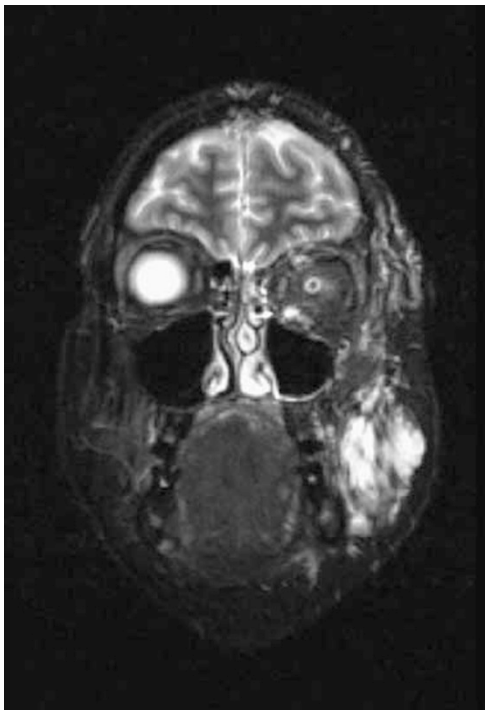

Figure 4 Coronal STIR MRI sequence showing the lymphatic-venous malformation as areas of abnormal high signal centred within the left masseter and extending superiorly to involve the temporalis and inferior rectus muscles. The left superior ophthalmic vein is enlarged.

identified a female preponderance, with most lesions presenting by childhood and occurring anywhere in the orbit. These malformations may remain clinically unapparent or enlarge slowly presenting with mass effects, haemorrhage, or pain.

Conjunctival and episcleral involvement is common.[3] Lesions can extend to ipsilateral hard or soft palate and face. Intervention is advised only in cases of compressive optic neuropathy, strabismus, or cosmetic deformity.[1,3]

MRI shows irregular, multi-loculated, heterogeneous masses that are not confined to any fascial planes.[4] Phleboliths occur in one-third of cases.[3] Histology can confirm the diagnosis by revealing irregular dysplastic endothelial channels, with a dense stroma containing smooth muscle, haemosiderin-laden macrophages, and lymphocytes.[3,4]

In this case, it is difficult to elucidate whether ocular involvement is due to a primary OVLM or secondary to extension. If it is due to primary involvement, an earlier ocular presentation might be expected, although it is possible that the decrease in left visual acuity is related to a long-standing OVLM, rather than 'true amblyopia'.

However, the clinical presentation and radiological evidence in our case favours the less common scenario whereby the orbital involvement is secondary to an active extension from the large venous-lymphatic malformation centred within the masseter.

We describe a case that clearly demonstrates the known clinical and radiological features of an orbital venous-lymphatic malformation, and emphasises the importance of searching for associated malformations and documenting progression.

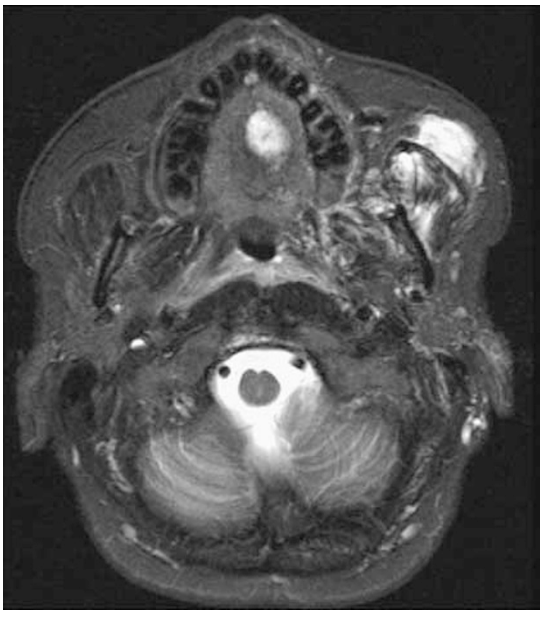

Figure 5 Axial STIR MRI sequence taken at the level of the maxilla. Abnormal high signal representing the lymphatic-venous malformation is present within the left maxilla, pterygoid, masseter, and tongue muscles.

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