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Sir,

Isolated superior ophthalmic vein thrombosis with orbital congestion: a variant of idiopathic orbital inflammatory disease?

Superior ophthalmic vein (SOV) thrombosis is a rare entity, which may present with dramatic clinical signs. We report an unusual case of isolated SOV thrombosis with overlapping features of idiopathic orbital inflammatory disease (IOID).

Case report

A 78-year-old lady presented with a 2-week history of painful and progressive right sided proptosis with accompanying blurring of vision. She also gave a history of intermittent retro-orbital pain and diplopia over a 2-year period. Her general health was otherwise good. She was on no regular medications and was also teetotal.

Ocular examination revealed right periorbital swelling with mild erythema. The affected globe was nonpulsatile but proptosed by 8 mm and also displaced inferiorly by 2 mm. Visual acuity was 6/60 (*vs* 6/9 in the left) and a brisk relative afferent pupillary defect was present. Significant conjunctival chemosis with dilated episcleral vessels was noted (Figure 1). The cornea was clear but the anterior chamber was shallow with evidence of mild activity. The intraocular pressure was slightly elevated at 28 mmHg. The pupil was fixed with posterior synaechia. Undilated fundoscopy showed generalized attenuation of the retinal vasculature with marked, apparently solid, elevation of the nasal and inferior retina. Ocular motility was restricted in all positions but more significantly in abduction and elevation.

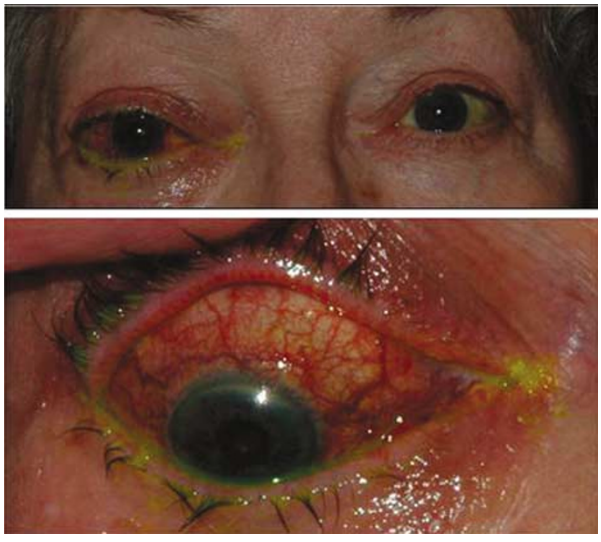

Figure 1 Acute proptosis with periorbital oedema, chemosis, and dilated episcleral vessels.

Suspecting intraocular malignancy with orbital involvement, an urgent CT scan of the orbits was performed (Figure 2). This showed a dilated and thrombosed SOV with associated swelling of the

Figure 2 CT scans showing (a) proptosis and thickening of extraocular musles, (b) dilated and thrombosed SOV (arrows), and (c) choroidal detachment with inferior displacement of globe.

extraocular muscles (predominantly superior and inferior rectii) (Figure 2a and b). The elevated area in the fundus appeared as a uniformly enhancing lesion (Figure 2c) and was subsequently shown to be a choroidal detachment by means of a B-scan (Figure 3). CT and MR angiograms excluded a dural cavernous sinus fistula and extension of thrombus into the cavernous sinus. Laboratory investigations (serum angiotensin converting enzyme, antineutrophilic cytoplasmic antibody, rheumatoid factor, antinuclear antibody, antimicrosomal antibody, antithyroglobulin, thyroid function tests, and inflammatory markers) for other inflammatory, granulomatous, and vasculitic disorders potentially causing this presentation were either negative or within normal range. Chest X-ray and abdominal ultrasound were also normal.

A diagnosis of SOV thrombosis with possible IOID, by exclusion, was made. The patient was not keen on an orbital biopsy and treatment was commenced with high-dose oral steroids and anticoagulants. Eight weeks later, ocular motility was normal, visual acuity was 6/9 and complete resolution of the choroidal effusion was noted. Although the globe was no longer inferiorly displaced, there was a small residual proptosis of 2 mm. This resolved completely after a further 8 weeks.

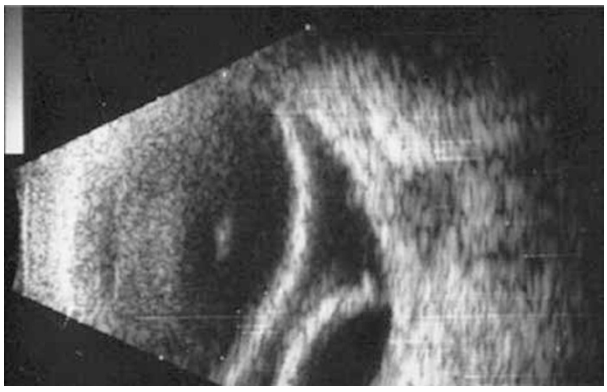

Figure 3 B-scan showing choroidal detachment.

Comment

IOID is a noninfective clinical syndrome with highly variable clinical features ranging from a diffuse to very focal inflammatory process targeting orbital tissue.[1] Proptosis, chemosis, optic neuropathy, and extraocular muscle dysfunction are nonspecific signs of orbital congestion also common to IOID. In contrast, anterior chamber shallowing and choroidal effusion have been shown to be strongly associated with SOV thrombosis.[2] The latter has been reported as a complication of sepsis and in association with dural-cavernous sinus fistulae.[2,3] Neither of these were present in our patient. To our knowledge, there is no established association between IOID and SOV thrombosis. However, colour doppler imaging studies in patients with active thyroid eye disease have shown significantly reduced blood flow within the SOV.[4] This has been attributed to compression of the SOV or its involvement in the inflammatory process. We postulate that the same mechanism, with a predominantly inflammatory component, may account for SOV thrombosis in IOID.

The importance of recognizing SOV thrombosis in association with IOID has implications for management and prognosis. IOID is known to be steroid-responsive and, although relapses and persistent inflammation can complicate the clinical course, most patients experience complete resolution over several weeks.[1] In our patient, a tapering regimen of steroids was used over 16 weeks and anticoagulation continued for 6 months to prevent extension of thrombus into the cavernous sinus.

References

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