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CORRESPONDENCE

Sir,

Treatment of postoperative uveal effusion in a nanophthalmic patient with posterior sub-Tenon's triamcinolone (PSTT)

Nanophthalmos is characterised by small, hypermetropic eyes and thickened sclera. Patients frequently develop glaucoma and primary uveal effusions (PUEs). Postoperative complications are common.[1,2] We describe the successful treatment of secondary uveal effusions (SUEs) in a nanophthalmic patient with posterior sub-Tenon's triamcinolone (PSTT).

Case report

A 72-year-old Caucasian man was referred with raised IOP (40 mm Hg OD, 30 mm Hg OS). Visual acuity was 6/9 bilaterally (+7.00 DS). Gonioscopy demonstrated open angles (Shaffer grade 1, 90°; grade 2, 270°) with normal anatomy. Central AC depth was 2.0 mm (A-scan measurement inclusive of corneal thickness). Medical treatment failed to control IOP OD, and he underwent uncomplicated trabeculectomy. Postoperatively he developed a large SUE (IOP 20 mm Hg, PI patent, AC shallow, bleb well-formed). Ultrasonography demonstrated thickened sclera with significant SUE (Figures 1 and 2), axial length 20 mm. Aqueous misdirection was excluded and nanophthalmos diagnosed. The SUE resolved initially with topical steroids, atropine, and oral acetazolamide (Figure 3). A recurrence required surgical drainage. A further, later, painful recurrence with dilated episcleral vessels was resolved following administration of oral prednisolone 40 mg. Gastric ulceration necessitated discontinuation with recurrence of SUE. PSTT 40 mg was subsequently successful with sustained effect. Later, cataract surgery was followed by SUE, responding rapidly to repeat PSTT. Final IOP was 20 mm Hg (angle open but pigmented, cup : disc ratio 0.7).

Comment

Nanophthalmic patients frequently develop SUE following surgery,[3] and it is therefore helpful to be aware of the diagnosis preoperatively. Various strategies have been proposed, from conservative management to surgical drainage.[4] Oral steroids can be effective. Deep sclerectomy, sclerotomy, intravitreal triamcinolone, and bevacizumab have been described.[4,5]

Here, trabeculectomy alone was appropriate, owing to the absence of cataract and open angle. In combined procedures, the higher cataract complication rate may compromise trabeculectomy success.

Differences may exist in pathophysiology of PUE and SUE. PUE is thought to result from increased resistance to uveoscleral outflow in thickened sclera through impedance of episcleral venous drainage. Surgical drainage is frequently indicated for PUE. SUE may have an inflammatory component. Perhaps, nanophthalmic eyes cannot compensate for increased circulation and exudation accompanying surgical inflammation, explaining an apparent role for steroids.

We have described repeatable SUE resolution following PSTT. Potential secondary IOP elevation should be considered. We propose that this novel approach offers a safe, effective management alternative and also a simple mode of surgical prophylaxis.

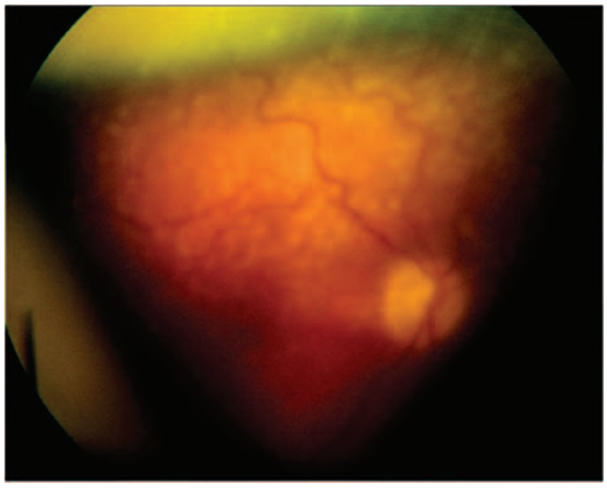

Figure 1 Fundus photograph of secondary uveal effusions.

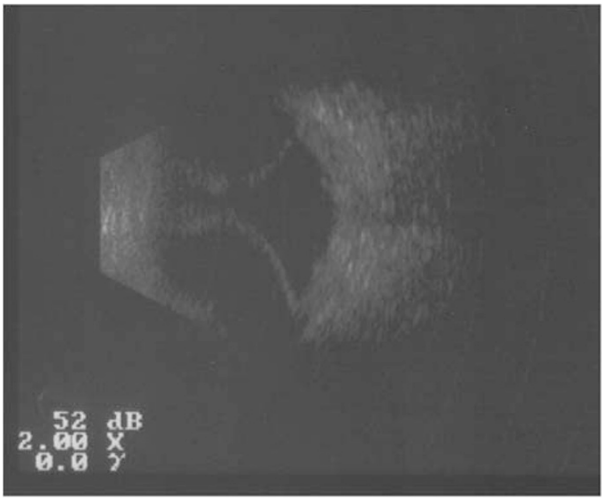

Figure 2 B-scan showing SUVs and thickened sclera.

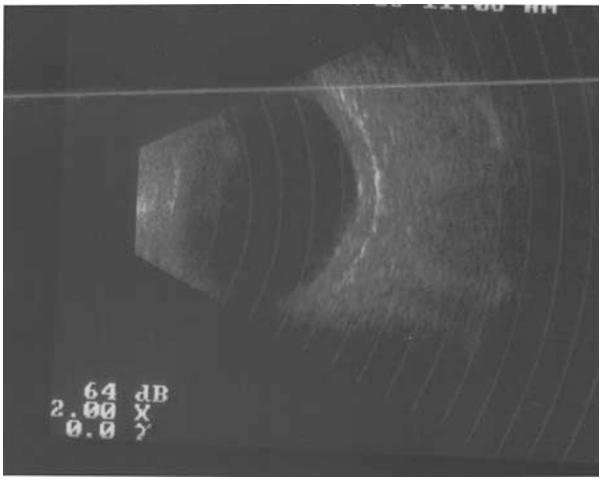

Figure 3 B-scan showing resolved uveal effusions and thickened sclera.

Conflict of interest
The authors declare no conflict of interest.

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Sir,
A possible genetic answer to a recently reported novel phenotype

We read with interest the recent article of Shen *et al*,[1] who report a unique Chinese pedigree with the features of ectopia lentis and varicose great saphenous vein. Marfan's syndrome (MFS), clinically diagnosed by characteristic multiple-system abnormalities, lies at one end of a phenotypic spectrum. At the other end of that spectrum are members of the general population who have one feature common to those with MFS.[2] Of those patients who fulfil the modified Ghent criteria for full MFS, up to 97% are found to have FBN1 mutations.[3] However, the patients presented by Shen *et al* are atypical. An alternative approach would be to try to postulate an all-encompassing molecular diagnosis that best fits the clinical signs. Venous varicosity is usually secondary to valvular incompetence, a condition that has been strongly associated with heterozygous mutations in the FOXC2 gene on chromosome 16.[4] Similarly, a range of anterior segment phenotypes have been described, with mutations in FOXC2 inherited in a dominant manner.[5] When aiming for a genetic diagnosis in this family, we would therefore advocate including FOXC2 in the screening set of genes. If a FOXC2 mutation were found in this family, this would represent an interesting extension to the associated phenotype.

Conflict of interest
The authors declare no conflict of interest.

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Sir,
Response to Khan *et al*

We thank Khan *et al*[1] for their insightful comments on our recent paper.[2] Their suggestion of including *FOXC2* in the screening set of genes is very interesting. It opens a possibly new aspect of an interesting extension to the associated phenotype in the reported Chinese family. We have already collected 18 genomic DNA samples from three generations of this family. Linkage to *FBN1* locus cannot be ruled out by microsatellite markers. A novel missense mutation was identified in *FBN1* gene, which co-segregated with the ocular phenotype (data not published). Association of single-nucleotide polymorphisms in *TGFβR2* gene was not confirmed. Obviously, this is different from the features associated to disposition to aortic dilatation and dissection of a UK family reported by Law *et al*.[3] *FOXC2*[4,5] or other genes may be the possible genetic factors as Khan *et al* pointed out. Whole-genome scanning using single-nucleotide polymorphism chips is our future strategy, which we hope can answer the question shortly.

Conflict of interest
The authors declare no conflict of interest.

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