

In most cases, MPNSTs present late with symptoms of enlarging mass and pain.<sup>2,7</sup> Our patient was fortunate that by presenting with a Horner's syndrome, further investigation resulted in the early diagnosis of MPNST. The subsequent prompt management of the MPNST will hopefully lead to a more favourable outcome in our patient.

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*Eye* (2004) 19, 351–353. doi:10.1038/sj.eye.6701478  
Published online 23 July 2004

quantifying agreement using the Bland–Altman graphical method.<sup>1</sup> In our paper we elected to use analogue measurements on the horizontal axis as this was regarded as the gold standard.<sup>2</sup> Re-plotting the graph using an average of analogue and digital on the horizontal axis did not make any difference to the limits of agreement.

We found that the limits of agreement for distance up to 5 mm were clinically acceptable, but we do accept that there appears to be a linear relationship between amount of disagreement and magnitude of distance measured. We are grateful to the authors for pointing this out, and would suspect that the most likely source of this bias might be the actual screen size (number of pixels) setting on the computer monitor. This would explain the similar gradient seen in group 1 and group 2 plots. We will conduct further studies to evaluate the influence of screen size setting as a confounding factor.

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*Eye* (2005) 19, 353. doi:10.1038/sj.eye.6701486  
Published online 23 July 2004

Sir,  
**Reply**

The authors of the letter made several points regarding the validity of our results. We agree that a correlation coefficient plot does not necessarily exclude systematic bias or disagreement between measurements obtained by the two methods being evaluated. This is the reason for

Sir,  
**Hemiretinal vein occlusion associated with pseudotumour orbit: an observational case report**

Pseudotumour orbit is a condition of idiopathic nonspecific orbital inflammation with associated retinal changes such as papilloedema, papillitis, choroiditis, and

exudative retinal detachment.<sup>1-4</sup> However, retinal vascular occlusions have hitherto never been reported in association with this condition. We present a case report of hemiretinal vein occlusion associated with pseudotumour orbit, which to the best of our knowledge is the first of its kind (as per medline search).

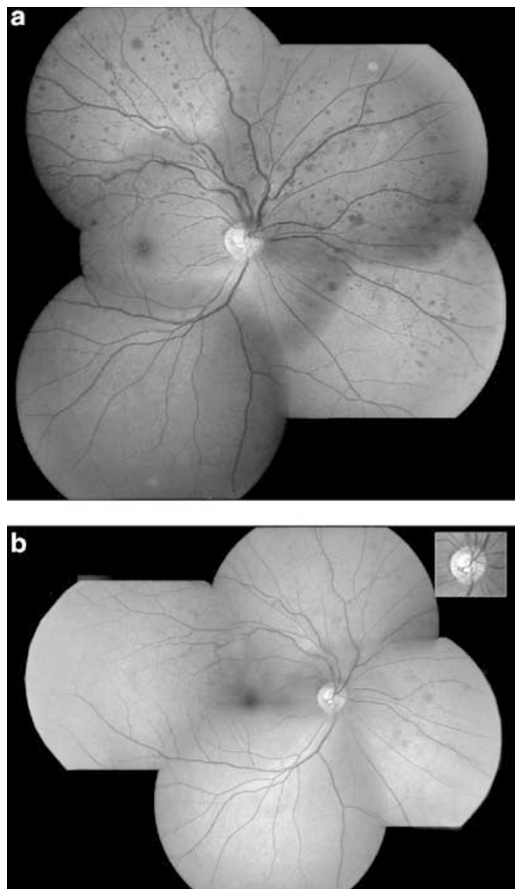
**Case report**

A 35-year-old male presented with complaints of progressively increasing protrusion and blurred vision of the right eye over the past 6 months and 1 week, respectively. Ocular examination revealed a visual acuity of 20/20, and N<sub>6</sub> in both the eyes. There was an axial proptosis of the right eye (Hertel's readings: 20 and 15 mm for the right and left eyes, respectively), with restriction of elevation. Fundus examination of the right

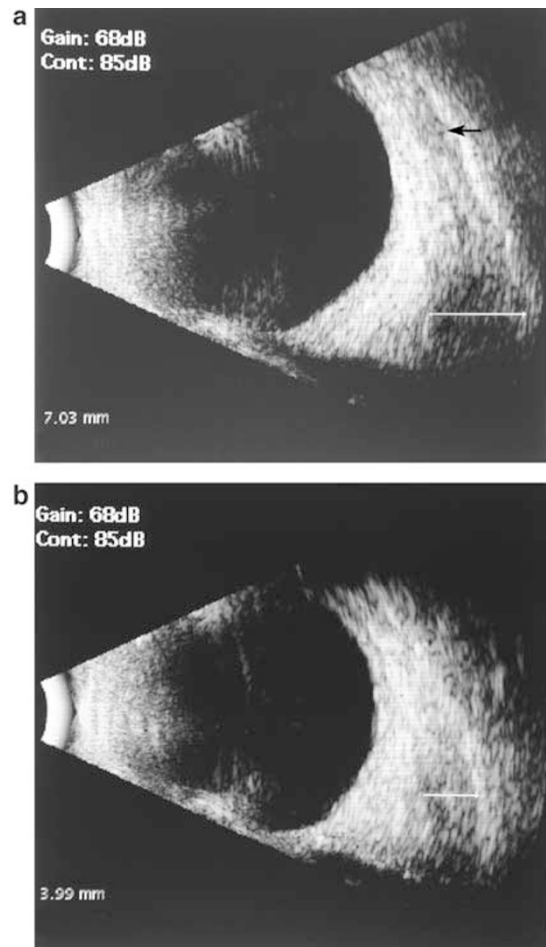
eye revealed a superior hemiretinal vein occlusion (HCRVO) (see Figure 1a), which was confirmed by fluorescein angiography. The left eye was essentially within normal limits. An electroretinogram showed the nonischaemic nature of the occlusion (*b/a* ratio of 2.1, scotopic 30 Hz flicker amplitude of 40.6 μV, and implicit time of 29.8 ms).

B and A scan ultrasonography of the right orbit was carried out, which revealed diffuse thickening of all the extraocular muscles involving both the belly fibres and the tendons, with the superior rectus-levator palpebrae superioris (SR-LPS) complex thickened the most (see Figure 2a).

Computerised tomography (CT) with contrast of the orbits was carried out, which revealed two enhancing soft tissue masses, most probably of inflammatory



**Figure 1** (a) Composite colour fundus photograph of the right eye at presentation showing venous dilatation and tortuosity and scattered superficial haemorrhages in the superior retina. The macula looks normal. (b) Composite colour fundus photograph of the right eye at 3 months of follow-up showing decreased venous dilatation and tortuosity and superficial haemorrhages in the superior retina. The inset picture shows the disc collateral.



**Figure 2** (a) B-scan echograph (longitudinal scan) at a gain of 68 dB of the patient at presentation showing the thickened SR-LPS complex (maximum thickness 7.03 mm). A Black arrow points to the thickened insertion. (b) B-scan echograph (longitudinal scan) at a gain of 68 dB of the patient at 3 months of follow-up after treatment showing the decreased thickness of the SR-LPS complex (maximum thickness 3.99 mm).

aetiology, with the larger soft tissue mass seen in the superior intraconal space superior to the optic nerve sheath and inferior to the SR-LPS complex. There was also minimal thickening and enhancement of the optic nerve sheath (see Figure 3a–d). A diagnosis of pseudotumour of the right orbit was made, based on the above findings and investigations.

General physical examination and laboratory investigations failed to reveal a thromboembolic source that could have resulted in the occlusion.

The patient was started on tapering doses of oral corticosteroids. Examination of the right eye at 3 months revealed a compensated and resolving vein occlusion as suggested by venous collateral over the optic disc, (see Figure 1b), and a resolved proptosis, which was corroborated by ultrasonography (see Figure 2b). The patient was stable and off steroids when last seen at 6 months follow-up.

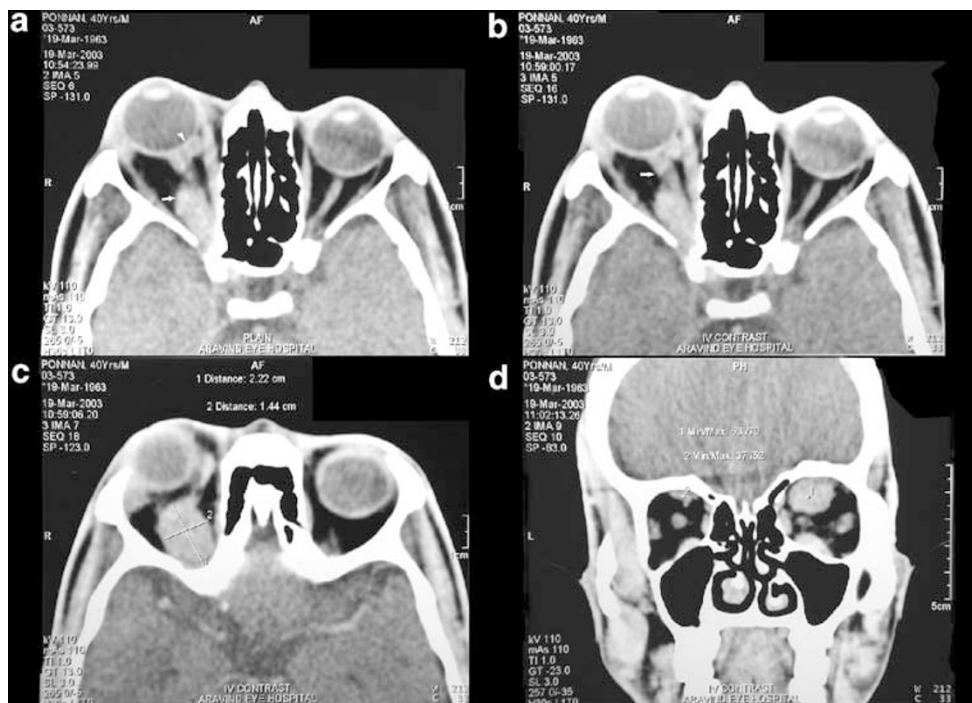
### Comment

HCRVO occur due to the persistence of the central retinal vein as twin trunks and systemic causes predominate.<sup>5,6</sup>

The failure to detect a systemic thromboembolic source in our patient to account for the superior HCRVO, along with the presence of orbital inflammation in the involved eye, suggests that the association of these two conditions is not merely by chance.

The HCRVO could have resulted from either direct compression or spill-over inflammation from the associated pseudotumour causing vasculitis. The former possibility is likely since the location of the main inflammatory mass of the pseudotumour was also superior. The pressure due to compression could have led to mechanical or haemodynamic changes in the retinal vein either at the level of lamina cribrosa or at the point of crossing the subarachnoid space around the optic nerve, resulting in its occlusion.

Alternatively, the venous occlusion could have been secondary to inflammation of the venous wall. Vasculitis has been reported in association with pseudotumour orbit.<sup>3,7</sup> However, these cases had an acute presentation with severe pain and visual loss and poor response to steroids, unlike our patient who had an insidious presentation and dramatic response to steroids.



**Figure 3** (a) CT scan picture (plain, axial scan) showing axial proptosis of the right eye, soft tissue mass in the intraconal space superior to the optic nerve sheath extending up to the orbital apex (white arrow). A white arrowhead shows a second soft tissue mass between the optic nerve sheath complex and the anterior aspect of the medial rectus just posterior to the globe. (b) Axial CT scan picture after contrast showing mild enhancement of the soft tissue masses. A white arrow shows mild thickening and enhancement of the optic nerve sheath. (c) Axial CT scan picture after contrast (superior cut) shows a mildly enhancing, globular well-defined soft tissue mass in the superior and posterior intraconal space superior to the optic nerve sheath. (d) Coronal CT scan picture after contrast showing thickening and enlargement of all the extraocular muscles of the right side and the globular soft tissue mass under the superior rectus in the intraconal space superior to the optic nerve.

Of note, vasculitis has also been reported in association with posterior scleritis.<sup>8,9</sup> As a corollary, although there was no ultrasonographic evidence of posterior scleritis in our patient, the inflammatory focus in the superior intraconal space could have spread to the adjacent superior hemiretinal vein. Additionally there was evidence of optic nerve sheath inflammation on CT scan, although this was not evident clinically or ultrasonographically. Since the superior hemiretinal vein, like the central retinal vein, crosses the subarachnoid space around the optic nerve, there is a high probability of this optic nerve sheath inflammation to spill over to the vein.

To the best of our knowledge, this is the first case report of HCRVO associated with pseudotumour orbit as per medline search. The possibility of the occurrence of such vascular occlusions due either to direct compression or spill-over inflammation should be borne in mind in cases of visual loss due to pseudotumour orbit.

### Acknowledgements

Proprietary interest: None

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*Eye* (2005) **19**, 353–356. doi:10.1038/sj.eye.6701495  
Published online 30 July 2004

Sir,

### An unusual appearance of limbal conjunctival follicles in a patient on brimonidine and dorzolamide

Conjunctival follicles are known to occur as a result of ocular allergy to topical glaucoma medication, and are located mainly in the inferior bulbar and palpebral conjunctiva. Ocular allergy to topical treatment is associated with symptoms of burning and stinging; however, these symptoms settle and the conjunctival follicles disappear after discontinuing the offending agent. We report a case of bilateral multiple limbal and palpebral conjunctival follicles in a glaucomatous patient.

### Case report

An 85-year-old Caucasian man presented in 1998 with hand movement (HM) vision in both eyes. A diagnosis of advanced open angle glaucoma with age-related macular degeneration (AMD) was given and he was registered blind. He had a presenting intraocular pressure (IOP) of 31 and 35 mmHg in the right and left eye, respectively. Retinal examination showed bilateral 0.95 cup/disc ratios with extensive AMD. He had a history of asthma, thus beta-blockers were contraindicated. After a 3-week trial, he became intolerant of Latanoprost 0.005%, which caused symptomatic blurring of vision. He was subsequently treated with topical dorzolamide 2% tds and brimonidine 0.2% to both eyes. No further blurring occurred, and IOP remained stable for 4 years. At this stage the right IOP control deteriorated, and the corresponding visual field showed progressive field loss. An augmented