References

1 Rauz S, Stavrou P, Murray PI. Evaluation of foldable intraocular lenses in patients with uveitis. *Ophthalmology* 2000; **107**: 909–919.
2 Suresh PS, Jones NP. Phakoemulsification with intraocular lens implantation in patients with uveitis. *Eye* 2001; **15**: 621–628.
3 Rojas B, Foster CS. Cataract surgery in patients with uveitis. *Curr Opin Ophthalmol* 1996; **7**: 11–16.
4 Foster CS, Stavrou P, Zafirakis P, Rojas B, Tesavibul N, Baltatzis S. Intraocular lens removal from patients with uveitis. *Am J Ophthalmol* 1999; **128**: 31–37.
5 Jones NP. Extracapsular cataract surgery with and without intraocular lens implantation in Fuchs' heterochromic uveitis. *Eye* 1990; **4**: 145–150.
6 Leatherbarrow B, Trevett A, Tullo AB. Secondary lens implantation: incidence, indications and complications. *Eye* 1988; **2**: 370–375.
7 Noble BA, Hayward JM. Secondary lens implantation: a perspective. *Dev Ophthalmol* 1991; **22**: 75–79.
8 Scott IU, Flynn HW Jr, Feuer W. Endophthalmitis after secondary intraocular lens implantation. A case-report study. *Ophthalmology* 1995; **102**: 1925–1931.

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Sir,

Unilateral acute idiopathic maculopathy (UAIM) masquerading as Best's disease

Eye (2002) **16,** 496–497. doi:10.1038/sj.eye.6700044

Acute onset of exudative maculopathy is a well-recognized feature of unilateral acute idiopathic maculopathy (UAIM).[1] We report a patient with unilateral acute idiopathic maculopathy (UAIM) characterized by predominantly macular involvement.

Case report

A 15-year-old, healthy female Caucasian patient was first noted to have a retinal abnormality during a routine visit to her optician. She was asymptomatic and had no significant ophthalmic or medical history or any history of medication. On a previous visit to her optician 6 months ago no fundal abnormality was detected. There was no history of amblyopia, squint or nyctalopia in the past. Her family history was unremarkable.

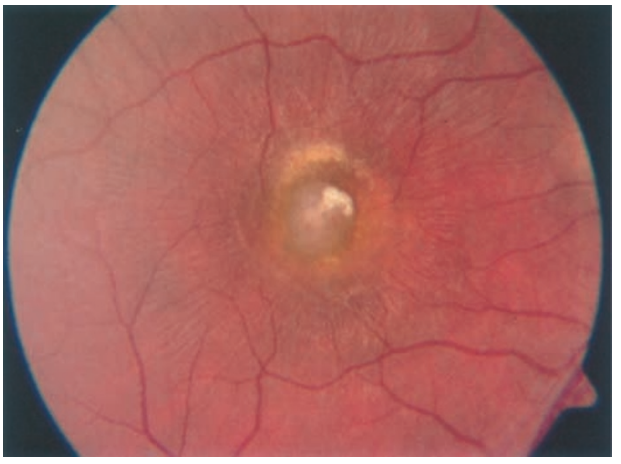

Figure 1 Fundus appearance at initial presentation.

Visual acuities were counting fingers at 1 metre in the right eye and 6/5, N5 in the left eye. Slit lamp biomicroscopy of the right eye revealed a normal vitreous cavity unaccompanied by signs of an active uveitis. The striking abnormality was the presence of a subfoveal grayish lesion at the level of the retinal pigment epithelium (RPE) measuring approximately 1/3 disc diameter in size surrounded by subretinal exudates associated with a fluffy white appearance. There was evidence of neurosensory retinal detachment overlying and surrounding the entire lesion as noted by the tenting up of the retina in the macular region (Figure 1).

Both optic discs were pink and healthy. Fluorescein angiography demonstrated hyper-fluorescence of a subfoveal lesion (Figure 2). The clinical picture was

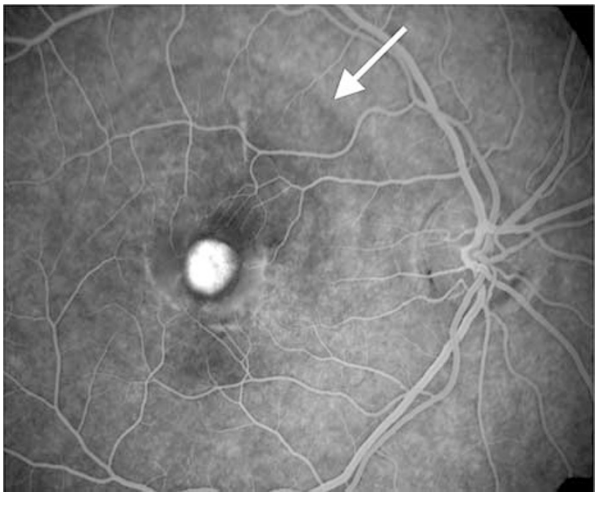

Figure 2 Fluorescein angiogram confirming a hyperfluorescent lesion at the fovea with pigmentary macular disruption (white arrow) corresponding to the neurosensory retinal detachment involving the macula.

thought to be consistent with atypical Best's disease. However, the Electro-oculogram (EOG) and Ganzfeld Electroretinogram (ERG) investigations, performed to the international standards, were normal, although in the right eye the five main responses of the ERG were slightly reduced in comparison to the left. The findings were not different with the photopic and scotopic ERG. In contrast the Wide Field Multifocal Electroretinogram (mfERG) showed marked reduction in the central responses in the right eye. Best's disease and a diffuse retinal dystrophy were therefore excluded. A revised diagnosis of unilateral acute idiopathic maculopathy was made.[2] Posterior uveitis investigations including toxoplasma and toxocara titres were within normal limits. Ultrasound B-scan of the globe and a CT scan of brain and orbits showed no abnormality. In view of her relatively young age, a periodic review was arranged. During the last 2 years the lesion has remained stable in the right eye with no involvement of the left eye.

Comment

A grayish, thickened subfoveal lesion with maculopathy is well recognized in patients with UAIM.[1] When viewed with a contact lens, vitreous cells have been noted to be a common but variable finding. In our patient, there was no evidence of activity in the vitreous cavity. Two recent series reported patients with absence of vitreous involvement in up to 50–60% of individuals.[2,3]

The yellowish gray nature of the subfoveal lesion led us to initially suspect an atypical presentation of Best's disease but the unilateral acute presentation in a white 15-year-old female patient together with normal electrophysiological studies made us revise the diagnosis. Other studies[4,5] together with the present report suggest that UAIM should be considered in the differential diagnosis of all patients with unilateral, acute onset, maculopathy of this nature.

UAIM is a distinct entity but other possible diagnoses such as dystrophies, degenerative, infiltrative, and infectious disorders should be eliminated by angiography, electrophysiology and imaging studies.[5,6] Every effort should be made to establish an accurate diagnosis, as there are implications in relation to genetic counselling for degenerative disorders such as Best's disease.

However with recent reports of several bilateral cases, it may be appropriate to rename this disorder simply acute idiopathic maculopathy (AIM) as suggested by Gass in the new edition of his atlas.[7]

We thank Professor GN Dutton, Tennent Institute of Ophthalmology, for his advice with this case.

References

1 Freund KB, Yannuzzi LA, Barile GB *et al*. The expanding clinical spectrum of Unilateral Acute idiopathic maculopathy. *Arch Ophthalmol* 1996; **114**: 555–559.
2 Yannuzzi LA, Jampol LM, Rabb MF *et al*. Unilateral acute idiopathic maculopathy. *Arch Ophthalmol* 1991; **109**: 1411–1416.
3 Hannutsaha P, Yannuzzi LA, Freund KB. The occurrence of uncommon inflammatory diseases; a survey of the macula society. *Retina* 1996; **16**: 437–439.
4 Fish RH, Territo C, Anand R. Pseudohypopyon in unilateral acute idiopathic maculopathy. *Retina* 1993; **13**: 26–28.
5 Yannuzzi LA, Slakter JS, Sorenson JA *et al*. Digital indocyanine green video angiography and choroidal neovascularization. *Retina* 1992; **12**: 191–223.
6 Guyer DR, Yannuzzi LA, Slakter JS *et al*. Digital indocyanine green video angiography and central serous chorioretinopathy. *Arch Ophthalmol* 1994; **112**: 1057–1062.
7 Gass JD. *Stereoscopic Atlas of Macular Diseases*, 4th edn. Mosby-Year Book: St Louis, 1997, p 676.

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Sir,

Bilateral ocular ischaemic syndrome in association with hyperhomocysteinaemia

Eye (2002) **16,** 497–500. doi:10.1038/sj.eye.6700010

Ocular ischaemic syndrome (OIS) is caused by chronic ocular hypoperfusion, usually secondary to severe carotid artery obstruction.[1] Iris neovascularization is the most common anterior segment finding at diagnosis and typical fundus signs include retinal arteriole narrowing, retinal venous dilatation without tortuosity, midperipheral retinal haemorrhages and microaneurysms, and peripheral vascular closure.[2–4] We report a case of bilateral OIS presenting in the absence of carotid artery obstruction and discuss possible aetiological factors, including elevated plasma homocysteine.