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]

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Sir,

Bilateral ocular ischaemic syndrome in association with hyperhomocysteinaemia

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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.

Case report

A 45-year-old man was referred to the eye department with gradual, painless loss of vision in the right eye. He had been a heavy smoker from 13 years of age (30–40 self-rolled cigarettes per day) and had previously consumed significant amounts of alcohol (up to 20 units per day). He had no symptoms of coronary, cerebral or peripheral vascular disease, but reported Raynaud's phenomenon in his hands and feet.

Initial ocular examination revealed best corrected Snellen visual acuity of right eye 6/36, and left eye 6/9. The right eye showed iris neovascularization, a relative afferent pupil defect, intraocular pressure of 22 mmHg, and mid peripheral retinal dot and blot haemorrhages. Left ocular examination was normal. Fluorescein angiography demonstrated signs of OIS with a leading edge of dye within the retinal arterioles, and midperipheral haemorrhages, microaneurysms and vascular closure.[2] Panretinal photocoagulation was applied to the right eye (argon green laser, 5000 burns at 200 μm diameter with the Volk SuperQuad 160 contact lens—magnification factor of 2.0), but over the next 3 years progressive neovascular glaucoma and cataract reduced vision to no light perception.

Seven years after initial presentation, fine iris neovascularization developed in the left eye and the intraocular pressure began to rise progressively. Left fundoscopy showed narrowed retinal arterioles and scattered dot and blot haemorrhages and microaneurysms in the midperiphery with a few extending to the posterior pole. Fluorescein angiography was again typical of OIS with a delayed arm to retina circulation time, a leading edge of dye

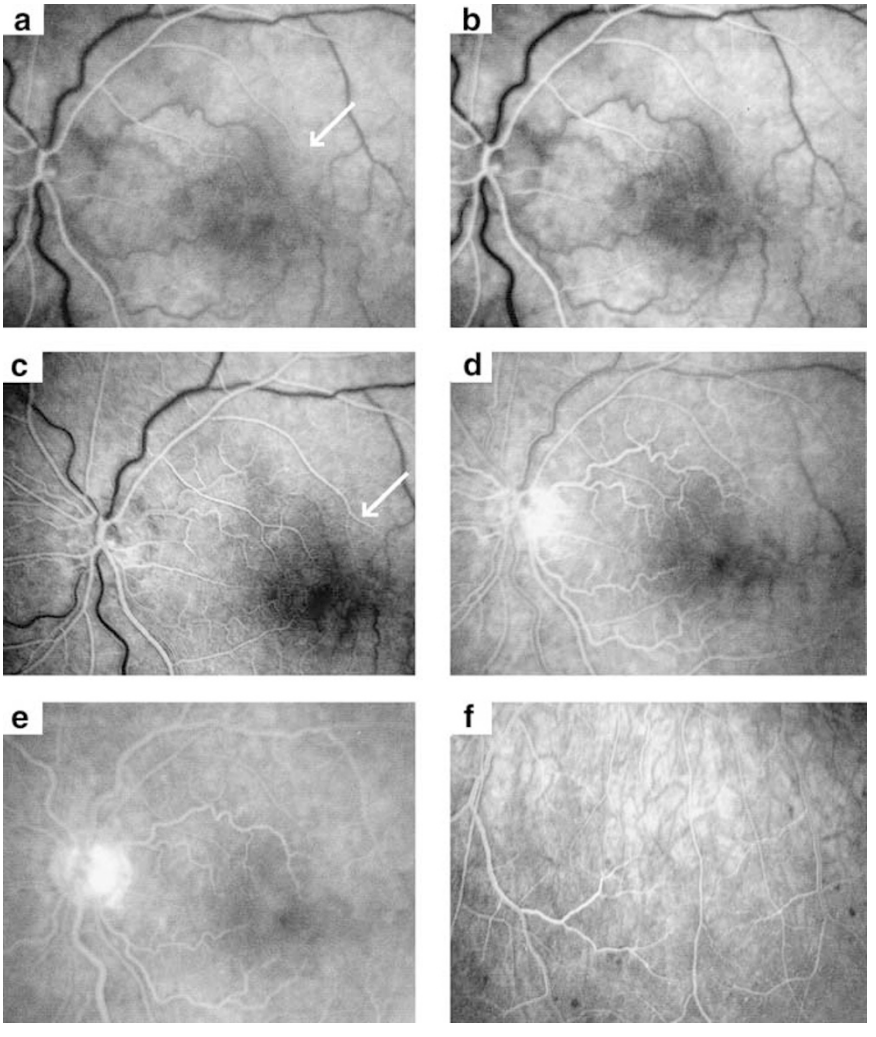

Figure 1 Left eye. Fluorescein angiogram. The times indicate the interval following injection of fluorescein. (a) 69 s. Arrow points to leading edge of fluorescein in arteriole. (b) 72 s. (c) 78 s. Arrow points to arteriole now filled with fluorescein. (d) 83 s. Note delayed venous filling temporal to fovea. (e) 97 s. Note venous filling now complete. (f) 163 s. Inferior mid periphery showing microaneurysms and blot haemorrhages.

within narrowed retinal arterioles, prolonged arteriovenous transit time (28 s) and midperipheral haemorrhages and microaneurysms (Figure 1a–f). Panretinal photocoagulation (argon green laser, 8000 burns at 200 μm diameter with the Volk SuperQuad 160 contact lens) supplemented by peripheral retinal cryotherapy caused regression of the iris new vessels, but the intraocular pressure remained elevated despite medical treatment. Following mitomycin trabeculectomy the left intraocular pressure was well controlled without medication. Eighteen months after development of iris neovascularization in the left eye, corrected acuity remained at 6/9.

Systemic investigations showed normal blood pressure (130/80) and sinus rhythm. Carotid doppler ultrasound showed normal flow in the common, external and internal carotid arteries on both sides. Ophthalmic doppler ultrasound showed normal flow in the ophthalmic arteries, the central retinal arteries and veins, and the posterior ciliary vessels of both eyes. The following blood tests were all normal: U&E, LFT, glucose, FBC, ESR, lipids, coagulation, plasma viscosity, serum immunoglobulins, serum electrophoresis, antithrombin III, protein C and S, ANA, rheumatoid factor, VDRL. Plasma vitamin B-12 was 192 ng/l (normal range 155–1100) and folate was 3.0 ng/ml (normal range 2.8–12.4). Plasma homocysteine was elevated at 20.8 μmol/l (normal range 5–15). He was treated systemically with low dose aspirin and folic acid, and was advised to stop smoking.

Comment

Our patient presented with classical signs of OIS but without carotid artery obstruction. Ocular hypoperfusion in OIS is usually secondary to severe carotid artery obstruction but a recent large prospective study found that 26% of eyes with OIS had mild or no ipsilateral carotid artery stenosis.[3] The authors of that study suggested vascular occlusive disease of the aortic arch, ophthalmic, central retinal or ciliary arteries as the cause for OIS in such patients. Doppler ultrasound studies showed normal flow in our patient's carotid, ophthalmic, central retinal and ciliary arteries. He also had no symptoms of macrovascular disease (angina, claudication or transient ischaemic attacks). We presume that widespread microvascular disease and undetected macrovascular disease (without areas of focal stenosis) had cumulatively impaired ocular blood flow.

Cigarette smoking is a well-known risk factor for vascular disease: our patient was counselled to stop smoking and low dose aspirin prescribed. As he presented at a comparatively young age, we screened for haematological risk factors for vascular occlusive disease. The only abnormality detected was a significantly elevated plasma homocysteine level. Elevated plasma homocysteine is considered an independent graded risk factor for arteriosclerotic vascular diseases,[5] and modestly elevated plasma homocysteine levels are known to cause vascular endothelial dysfunction.[6] Hyperhomocysteinaemia has recently been reported as a possible risk factor for retinal vascular disease,[7] and we now suggest that it may be an independent risk factor for ocular ischaemic syndrome.

Daily folic acid and vitamin B-12 supplementation has been shown to reduce blood homocysteine concentrations by about a quarter to a third, with the majority of the effect attributable to folic acid.[8] Although there seem to have been no large-scale trials examining the effect on vascular morbidity of lowering plasma homocysteine levels, we prescribed daily low dose folic acid for our patient as his plasma folate was in the lower end of the normal range and he had a high risk of progressive visual loss.

Panretinal photocoagulation (PRP) in OIS has been shown to cause regression of iris new vessels in 36% of eyes.[9] Moderate PRP alone was unsuccessful in stabilizing our patient's right eye. However, more extensive PRP and retinal cryotherapy followed by mitomycin trabeculectomy has currently stabilised his left eye. Through the above surgical treatments and risk factor modifications we hope this patient will preserve useful sight in his left eye.

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Sir,

Advanced Coats' disease successfully managed with vitreo-retinal surgery

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Coats' disease or idiopathic retinal telangiectasia is characterized by unilateral retinal telangiectasia associated with subretinal lipid exudation and exudative retinal detachment usually seen in young boys. The natural history is variable but is typically progressive. Patients who develop extensive exudative retinal detachment commonly progress to total retinal detachment, neovascular glaucoma, and phthisis bulbi despite use of laser photocoagulation, cryotherapy, or diathermy.[1] We describe a case of advanced Coats' disease with exudative retinal detachment in which vitreo-retinal surgery was effective in providing retinal reattachment.

Case report

A 6-year-old boy first presented to our office for evaluation of poor visual acuity in his right eye of one year's duration. Past medical history and family history were unremarkable. Visual acuity was counting fingers at two feet OD and 20/20 OS. There was an afferent pupillary defect OD. Slit lamp examination revealed quiet anterior segments OU. Ophthalmoscopy of the right eye revealed severe submacular lipid exudation (Figure 1). A lesser amount of exudation was seen in the nasal retina. An exudative retinal detachment extended from 6 o'clock to 8 o'clock peripherally. Multiple telangiectatic vessels were found on the temporal retina. The left fundus was normal. A clinical diagnosis of Coats' disease was made.

He underwent pars plana posterior vitrectomy and removal of vitreous and preretinal membranes. However, the posterior hyaloid face was firmly adherent to the retina and difficult to remove completely. Drainage of subretinal fluid through a temporal retinotomy was completed following air-fluid exchange. Endolaser photocoagulation was then applied to all telangiectatic vessels around the retinotomy and circumferentially for 360 degrees around the midperiphery. At 2 and 4 months postoperatively his retina was flat and attached, and visual acuity improved to 20/400. However, 7 months after surgery he developed recurrent vitreous membranes associated with a tractional detachment of the macula. He underwent repeat pars plana vitrectomy, removal of preretinal and epiretinal membranes and cryoablation of residual telangiectatic vessels temporally. His retina has remained attached with decreased submacular lipid exudation and absence of telangiectatic retinal vessels. At last follow-up, 27 months after initial surgery, his vision was 20/400. The retina was attached, but a significant posterior subcapsular cataract had developed (Figure 2).

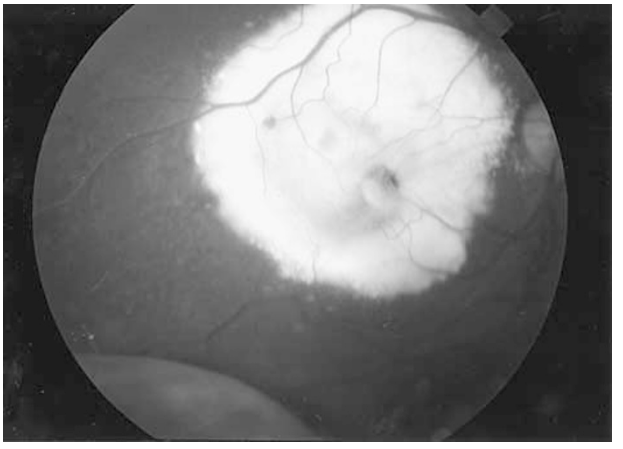

Figure 1 Preoperative fundus photograph: severe submacular lipid exudation with exudative retinal detachment.

Comment

The management of early Coats' disease is best achieved with prompt laser treatment or cryotherapy of leaking telangiectatic vessels. The best form of management of advanced Coats' disease after exudative retinal detachment has occurred is unclear. Siliodor *et al*[2] showed a series of 13 patients for which neovascular glaucoma could be prevented through subretinal drainage and cryotherapy. In their study all untreated eyes required enucleation. However, eyes that were salvaged had no useful vision. Wessing[3] has pointed out that advanced disease with detachment in two quadrants is almost untreatable.