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Sir,

Ischaemic retinal vasculitis in biopsy-proven sarcoidosis

Retinal periphlebitis is a well-recognised feature of ocular sarcoidosis, but ischaemia and vascular occlusions are only rarely reported. We present a case of biopsy-proven sarcoidosis with ischaemic retinal vasculitis.

Case report

A 47-year-old man presented in 1993 after 1 month of blurred vision in both eyes. He also had a history of malaise, tiredness, loss of weight and non-productive cough of 9 months duration. Examination showed a visual acuity of 6/9 in each eye. There was a granulomatous anterior uveitis, vitreous cells and snowballs, and multiple retinal perivenous infiltrates. On investigation abnormal results included a raised serum angiotensin converting enzyme (ACE) at 217 IU/ml, raised γGT and AST levels, and a chest radiograph which revealed bilateral hilar lymphadenopathy and increased interstitial markings. A transbronchial biopsy showed non-caseating granulomata consistent with a diagnosis of sarcoidosis. The patient was treated with topical steroids and systemic prednisolone, the latter commencing at 40 mg/day and reducing according to disease activity. Substantial improvement ensued and systemic treatment was discontinued by the end of 1996, at which time the minimal uveitis was well controlled using only topical steroids. The visual acuity was 6/6 in both eyes.

In 1998 the patient presented urgently with sudden blurring of vision in the right eye. Examination showed a visual acuity of 6/6 in each eye. There was no afferent pupillary defect. Colour vision was marginally reduced in the right eye, the patient being able to read 19/21 Ishihara plates. The left eye was entirely normal. The right anterior segment was uninflamed but there was active vitritis and some areas of substantial perivenous exudate. There were multiple small poorly defined pale retinal lesions at the posterior pole. Reactivation of sarcoidosis was presumed and prednisolone 80 mg/day was commenced. The visual acuity fell to 2/60 within 3 weeks, the optic disc becoming hyperaemic and cotton-wool spots becoming evident at the posterior pole (Fig. 1). There were no retinal haemorrhages. A mild afferent

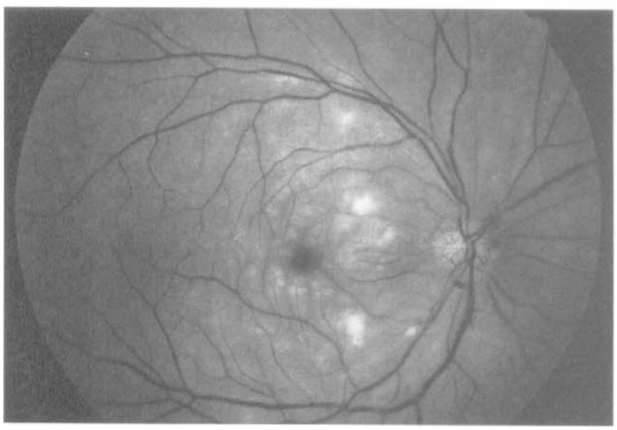

Fig. 1. *A red-free photograph of the right posterior pole shows multiple cotton-wool spots and minor calibre changes to the superotemporal venous circulation.*

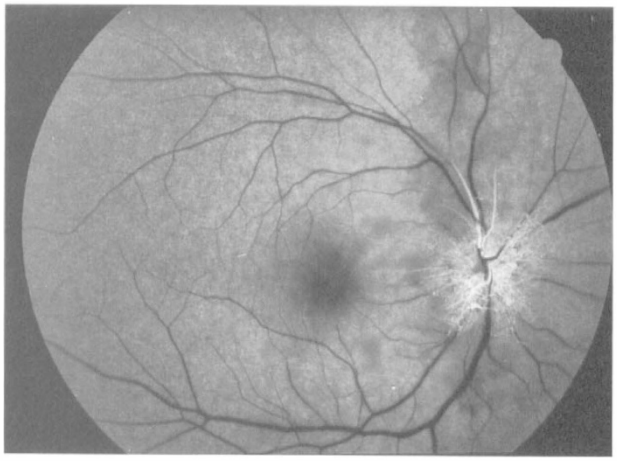

Fig. 2. *The onset of the arterial phase fluorescein angiogram (10.2 s) shows filling of optic nerve head vessels prior to complete choroidal filling.*

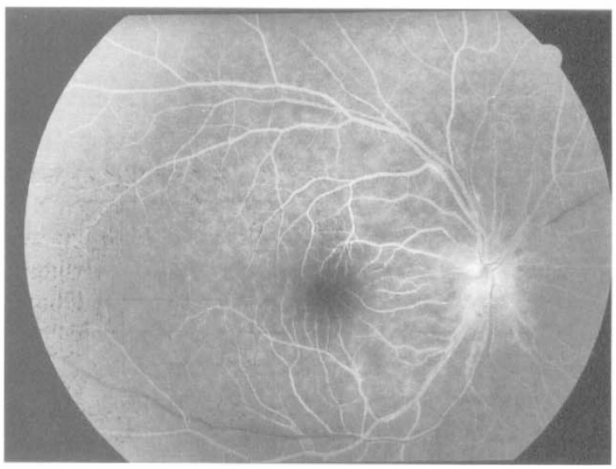

Fig. 3. *A late phase angiogram (70 s) shows that venous filling remains incomplete. Two areas of phlebitis are seen in the superotemporal vessels. There is some leakage from the optic disc.*

pupillary defect was present. Intraocular pressures were normal and symmetrical. Fluorescein angiography showed profoundly delayed retinal circulation (Figs. 2, 3) and evidence of active vasculitis. A CT scan of brain and orbits was normal, as was serum ACE. There were no symptoms or signs of active systemic sarcoidosis. There was no clinical or investigative evidence of systemic vasculitis, with ANCA, ANA, liver and renal function being normal. The patient developed disc neovascularisation and iris rubeosis, which regressed following panretinal photocoagulation, the right visual acuity eventually improving to 6/12. Currently there is minimal bilateral panuveitis and the patient is maintained on prednisolone 4 mg/day.

Comment

Retinal vasculitis is a well-recognised feature of some forms of intraocular inflammatory disease, including sarcoidosis.[1] This disease is generally associated with non-ischaemic retinal vasculitis,[2] although there are rare reports of vascular occlusion.[3–5]

Our patient has biopsy-proven sarcoidosis with uveitis and retinal vasculitis. At the time of acute visual loss there was no evidence of systemic sarcoidosis. However, the fundal appearance suggested an ischaemic event secondary to vasculitis. The profound delay in retinal circulation (incomplete venous filling after 90 s) and the presence of cotton-wool spots suggest slow flow through the central retinal artery. There was no evidence of another cause of vasculitis. These circumstances imply sarcoidosis as the cause of retinal ischaemia in our patient, and the presumed pathological process is a pressure effect caused by granulomatous inflammation within the optic nerve. This case highlights the possibility of ocular sarcoidosis as a cause of occlusive vasculitis, even in the absence of systemic inflammation.

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Sir,

Lens notching in association with presumed Marfan's syndrome

Lens notches, often called coloboma, are not true coloboma as there is no focal absence of a tissue layer due to failure of closure of a fetal fissure. They are, instead, notches in the equator of the lens due to an absence of zonular fibres. If there are insufficient zonules present the lens becomes subluxed. Marfan's syndrome is the most frequent cause of heritable lens dislocation, with intact and stretched, or focally absent zonules. We present a case of prominent lens notches that were otherwise asymptomatic, not leading to lens dislocation. This later turned out to be Marfan's syndrome.

Case report

A 36-year-old man presented to the eye clinic having been noticed by his optometrist to have bilateral lens changes. The patient had not noticed any visual problems. Ocular examination revealed unaided visual acuities 6/5 right and left. On mydriasis, lens changes were obvious (Fig. 1); there were no signs of uveal coloboma.

The patient had been reviewed when a child for chest wall asymmetry, thought to be insignificant. He was not known to be hypertensive. His father, however, had died at the age of 33 years of a sudden heart attack; no autopsy was performed at that time. Our patient, after a brief history of back pain, suffered acute back pain and collapsed and died when he was 44 years old.

An autopsy showed the cause of death to be a dissecting aortic aneurysm. Left ventricular hypertrophy was noted. Subsequent histological examination confirmed cystic medial necrosis in the wall of the aorta.

Comment

The true incidence of aortic dissection is unknown, as up to one-third of cases go undiagnosed but autopsy and population studies suggest an incidence of between 5 and 27 per million people per year.[1] The most important risk factors are untreated hypertension, advanced age and disease of the aortic wall.

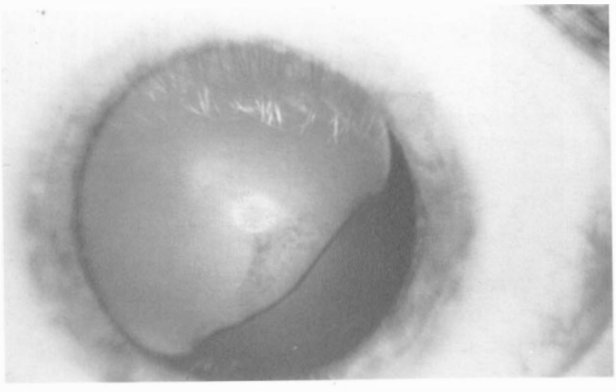

Fig. 1. *Photograh of the left eye showing lens notching.*