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.

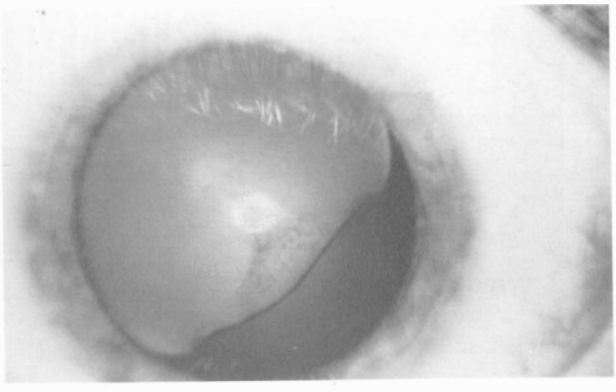

Fig. 1. *Photograh of the left eye showing lens notching.*

Marfan's syndrome is an autosomal dominant generalised disorder of connective tissue, with variable expression. There is also a variable biochemical picture; abnormal scleroproteins are produced, which perform poorly under tension, and urinary hydroxyproline is also increased, suggesting increased catabolism. The defective gene for some forms of Marfan's syndrome has been mapped to chromosome 15, band 21, the site of the fibrillin gene, the protein found in the microfibrillar system and in the zonules of the lens. The lesion seen in the aortic wall, termed 'cystic medial necrosis' by Erdheim, actually exhibits no cysts or necrosis, but the normal pattern of medial lamellar units is destroyed, the wall is thinned and pools of glycosaminoglycans are seen where the elastic structure of the wall is lost. This progressive change has also been identified as an ageing phenomenon which may be exaggerated by stress, such as hypertension.[2]

Many genotypes and phenotypes of Marfan's syndrome are being identified and it is likely that there is a spectrum of disorders. The skeletal phenotype shows considerable variation; not all patients are tall and thin. Clinically, for diagnosis, in the absence of one unequivocally affected first degree relative, involvement of the skeletal system must be present, together with the involvement of at least two other systems, with one major manifestation.[3] This patient had chest wall asymmetry noted, he had involvement of the cardiovascular and ocular systems, and had at least one major manifestation in his aortic dissection. Marfan's syndrome is therefore very likely to be his diagnosis. This conclusion has obvious implications for the counselling of his family, which has subsequently been arranged.

In conclusion, the clinician identifying lens notching or absent zonules should entertain Marfan's syndrome as a possible diagnosis.

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Sir,

CSR-like presentation in epidemic dropsy

Epidemic dropsy is an acute toxic disease caused by the consumption of mustard oil adulterated with *Argemone mexicana* oil.[1] This is due to the deliberate or inadvertent mixing of these two similar seeds during oil processing. The primary toxicity of argemone oil is due to the presence of the toxins sanguinarine and dihydroxy-sanguinarine.[2] These toxins block pyruvic acid metabolism, leading to an increase in blood levels of pyruvate – a potent vasodilator and endothelial toxin – increasing capillary permeability throughout the body.[3] The release of prostaglandins and histamine has also been suggested to play a significant role in the pathogenesis of dropsy-induced glaucoma.[4]

During an epidemic of epidemic dropsy in Delhi and adjacent states, we observed a central serous retinopathy (CSR)-like picture in two cases. To the best of our knowledge, this is the first report in the world literature of CSR related to epidemic dropsy.

Case reports

Case 1. A 40-year-old male tailor presented with sudden diminution of vision in both the eyes for 20 days. The patient had consumed a fresh stock of mustard oil sold loose, following which he reported severe headache, nausea, vomiting and diarrhoea, associated with bilateral severe swelling of both the lower limbs. He came from an area where a large number of people had suffered from epidemic dropsy and had been purchasing his cooking oil from the same sources. He was admitted to a local hospital and given intensive rehydration treatment for the acute condition. Although he stabilised systemically, he noticed a dimness of vision in the left eye, followed by the right eye after 3 days, and was referred to our centre.

On examination he had a vision of 3/60 in the right eye and counting fingers close to face in the left eye, with accurate projection. He had bilateral ill-sustained pupillary reactions. Fundus examination showed bilateral mild blurring of disc margins with temporal pallor and dull foveal reflex. The intraocular pressure (IOP) was 14 mmHg in both the eyes. The systemic examination was normal except for the presence of pedal oedema, which increased on exertion. The electrocardiogram showed clockwise electrical rotation of the inferior QRS axis. The echocardiogram was normal.

As toxic optic neuropathy was suspected the patient was given two doses of intravenous steroid pulse therapy: dexamethasone sodium phosphate 100 mg in 5% dextrose over 1 h daily. After two doses, the vision improved to 6/9 in both eyes. Automated perimetry (Humphrey central 30-2 threshold test) revealed a diffuse central field loss with a superior arcuate pattern field defect in the right eye and a patchy central scotoma in the left eye (Fig. 1).

On reviewing the fundus under dilation, we detected pale, diffuse subretinal lesions in both eyes in the macular areas. Fluorescein angiography showed focal leaks at the macula beginning in the arteriovenous phase, which enlarged in size in both the eyes, suggestive of a central serous retinopathy (Fig. 2).

A final diagnosis of epidemic dropsy with optic neuropathy with bilateral central serous retinopathy was made. At final follow-up at 3 months, the vision was