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

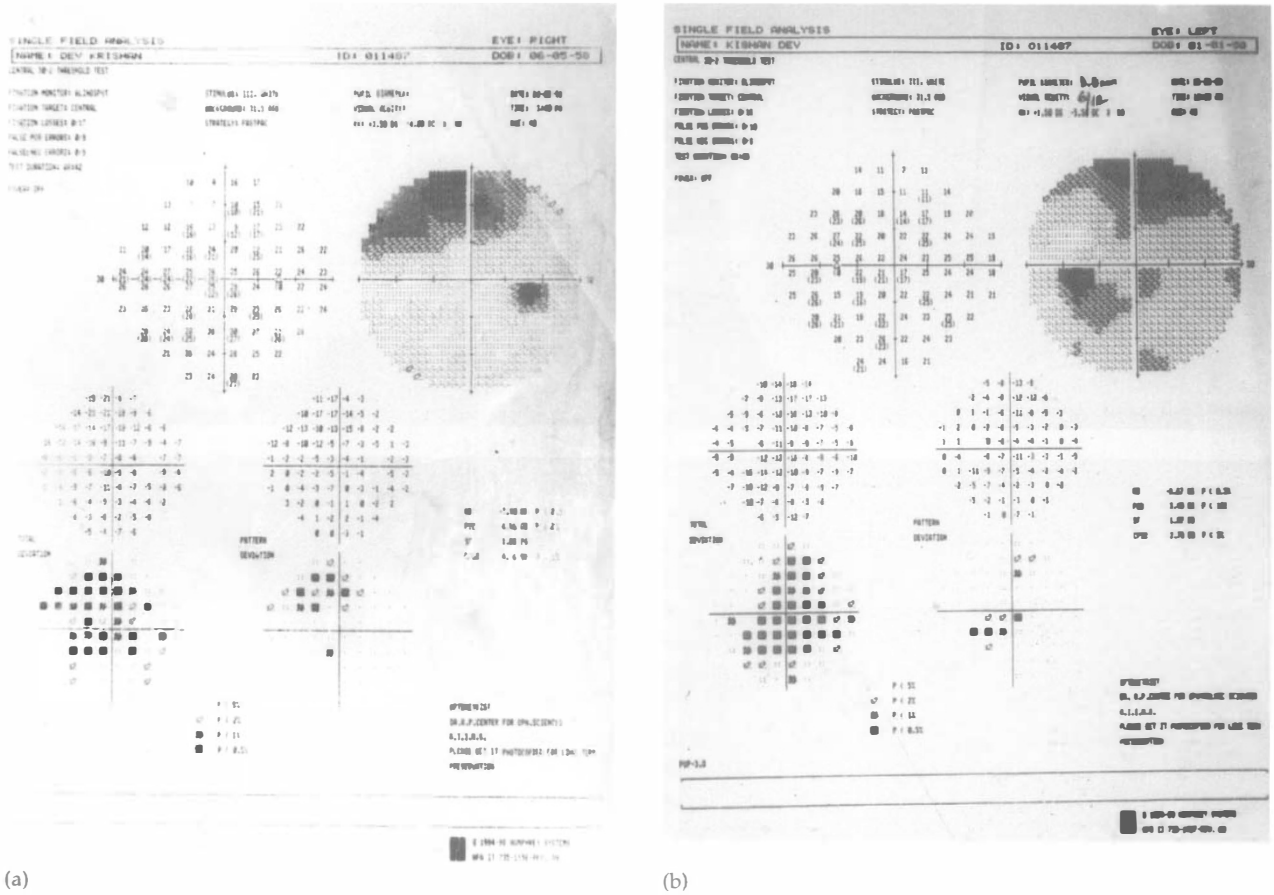
Case 1. A 40-year-old male tailor presented with sudden diminution of vision in both the eyes for 20 days. The patient had consumed a fresh stock of mustard oil sold loose, following which he reported severe headache, nausea, vomiting and diarrhoea, associated with bilateral severe swelling of both the 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



(a) (b)

Fig. 1. *Case 1. Humphrey central 30-2 threshold tested fields showing (a) right diffuse central field loss with a superior arcuate pattern field defect and (b) left central scotoma.*

stable at 6/6 with spectacles. The fundus examination showed only patchy pigment epithelial disturbance in both eyes.

Case 2. A 30-year-old male carpenter reported to our centre with complaints of sudden dimness of vision in both the eyes for 3 weeks. He had been suffering from pedal oedema for the past 6 weeks and had been diagnosed as having epidemic dropsy. His wife and 10-year-old son were similarly affected and they had all been treated with diuretics and multivitamins in a local hospital. Three weeks after the onset of the systemic symptoms of dropsy, he had developed blurred vision for which he consulted a private practitioner, who prescribed oral steroids. His vision deteriorated further after taking the steroids and he was referred to our centre.

On examination, his best corrected visual acuity was 6/18 in the right eye and 6/24 in the left eye. The IOP was 16 mmHg in both eyes. Ophthalmoscopy revealed disc hyperaemia, tortuous retinal vessels and focal central serous retinal detachments in both eyes. The visual fields showed patchy central field defects. The fluorescein angiogram showed bilateral macular leaks, suggestive of CSR (Fig. 3). There was also a focal area of perivenous leakage involving a superotemporal venule in the left eye. The patient was given only supportive treatment with an oral multivitamin-antioxidant combination daily. Three weeks later on routine follow-up examination, the IOP was high: 28 mmHg in the right eye and 26 mmHg in the left eye. The IOP responded to topical timolol 0.5% eye drops twice daily. Over a period of 2–3 weeks the vision improved to 6/9 in both eyes. Repetition of the fluorescein angiogram revealed no leakage of dye with patchy window defects of the retinal pigment epithelium.

Comment

The clinical presentation of epidemic dropsy is usually with gastrointestinal upset in the form of nausea, vomiting, diarrhoea and low-grade fever. This is associated with a characteristic bilateral non-inflammatory swelling of both the lower limbs, with rashes, erythema and telangiectasias. High-output congestive heart failure is the most common cause of death in these patients. Ocular manifestations include glaucoma,[3–5] engorgement and tortuosity of retinal veins,[5,6] retinal haemorrhages,[5,6] papillophlebitis,[7] disc oedema[8] and central retinal vein occlusion.[7–9]

During August and September 1998 there was a severe outbreak of epidemic dropsy in Delhi, in which approximately 3000 cases were affected and 67 died (according to press reports). Random testing of the oil samples revealed an unprecedented high level of contamination. New features reported during this epidemic were soft exudates, extensive haemorrhages (macular, subhyaloid, vitreous) and fibrous bands near

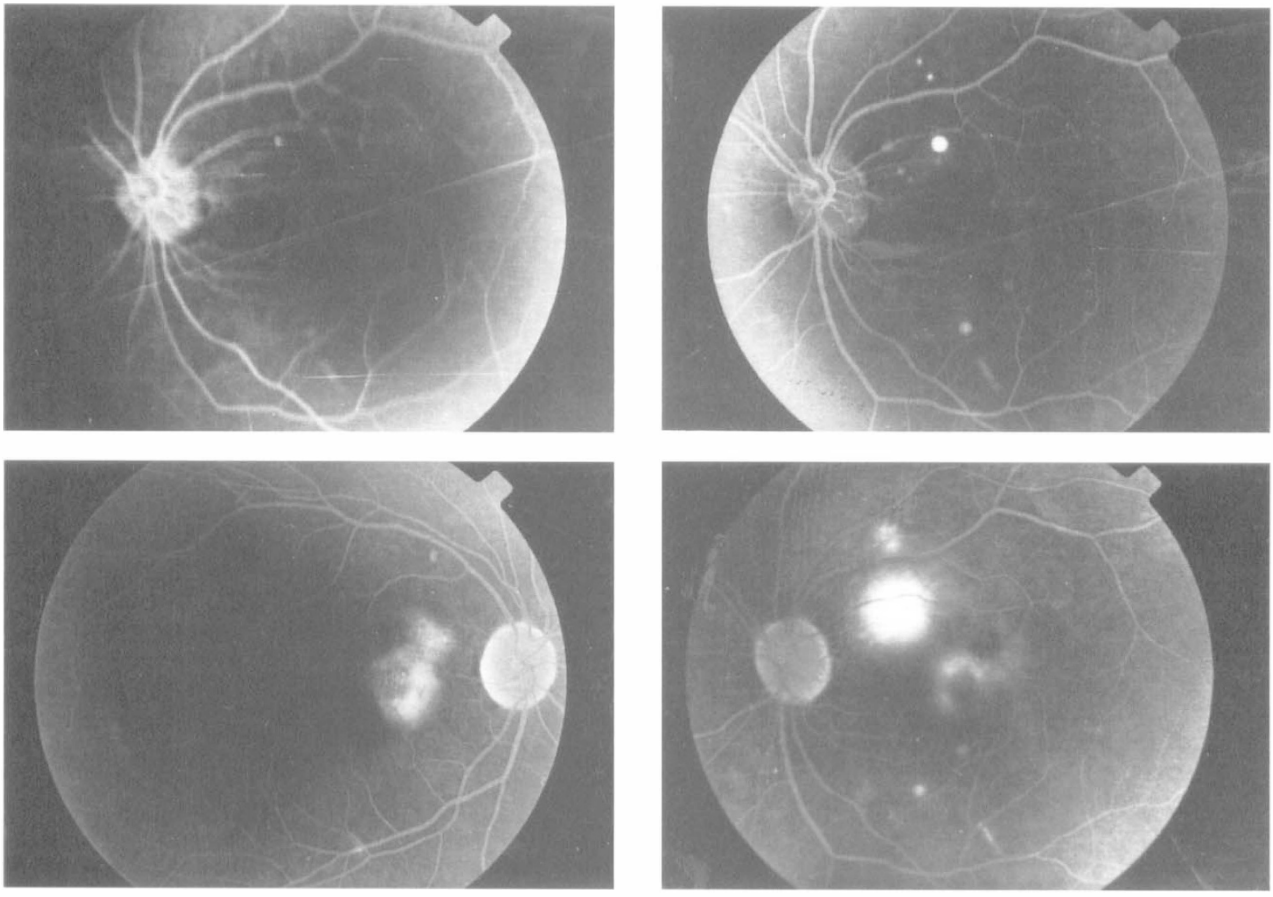
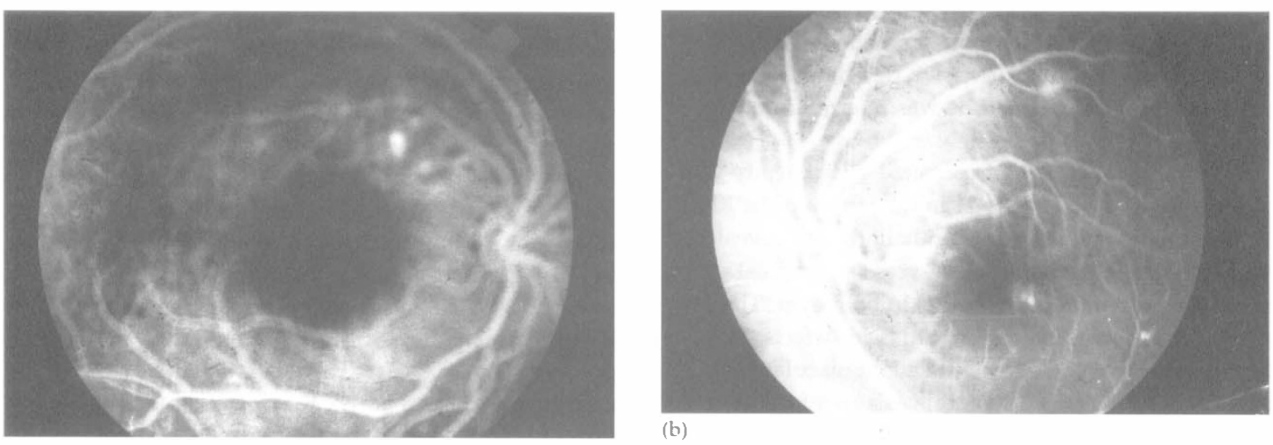


Fig. 2. *Case 1. Fluorescein angiogram (clockwise from top left). Serial photographs show multiple ink blot leaks in the left macula and the late-phase picture of the right eye shows an area of diffuse leak involving the papillomacular bundle.*

the disc.[10] The two cases reported here represent the first description of fluorescein angiographic features resembling CSR.

An increased permeability at the retinal pigment epithelium–choriocapillaries level due to a toxic breakdown of the blood–aqueous barrier induced by the sanguinarine compounds is the most likely mechanism. Another possibility is that the CSR-like picture was either precipitated or aggravated by the use of steroids in both cases. The role of steroids in the pathogenesis of CSR is well known.[11] The optic neuropathy seen in the first case could have been toxic, as bilateral involvement was noticed very soon after the onset of severe systemic disease. Improvement of vision to some extent with intravenous steroids supports the possibility of reversible inflammation or toxin-induced optic neuropathy.

The toxicity of argemone oil is known to be dose dependent and cumulative,[12] therefore a higher level of oil contamination in this epidemic may account for these two new features having manifested for the first time, namely optic neuropathy and CSR.



(a) (b)

Fig. 3. *Case 2. Fluorescein angiogram demonstrating (a) ink blot leak in the right superonasal macula and (b) two focal ink blot leaks in the left macula. An area of perivenous leakage involving the left upper temporal venule is also evident.*

The treatment is to discontinue the consumption of the adulterated mustard oil and give supportive therapy with diuretics, vitamins, anti-histaminics and anti-prostaglandin agents. The inadvertent use of systemic steroids in this instance probably contributed to the CSR-like lesions. Systemic steroids are not routinely given to control the acute phase of epidemic dropsy. This report highlights the potential problems of using oral steroids in these patients. It is also essential to screen other non-symptomatic members (who consumed the oil) for glaucoma and to continue IOP monitoring over a period of 2 months for the exposed population.

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Sir,

One-and-a-half syndrome: a different type

In 1967, Fisher[1] first described two patients with clinical disorder of extraocular movements characterised by a conjugate gaze paresis on attempted gaze to the lesion side and impaired adduction of the ipsilateral eye (an internuclear ophthalmoplegia) on attempted gaze to the contralateral side (one-and-a-half syndrome). Since then, similar cases have been reported in the literature and have confirmed the specificity of this syndrome.[2–6] We describe here a different type of one-and-a-half syndrome in a 61-year-old man in whom the preserved eye movement was adduction.

Case report

A 61-year-old man was admitted to the hospital because of gradual progression of marked weakness and hypoesthaesia of the right side. The patient was well until 2 days earlier, when he experienced the onset of weakness and hypoesthaesia of the right side that made it impossible for him to walk. Cranial computed tomography (CT) findings were normal. Two days after the scan he became right hemiplegic, and he complained of diplopia. There was a 6-year history of hypertension, which was managed with enalapril maleate (10 mg/day).

On physical examination, blood pressure was 180/120 mmHg, heart rate was 84/min and his body temperature was 36.5 °C. General physical examination was normal. On neurological examination the patient was fully oriented and his speech was fluent. There was 3 mm of ptosis of the left eyelid. Fundus examination found no papilloedema bilaterally. His pupils were anisocoric (right: 2.5 mm, left: 4 mm) and pupillary reflex was absent on the left side. Decreased corneal reflex on the right side was established. Horizontal gaze to the right showed a total conjugate gaze paresis. On left gaze there was paresis of abduction of the left eye, although the right eye did adduct with gaze-evoked horizontal jerk nystagmus (Fig. 1). Vertical eye movements and vestibulo-ocular responses were intact in right eye and his other cranial nerve functions were normal. Visual fields demonstrated a right homonymous hemianopia.

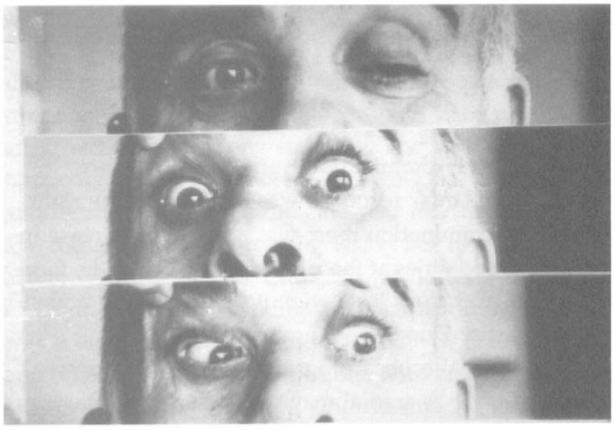

Fig. 1. *Top: Note marked ptosis of the left eye in the primary position of gaze. Centre: Horizontal conjugate gaze palsy looking right. Bottom: On left gaze there was paresis of abduction of the left eye, although the right eye did adduct.*