PostScript

LETTERS

Transient macular dysfunction determined by focal macular electroretinogram

Rapid diagnosis of patients with acute visual loss is critical[1] but is difficult if the retina appears normal ophthalmoscopically. We report the case of a patient who presented with acute unilateral visual loss and a central scotoma.

Case report

A 75-year-old man complained of a sudden and painless decrease of vision in his left eye. He had undergone surgery for an unruptured intracranial aneurysm 20 years earlier and was taking 7 mg/day of systemic prednisolone for rheumatoid arthritis. He had also had diabetic mellitus without retinopathy for 5 years. He was being followed for a left hemianopsia and normal tension glaucoma for the previous 3 years.

On examination, a left relative afferent papillary defect (RAPD) was observed, and visual acuity (VA) was 20/30 OD and 20/2000 OS. All of the ocular findings were normal except for enlarged disc cupping OU (fig 1). Fluorescein angiography showed a delay in the arm-to-retina circulation time of 20.0 s (fig 1). Neither retinal emboli nor localised filling delay were observed. Goldmann perimetry showed a left quadrantic homonymous hemianopsia and a small central scotoma with peripheral constriction. The alterations in the left eye were new (fig 1). Focal macular electroretinograms (FMERGs) were recorded 2 h after onset as described (see the supplemental figure available at http://bjo.bmj.com/supplemental).[2 3]

The FMERGs were decreased in the left eye (fig 2) indicating that the visual dysfunction was retinal in origin. Left VA improved to 20/250 spontaneously 3 h later. On the following day, VA had improved to 20/30, and the amplitudes of the FMERGs, full-field ERGs and pattern visual evoked response (VEP) were normal (fig 2). The visual field obtained 3 months later showed that the central scotoma was not present and an expansion of the peripheral visual fields. The fundus remained normal. Ultrasound echography revealed no stenosis of the carotid artery. Blood examination revealed rheumatoid factor and high HbA1c but was otherwise normal.

We conclude that the acute visual loss and central scotoma with reduced FMERGs were consistent with transient macular ischaemia, and prophylactic anti-coagulation treatment was considered.

Comments

Our patient had an acute monocular visual loss and a fundus that appeared normal except for the relatively delayed angiographic retinal filling time and enlarged disc cups. The RAPD, central scotoma and systemic complications made it difficult to determine the site of the alterations. The reduced FMERGs pointed to the retina as the site.

However, an abnormally long-lasting visual decrease is not typical for amaurosis fugax,[1] and ophthalmoscopy did not show retinal oedema typical of arteriolar occlusion. Conventional electrophysiological examinations such as full-field ERGs and VEPs might be useful except when the ischaemic site is in the macular region.

The clinical course in our case was compatible with a transient retinal ischaemia with a possibility of a transient central retinal artery occlusion, although additional more generalised abnormalities cannot be completely excluded. The FMERG within the 5° area is similar to that of the ERG of a monkey treated with 2-amino-4-phosphonobutyric acid (APB) and cis-2,3-piperidine dicarboxylic acid (PDA) to suppress both on and off synapses.[4 5] This implies that the inner retinal layers have serious dysfunction. Because of the systemic complications, the risk of utilising prophylactic anticoagulant agents was discussed. However, the VA and central scotoma quickly recovered accompanied by an improvement in the FMERGs without any intervention.

These findings indicate that clinicians should consider focal macular dysfunction in cases of acute vision loss and normal retinal

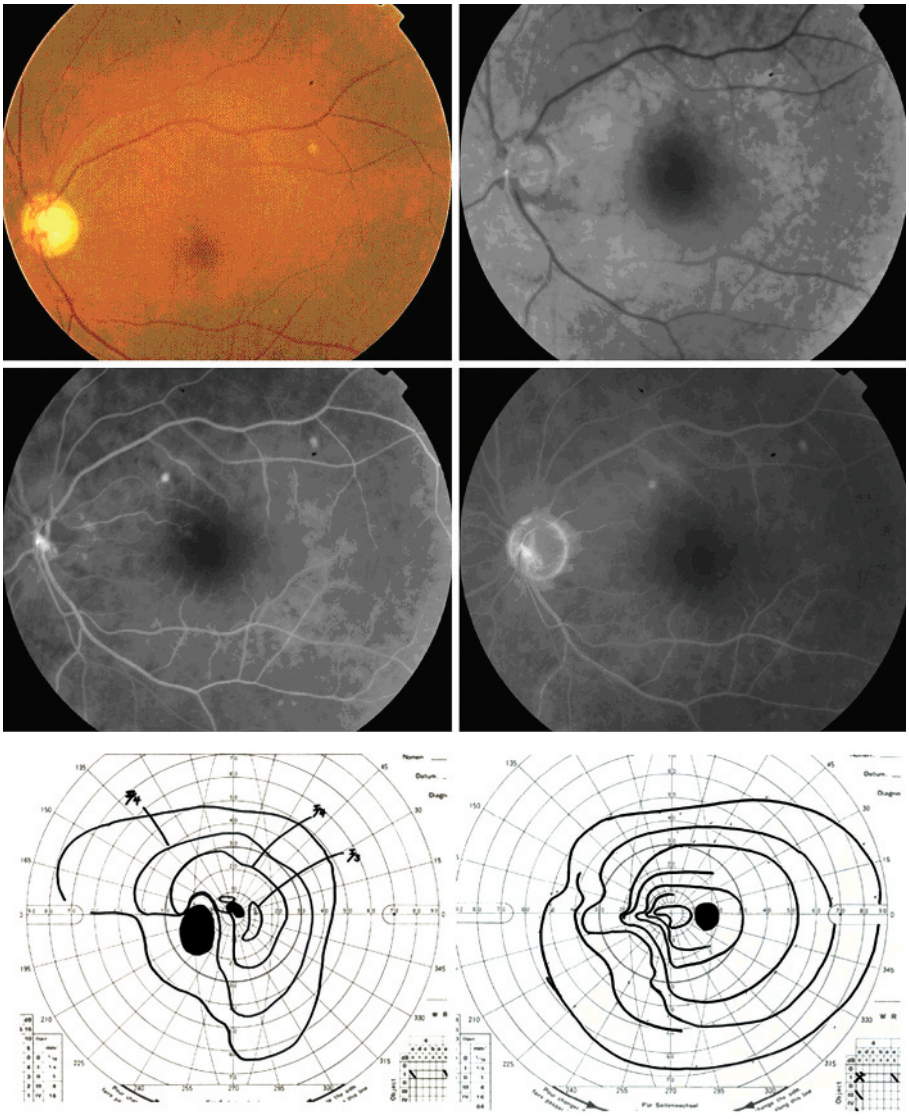

Figure 1 Fundus photograph and fluorescein fundus angiograms of the patient's left eye. Top left: Fundus photograph showing enlarged cupping of the left optic disc. Top right: Fluorescein angiogram with delayed arm-to-retina circulation time. Second left and right: Fluorescein angiogram showing neither localised arteriolar filling delay nor retinal emboli in the middle (left) and late (right) phases. Third left: Goldmann visual field in the acute phase shows small central scotoma and peripheral constriction in addition to a decrease in the isopters in the left temporal inferior area (quadrantic homonymous hemianopsia) which had existed for years in the left eye. The central scotoma and peripheral constriction were new. Visual acuity (VA) was 20/2000 OS. Third right: Goldmann visual field performed 6 months earlier showing a decrease in the isopters in the left nasal inferior area (homonymous hemianopsia) in the right eye. VA was 20/30 OD.

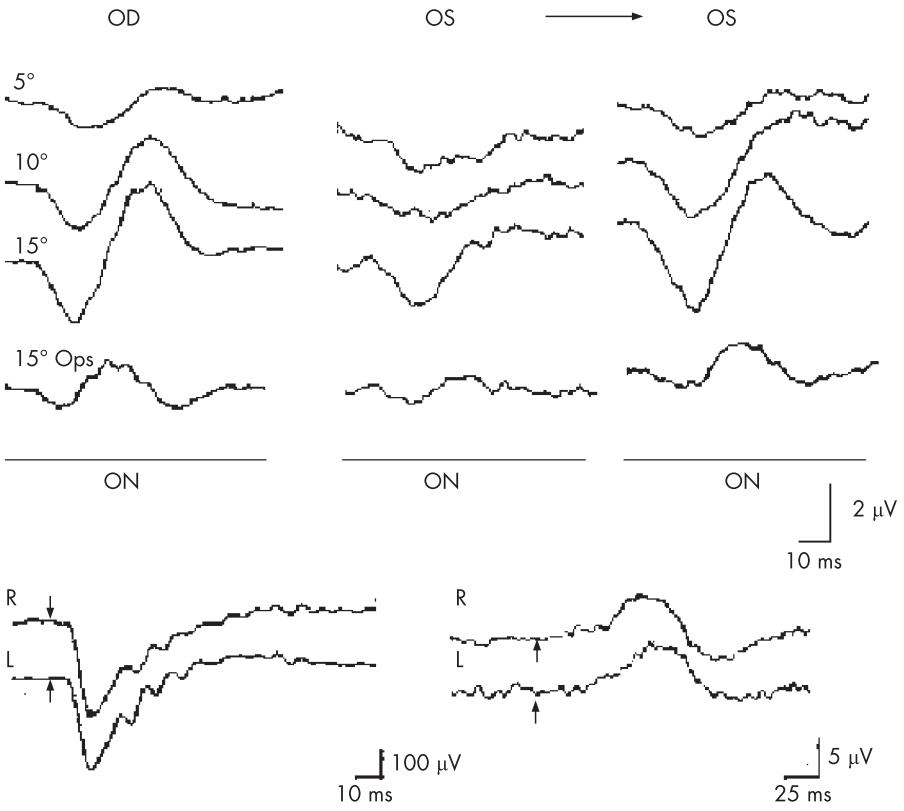

Figure 2 Focal macular electroretinogram (FMERG, top), full field electroretinogram and visual evoked response (VEP) from both eyes (bottom). Top: Upper three recordings show the photopic a- and b-waves, and lowest recordings show the oscillatory potentials (Ops). The stimulus spot size was 5°, 10° and 15° as indicated in the figure. Left and centre: FMERGs recorded during the acute phase. The ERGs of the left eye (centre) are reduced compared with those of the right eye. Right: FMERGs recorded on the following day showing the recovery of all components of the left eye. The a-wave is not affected as much as the b-waves and Ops. Bottom left: Bright-flash ERG showed no significant difference in the two eyes even in the oscillatory potentials which are sensitive to retinal ischaemia. Bottom right: The VEPs showed no significant difference between the two eyes. The stimulus onset of full field ERG and VEP is indicated by arrows.

appearance; multifocal ERGs or FMERGs are useful in determining the site of the pathology.

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Naoki Terauchi, Kaoru Fujinami, Kei Shinoda, Kazushige Tsunoda, Gen Hanazono, Yozo Miyake

Laboratory of Visual Physiology, National Institute of Sensory Organs, National Hospital Organization, Tokyo Medical Center, 2-5-1 Higashigaoka, Meguro-ku, Tokyo 152-8902, Japan

Koichi Inomata

Department of Ophthalmology, School of Medicine, Nihon University, 1-8-13 Surugadai, Kanda, Chiyoda-ku, Tokyo 101-8309, Japan

Correspondence to: Dr Kei Shinoda, Laboratory of Visual Physiology, National Institute of Sensory Organs, National Hospital Organization, Tokyo Medical Center, 2-5-1 Higashigaoka, Meguro-ku, Tokyo 152-8902, Japan; shinodakei@kankakuki.go.jp

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The supplemental figure is available at http://bjo.bmj.com/supplemental.

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In-vivo scanning of Ascher intrastromal corneal ring opacity

Case report

An 80-year-old man was seen with abnormal corneal appearances. He was asymptomatic with

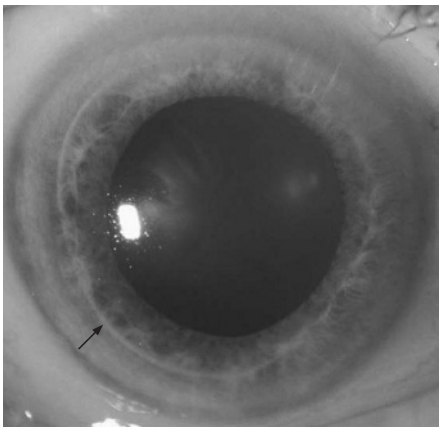

Figure 1 Right eye: intrastromal corneal ring 0.1 mm in width, 7.8 mm in vertical diameter.

no previous ocular, medical or drug history. Visual acuities were 6/6 in both eyes unaided; with ocular examination revealing bilateral 0.1 mm wide white intrastromal corneal ring opacities 7.8 mm and 7.7 mm in diameter in the right and left eyes, respectively (fig 1). Corneal sensation was normal with no evidence of thinning, scarring or vascularisation. The remaining ocular examination was normal. Blood tests for lipid, protein and autoimmune markers were normal. Corneal pachymetry and topography was unremarkable. Cross-sectional imaging of the cornea with anterior segment optical coherence tomography (Visante OCT, Carl Zeiss Meditec, Inc., USA) revealed opacification affecting the entire thickness of the stroma (fig 2). Confocal microscopy (HRT II/Rostock Cornea Module; Heidelberg Engineering GmbH, Germany) of the ring opacity revealed microdot extracellular deposits within the corneal stroma (fig 3). The epithelium and endothelium of the cornea appeared normal. Ocular examination of the patient's sister was unremarkable, with another sister unavailable for examination. There has been no progression with over 12 months follow-up with the patient remaining asymptomatic.

Comment

Originally reported by Ascher[1] in 1963, intracorneal rings are rare;[2–8] occurring both unilaterally

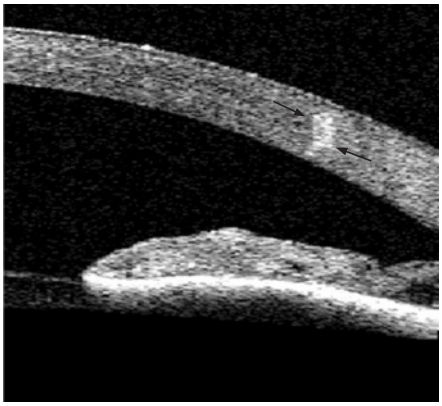

Figure 2 Optical coherence tomography of left cornea: highly reflective deposits are seen throughout the entire thickness of the corneal stroma.