Sir,

Acute annular outer retinopathy with systemic symptoms

Acute annular outer retinopathy (AAOR)[1,2] is a condition of unknown aetiology, which is considered by many to be a variant of acute zonal occult outer retinopathy (AZOOR).[3] This condition typically affects young adults and causes photopsia, scotoma, and electroretinographic abnormalities; it is rarely associated with systemic symptoms.

Case report

A 45-year-old woman presented with a 2-day history of right temporal photopsia in dim illumination and scotoma in bright illumination. Visual acuity—with a myopic correction—was 6/7.5 with the right eye and 6/6 with the left eye. Fundoscopy revealed a grey-white peripapillary deep retinal demarcation line in the right eye, which roughly corresponded to a scotoma on Goldmann perimetry (Figure 1). Subtle peripapillary hyperfluorescence was observed on fluorescein angiography and fundus autofluorescence using the Heidelberg Retinal Angiograph 2 revealed hyperfluorescent spots in the same region (Figure 2).

The patient was reviewed 5 days later, by which time she had symptoms of meningism and right-sided hearing loss. Her scotoma had subjectively expanded, but had become 'patchy'. Examination revealed extension of the retinal demarcation line and a new isolated

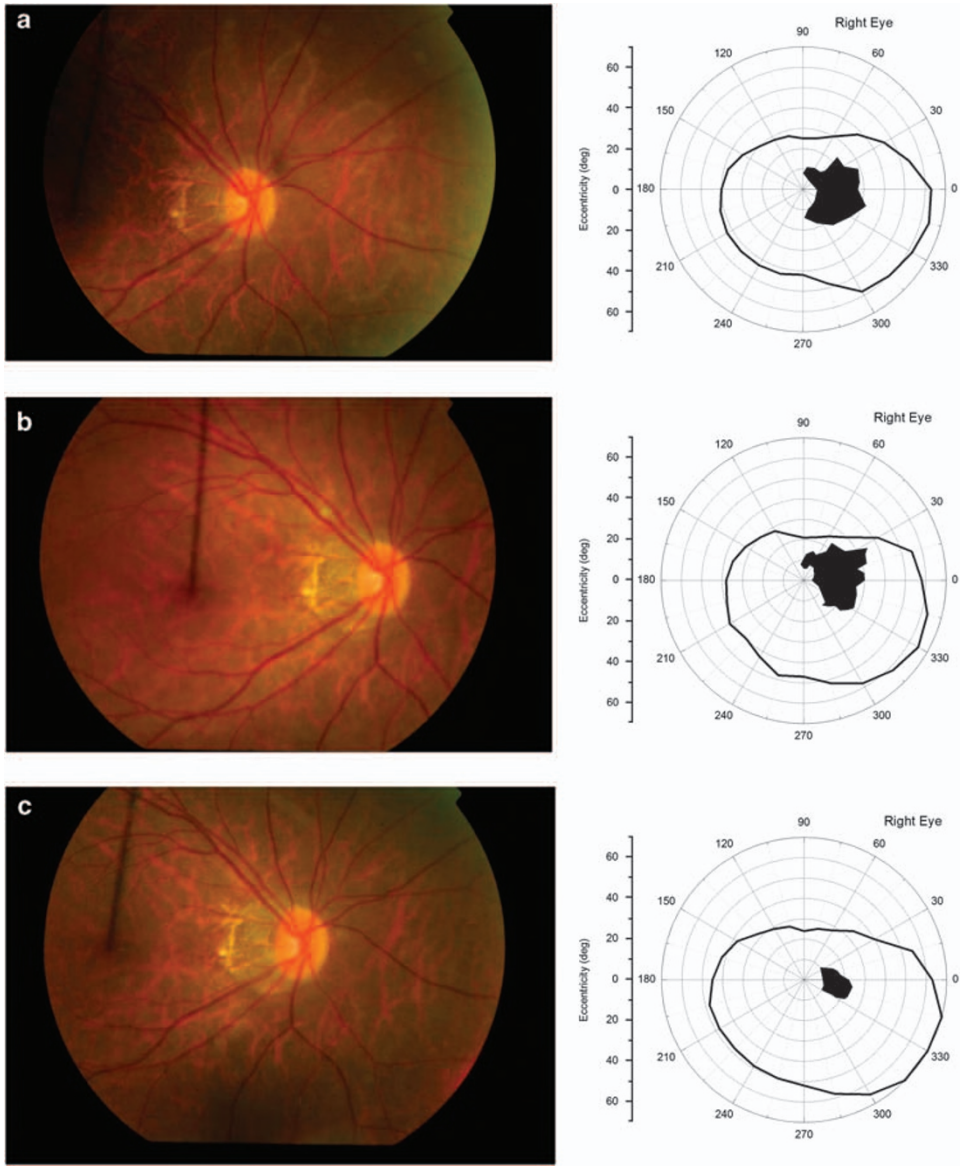

Figure 1 Fundus appearance (left) and the corresponding visual field (right) to a Goldmann Size I, intensity 4e stimulus. (a) At presentation, where there is a prominent demarcation line. (b) At 7 days, by which time the demarcation line had spread and become less prominent and a new cotton–wool spot (superior temporal to the disc) was observed. (c) At 4 weeks. The outer black lines of the field plots represent isopter lines, while the filled lines temporal to fixation represent the area of scotoma.

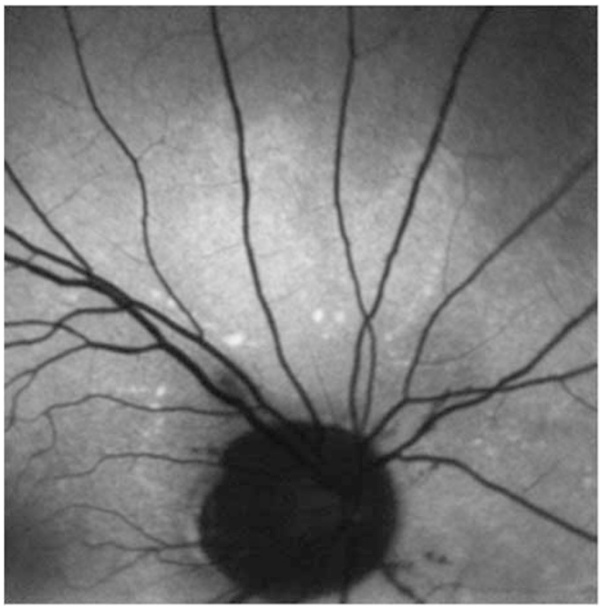

Figure 2 Fundus autofluorescence showing hyperfluorescence in the peripapillary area.

cotton–wool spot. Goldmann perimetry confirmed a change to the extent of the scotoma (Figure 1). Outer retinal involvement was suggested by the decreased multifocal electroretinogram amplitudes in the affected portion of the right visual field. She was referred for formal neurological and ENT assessment; neither revealed any further findings that were thought to be acute. Extensive laboratory testing revealed that she was anti-nuclear antibody positive (titre 1:80); there was no serological evidence of recent infection with HSV, HZV, EBV, CMV, Coxsackie virus or echovirus, and her VDRL was negative. Resolution of her fundus lesions occurred by day 31, with a corresponding contraction of her scotoma (Figure 1). Her systemic symptoms resolved after 2 weeks.

Comment

AAOR shares many common features with AZOOR:[3] the two are distinguished by fundoscopy—AAOR is characterised by annular deep retinal grey–white spots in the peripapillary area, whereas AZOOR causes no fundus changes early in the disease. AAOR is usually unilateral and visual acuity can vary from normal to hand movements. The collective evidence—gleaned from electrophysiology, high-resolution optical coherence tomography,[4] and indirect immunohistochemistry[5]—suggests outer retinal involvement. Autofluorescence shows the deposition of autofluorescent material in the peripapillary fundus.[6] The aetiology of AAOR remains unknown, although an autoimmune mechanism has been suggested.[2] Our case is noteworthy for several reasons—the presence of temporally associated systemic symptoms, positive serology for anti-nuclear antibodies, the observation of a cotton–wool spot (suggesting a retinal vascular component to the disease), and the remarkably rapid speed of recovery.

Conflict of interest

The authors declare no conflict of interest.

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