Multimodal imaging of acute zonal occult outer retinopathy in a Indian male

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Key words: Acute zonal occult outer retinopathy, multimodal imaging, white dot syndrome

Case Report

A 24-year-old Indian male presented with a temporal field defect in his right eye (RE) since 1 month. There was no associated photopsia, redness, pain, and no preceding viral illness. Unaided visual acuity (UAVA) was 6/6 in both eyes (BE). Anterior segment examination, pupillary reflexes, and fundus examination of the left eye (LE) were unremarkable. Fundus examination of the RE revealed a hypopigmented amoeboid lesion in the peripapillary area extending upto the nasal margin of the fovea [Fig. 1]. RE fundus autofluorescence (AF) showed stippled hypo- and hyper AF with a hyper AF transition zone between the involved and uninvolved retina [Fig. 2a]. Humphrey's visual field (VF) testing revealed a temporal hemianopic defect in the RE [Fig. 2b]. Spectral domain optical coherence tomography showed absence of ellipsoid and interdigitation zones in the area of the lesion with normal inner retinal and choroidal architecture [Fig. 3a]. While fluorescein angiography showed transmission fluorescence [Fig. 3b]. Multimodal imaging (MMI) of the LE was normal. Clinical diagnosis of RE acute zonal occult outer retinopathy (AZOOR) was made and the patient was subjected to full-field electroretinogram (ERG). RE ERG showed increased latency and significantly reduced amplitude of 30 Hz flicker response, suggestive of generalized cone dysfunction. The patient refused to take oral steroids, and at 6 months follow-up, he was maintaining a UAVA of 6/6 with no change in MMI.

Discussion

AZOOR was first described by Donald Gass as acute onset VF defect with or without photopsias due to outer retinal dysfunction.[1] It is an idiopathic disorder whose demography, clinical features, and outcomes are different in Asians when compared with Caucasians.[2] Asian patients have lesser retinal pigment epithelium (RPE) atrophy, more chances of spontaneous resolution, and better visual acuities.[2,3] Most of the data on AZOOR in Asians are from China and Japan.[2,4] There is just one case report from India in which a polycythemia vera patient developed AZOOR-like features.[5] About 76% of AZOOR patients have no findings on fundus examination at presentation; hence, MMI is invaluable to clinch

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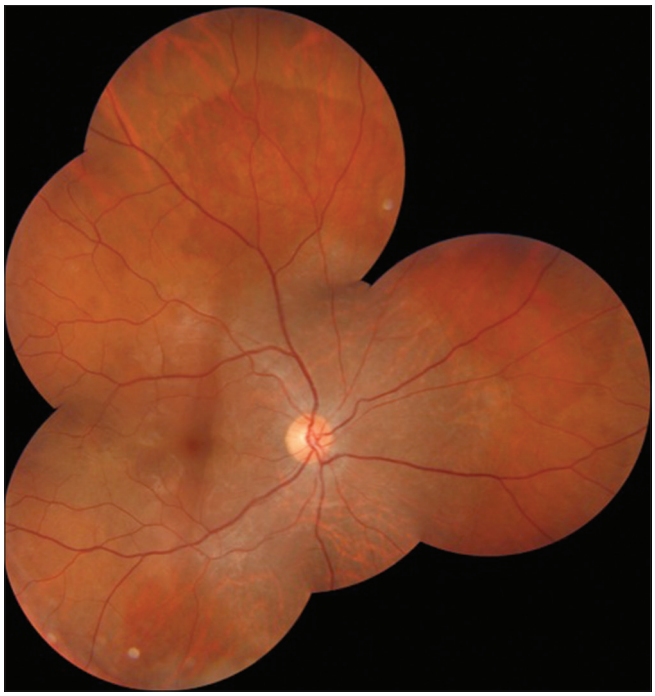

Figure 1: Montage fundus photograph of the right eye showing media clarity grade 1 with a circumpapillary hypopigmented lesion, suggestive of RPE atrophy

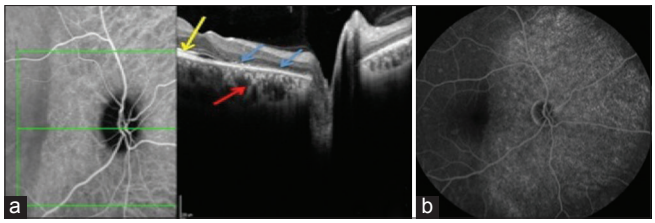

Figure 3: (a) Spectral domain OCT of the right eye showing disruption of the ellipsoid zone and interdigitation zone in the area corresponding to the lesion (blue arrows) with normal inner retinal (yellow arrow) and choroidal architecture (red arrow); (b) late-phase fluorescein angiogram of the right eye showing transmission fluorescence

the diagnosis.[3] MMI also helps differentiate early stages of AZOOR from autoimmune retinopathy, carcinoma-associated retinopathy, toxic retinopathy, serpiginous choroiditis, and acute idiopathic blind spot enlargement. Younger age of onset and zonal retinal dysfunction help distinguish AZOOR from other entities.[2,3]

This case conveys two important findings – first, Indian patients with AZOOR may have slow progression of this disease compared with Caucasians. Second, MMI is essential to diagnose and monitor AZOOR.

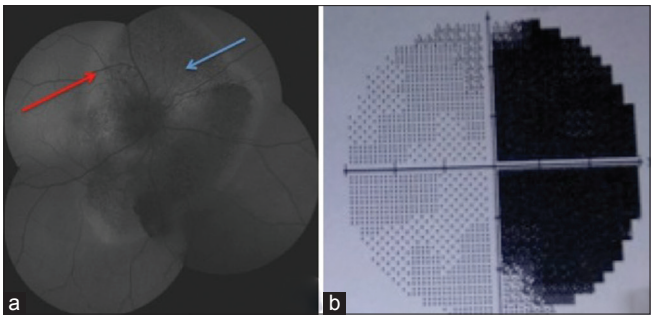

Figure 2: (a) Short-wave autofluorescence of the right eye showing speckled hypo- and hyper autofluorescence (blue arrow) with a hyper autofluorescence transition zone (red arrow); (b) Humphrey's automated visual field grey-scale printout of the right eye showing temporal hemianopic field defect

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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