Multimodal imaging signatures in a case of acute zonal occult outer retinopathy

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Acute zonal occult outer retinopathy (AZOOR) is a retinal disease characterized by a slow onset loss of retinal function with minimally evident fundus changes. Patients with AZOOR present with initially progressive scotoma and photopsia. Its pathogenesis has not been definitively determined as of yet. Characteristically, the extent of the visual field defect is unexplained by fundus examination, but there is marked retinal dysfunction, which is evident on multimodal imaging and electrophysiological testing. We herein describe multimodal imaging signatures of AZOOR, in a patient of Indian origin, highlighting the hitherto unreported multicolor channels and near-infrared autofluorescence.

Key words: Acute zonal occult outer the retinopathy, multimodal, near-infrared autofluorescence

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Acute zonal occult outer retinopathy (AZOOR) is a retinal disease characterized by a slow onset loss of retinal function with minimally evident fundus changes.[1] Although infectious and autoimmune mechanisms have been proposed as the cause of AZOOR, its pathogenesis has not been definitively determined.[2] Characteristically, the extent of the visual field defect is unexplained by fundus examination, but there is marked retinal dysfunction, which is evident on imaging and electrophysiological testing. We herein describe multimodal imaging signatures of AZOOR, in a patient of Indian origin, highlighting the hitherto unreported multicolor channels and near-infrared autofluorescence (IR-AF).

Case Report

A 31-year-old male patient presented to a tertiary eye center in Western India with complaint of gradual painless decreasing vision in both eyes, predominantly in the right eye, for the past 2 years. The patient was previously diagnosed elsewhere with vasculitis before 2 years and was treated for the same with oral steroids, of which no details were available. The patient discontinued the treatment after 2 months by himself and was on no treatment since then. Best-corrected visual acuity in the right and left eyes were 6/36, N12 and 6/9, N6, respectively. IOP was 14 mmHg in both eyes. The anterior segment was unremarkable. Dilated fundus evaluation [Fig. 1] showed areas of peripapillary retinal pigment epithelium (RPE) atrophy along with peripheral bony spicule pigmentation in both eyes. Multicolor imaging [Fig. 1] modality highlighted the areas of atrophy. The infrared reflectance channel [Fig. 2] revealed the areas of atrophy, while the green and blue reflectance channels [Fig. 2], which draw attention to the

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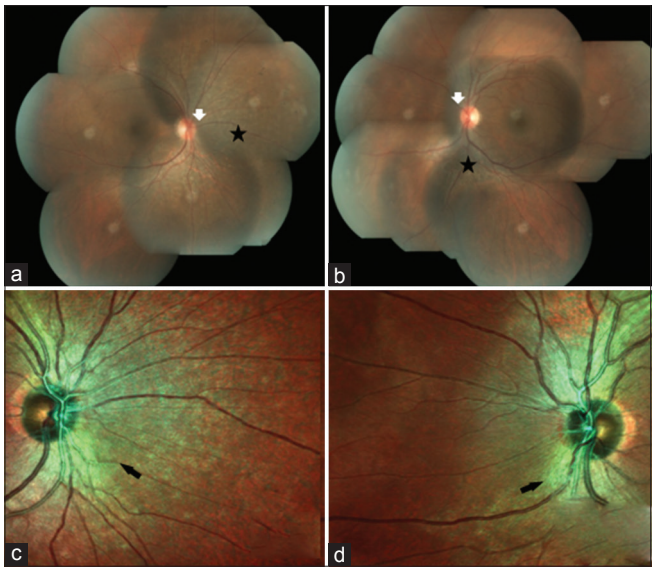

Figure 1: Color fundus montage (a and b) showing area of peri papillary atrophy (white arrow) and bony spicule pigmentation (star) in both eyes; multicolor imaging (c and d) highlighting the area of atrophy (black arrow)

superficial retinal lesions, were relatively silent. Fundus autofluorescence (FAF) [Fig. 3] of the right eye showed a hyperautofluorescent patch with a distinct margin surrounding the disc and involving the posterior pole, whereas the left eye showed a hyperautofluorescent patch with a distinct margin mainly surrounding the disc while sparing the macula. The IR-AF channel [Fig. 3] in both eyes highlighted the boundaries of the hyperautofluorescent patch and revealed multiple dark spicules nasal, inferior and superior to the disc. The spectral domain optical coherence tomography (SD-OCT) [Fig. 4] showed foveal thinning in the right eye along with the loss of external limiting membrane (ELM) and ellipsoid zone in the parafoveal region in both eyes, along with the presence of multiple hyperreflective foci in the outer retina and inner choroid. Fundus fluorescence angiography showed window defects with no leaks, and indocyanine green angiography showed no abnormality. The Humphrey visual field 30-2 showed an enlarged blind spot and temporal scotoma in both eyes, not respecting the horizontal meridian. Both eyes full-field electroretinographic (ERG) showed reduced photopic and scotopic response, right eye more than the left eye. With the above multimodal imaging signatures, a diagnosis of both eyes AZOOR was made more definitively. The patient was referred to a physician for steroid clearance for commencing further treatment.

Discussion

AZOOR is a rare retinal disease, especially in the Indian subcontinent, marked by photopsia and scotoma with subtle fundus changes in young myopic females.[1,3] It was first described by Gass, who described it as minimal fluorescein angiographic changes, loss of zones of outer retinal function, irreversible ERG abnormalities, and permanent visual field loss.[1,3] Late disease changes include photoreceptors atrophy, narrowing of arterioles, and pigment migration in a bone spicule pattern.[1,3,4] The study of Mrejen *et al*. described a zonal

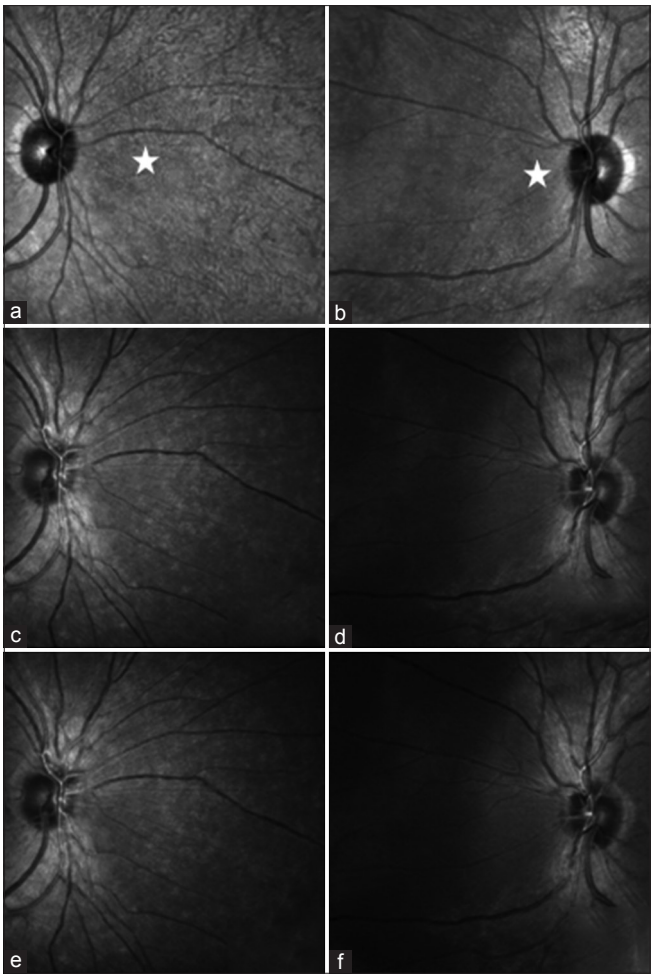

Figure 2: Infrared reflectance image (a and b) highlighting the area of peri papillary atrophy (star); while the green reflectance (c and d) and blue reflectance (e and f) showing no apparent changes

classification of AZOOR based on multimodal imaging.[5] Similar trizonal distribution is seen in our case, with additional novel features of near IR-AF and multicolor reflectance channels.

FAF is a noninvasive retinal imaging modality used in clinical practice to provide a density map of lipofuscin, the predominant ocular fluorophore, in the RPE.[5,6] In our case, FAF showed normal autofluorescence in the area outside of the demarcating line (zone 1), speckled hyperautofluorescence within the lesion (zone 2), and hypo auto fluorescence areas which corresponded to the RPE atrophy (zone 3). Such a trizonal distribution is distinctly illustrated in literature for AZOOR.[5]

Near IR-AF is a noninvasive imaging technology that provides information on the distribution of melanin within the retinal pigment epithelial cell/choroid complex.[7] The IR-AF channel, hitherto unreported for AZOOR lesion in literature, clearly delineated the margin of the lesion and revealed the typical pigment migration in a bony spicule pattern. Near IF AF is a hitherto unreported imaging modality used in AZOOR, which can facilitate in clearly marking the advancing edge of the lesion and guide regarding the progression of the disease.

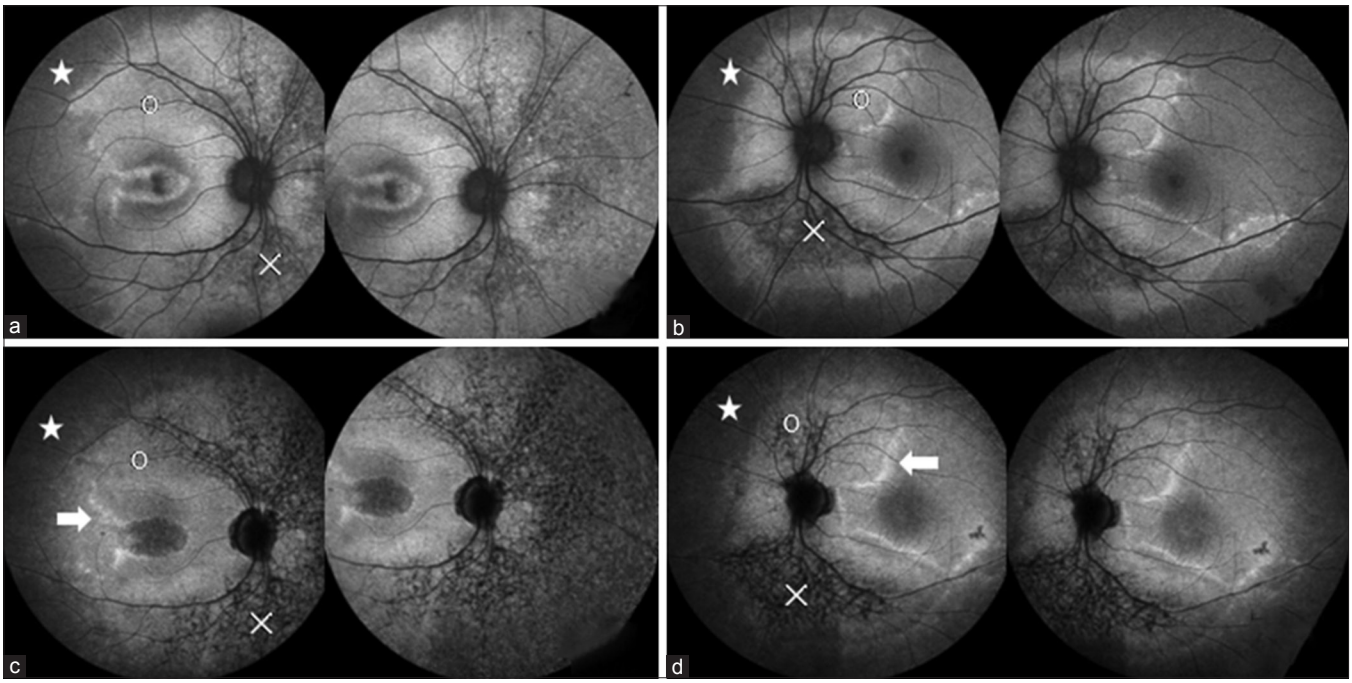

Figure 3: Blue fundus autofluorescence imaging both eyes (a and b) showing normal auto fluorescence in the area outside of the demarcating line (zone1-star), speckled hyperautofluorescence within the lesion (zone 2 - circle), and hypo auto fluorescence areas which corresponded to the retinal pigment epithelium atrophy (zone 3 - cross); infrared autofluorescence channel (c and d) clearly delineates the margin of the lesion (white arrow) also demonstrates the typical pigment migration in a bony spicule pattern (cross)

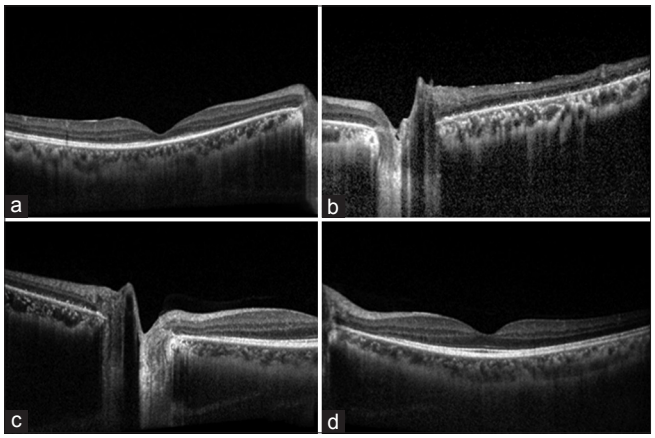

Figure 4: Spectral domain optical coherence tomography in right eye (a) showing foveal thinning, both eyes (a-d) showing trizonal distribution; normal outside of the demarcating line (zone 1), multifocal hyper reflective material present inside the demarcating line (zone 2), and loss of external limiting membrane and ellipsoid zone in peri papillary region corresponding to atrophic areas (zone 3)

A trizonal appearance was also seen on SD-OCT. The SD-OCT was normal outside of the demarcating line (zone 1). Inside the demarcating line, the multifocal hyper-reflective material was present resembling subretinal drusenoid deposits (zone 2). Photoreceptor, RPE, and choroidal atrophy were evident in the more advanced or long-standing area of the lesion, corresponding to loss of ellipsoid zone and ELM (zone 3).

In this era of multimodal imaging, AZOOR can be clearly differentiated from multiple evanescent white spot syndromes, acute macular neuroretinopathy, and other white spot diseases. The trizonal pattern of FAF and IR-AF imaging is a defining feature of AZOOR, particularly the hyperautofluorescent line demarcating the normal retina from the AZOOR lesion, pointing toward ongoing persistent activity in the disease. The trizonal pattern seen on SD-OCT showing disruption of both the ellipsoid and ELM is characteristic.

Conclusion

The multimodal imaging signatures provide striking information in detecting the AZOOR pathology, differentiating it from other white dot syndromes, and monitoring disease progression.

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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Conflicts of interest

There are no conflicts of interest.

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