Acute zonal occult outer retinopathy: Is optical coherence tomography angiography useful?

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A 44-year-old female presented with central vision loss and photopsia in both eyes since 2 months. Multimodal imaging, field defects, electroretinogram abnormalities, and spectral domain optical coherence tomography changes were all suggestive of acute zonal occult outer retinopathy. En-face optical coherence tomography angiography (OCTA) images demonstrated hyper-reflective dots at the level of ellipsoid zone in both eyes in the presence of normal retinochoroidal vasculature. The patient was started on oral azathioprine and prednisolone. On two consecutive monthly follow-ups, en-face OCTA images demonstrated serial changes in the hyper-reflective dot morphology at ellipsoid zone level that have not been previously reported in the literature.

Key words: Acute zonal occult outer retinopathy, optical coherence tomography angiography, subfoveal choroidal thickness

Acute zonal occult outer retinopathy (AZOOR) presents with central vision loss with or without photopsia due to outer retinal dysfunction. The exact etiopathogenesis of this condition is still not known. We report a case of AZOOR in a young adult female on the basis of multimodal imaging and novel findings on en-face OCTA imaging and their course over a 2-month follow-up.

Case Report

A 44-year-old female presented with sudden onset diminution of vision and photopsia in both eyes since 2 months. The best corrected visual acuity (BCVA) in her right eye was 6/36, N24 and in the left eye was 6/6, N6. Anterior segment findings were within normal limits. Fundus examination demonstrated an annular ring of altered reflex at the posterior pole in both eyes [Fig. 1a and b]. Fundus autofluorescence (FAF) imaging demonstrated gradually increasing hyperautofluorescence centrifugally from the fovea corresponding to the area of altered annular reflex, with the edge having maximum hyperautofluorescence [Fig. 1c and d]. Spectral domain optical coherence tomography (SDOCT) revealed thinning of the outer nuclear layer and loss of ellipsoid zone in both eyes [Fig. 2a, b, e, and f], the extent corresponding to the clinically visible lesion. Fluorescein angiography (FA) [Fig. 2c and g] and indocyanine green angiography (ICGA) [Fig. 2d and h] revealed parafoveal hyperfluorescence evident in the early phases and persisting through all phases of the angiogram. The subfoveal choroidal thickness (SFCT) was 385 μ and 399 μ in the right and left eye, respectively. Perimetry demonstrated central scotoma in both eyes [Fig. 1e and f]. A full-field electroretinogram (ERG) revealed reduced scotopic and photopic responses in both eyes [Fig. 1g and h]. Based on the history, imaging, and ancillary tests, a diagnosis of AZOOR was made. The patient was started on oral azathioprine [50 mg thrice daily] and a weekly tapering course of oral prednisolone [starting at 1 mg/kg body weight] and asked to follow-up after a month.

An OCTA was also performed at presentation. En-face OCTA structural analysis revealed hyper-reflective dots at the level of ellipsoid zone in the right eye in a "starry-sky" pattern [Fig. 3a] and in a "torpedo-like"' pattern in the left eye [Fig. 3d], corresponding to the area of ellipsoid zone loss on SDOCT. At 1 month, BCVA in the right eye dropped to 6/60, N36 but that in the left eye was 6/6, N6, which was preserved at 2-month follow-up. Serial en-face structural OCTA images demonstrated an increase in the density of hyper-reflective dots in the right eye at 1 month [Fig. 3b] which was more or less the same at 2-month follow-up [Fig. 3c]. However, in the left eye, the reflectivity of the dots showed a progressive decrease at the fovea [Fig. 3e and f, blue arrows] over the course of 2 months. At the same time, hyper-reflective dots around the subfoveal area increased over the first and second month follow-up [Fig. 3e and f, red arrows]. The vasculature of the retina (both superficial and deep), choriocapillaris, and the choroid at presentation [Fig. 4] and over two consecutive monthly follow-ups was within normal limits.

Discussion

First reported by Gass,[1] AZOOR typically affects young adult women who present with sudden onset of central scotomas usually accompanied by photopsias. The basic pathogenic mechanism is yet elusive, with both infectious[2] and autoimmune/inflammatory[3] etiologies being proposed. The diagnosis of AZOOR is made on the basis of a combination of fundus findings (especially FAF), field defects, ERG abnormalities, and SDOCT features.[4] FA may be normal or

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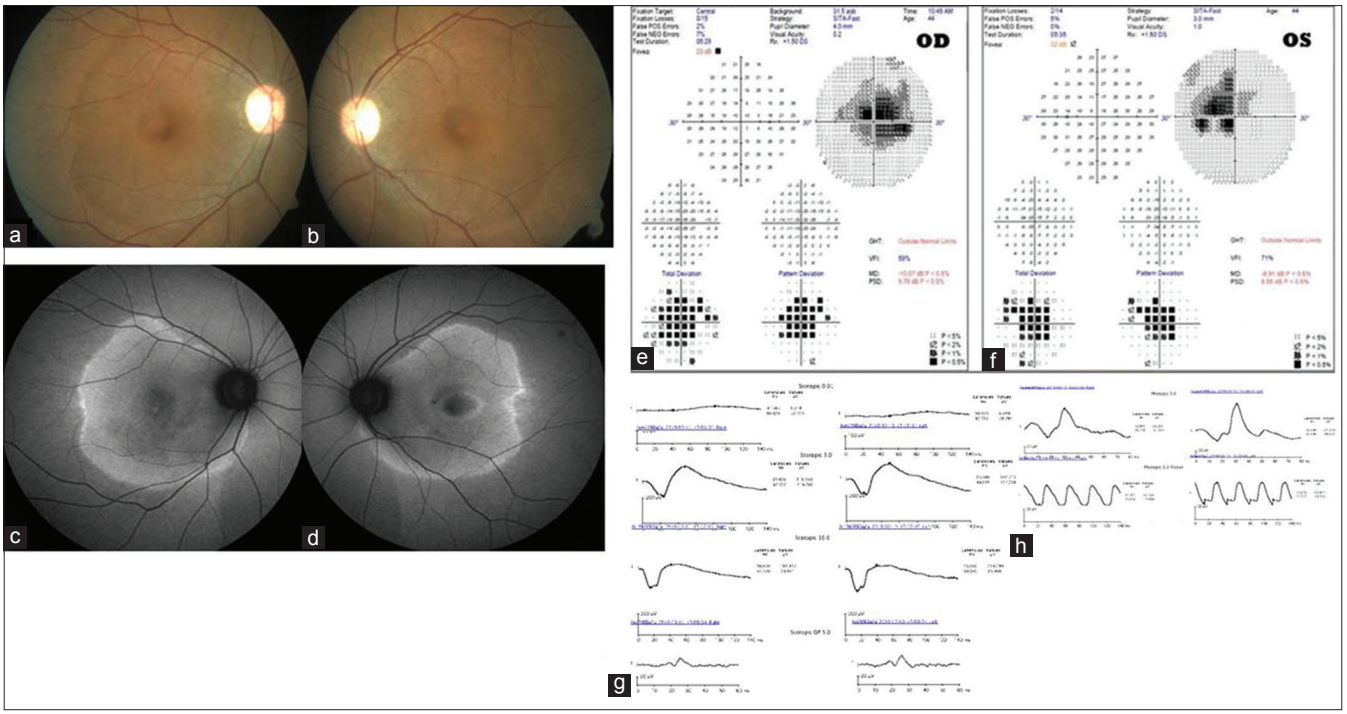

Figure 1: Fundus findings at presentation. (a and b) Fundus photograph at presentation demonstrating an area of altered retinal reflex at the posterior pole. (c and d) Autofluorescence images demonstrating gradually increasing hyperautofluorescence from the fovea to the edge of the lesion. (e and f) Visual fields demonstrating central scotoma. ERG demonstrating reduced scotopic (g) and photopic (h) responses

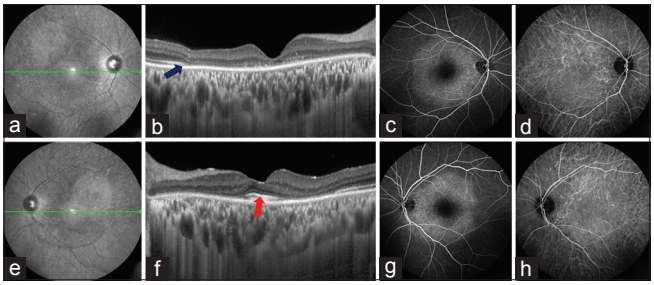

Figure 2: Ancillary investigations at presentation. (a, b, e, and f) spectral domain optical coherence tomography images – demonstrating outer nuclear layer thinning, loss of ellipsoid zone (b, blue arrow) corresponding to the clinically visible lesion. Hyper-reflectivity was noted at the level of inner choroid with increased sub-foveal choroidal thickness of 385 μ and 399 μ in the right and left eye, respectively. Note the intact subfoveal ellipsoid zone in the left eye (f, red arrow). (c and g) Fluorescein angiography demonstrating hyperfluorescence and (d and h) indocyanine green angiography demonstrating parafoveal hypercyanescence corresponding to the clinically visible lesion

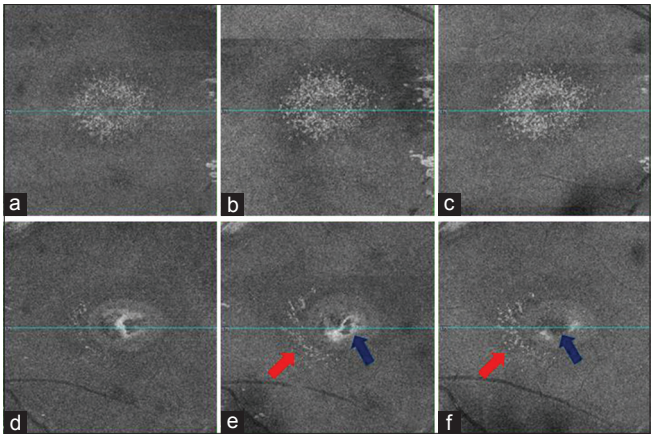

Figure 3: En-face optical coherence tomography angiography findings. (a) Starry-sky pattern of hyper-reflective dots at the ellipsoid zone slab in the right eye. Note the increase in density of hyper-reflective dots at 1 month (b), which was almost similar at 2 months (c). (d) Torpedo pattern of hyper-reflective dots in the left eye at the ellipsoid zone slab. Note the progressive decrease in the hyper-reflectivity at the fovea (e and f, blue arrows) with corresponding increase in the hyper-reflective dots in the parafoveal area over 1- and 2-month follow-up (e and f, red arrows)

show abnormalities related to the retinal pigment epithelium.[4] ICGA may show variable findings.[4] Ancillary investigations may demonstrate a wide range of abnormalities, none of which have been proven to be pathognomonic of AZOOR. However, it is certain that the outer retina is involved and the photoreceptors are maximally affected.

OCT angiography findings at presentation have not been reported in AZOOR. However, choroidal neovascularization in AZOOR was picked up only by OCT angiography in one report.[5] The structural en-face OCTA images clearly demonstrated the hyper-reflective dot structures at presentation. This has not been reported previously and hence the nature of these dots represents a matter of speculation. We postulate that these represent the degenerating photoreceptor segments. The cause of decreased visual acuity in the right eye but normal vision in the left eye in this patient may be because of the retained subfoveal ellipsoid zone integrity in the left eye [Fig. 2f, red arrow]. The difference in the pattern of hyper-reflectivity in both eyes is also unclear. We believe that the onset might

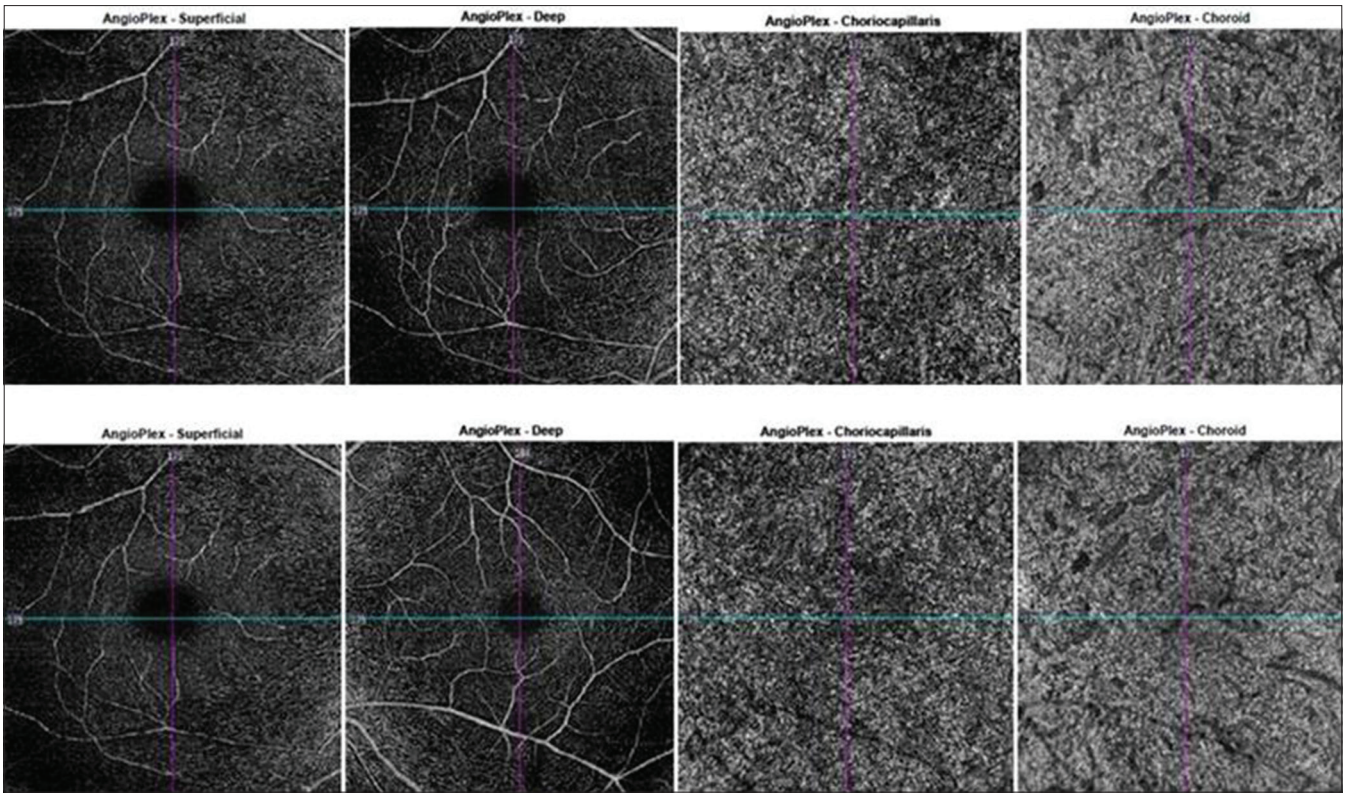

Figure 4: Optical coherence tomography angiography images of the vasculature at presentation - demonstrating normal retinal (superficial and deep), choriocapillaris, and choroidal vasculature in the right eye (top row) and the left eye (bottom row) (Images captured using AngioPlex™ optical coherence tomography angiography; Carl Zeiss Meditec Inc., Dublin, CA, USA)

have been asymmetric with the right eye being involved first. As a consequence, progressive photoreceptor damage in the right eye at an advanced stage revealed the "starry-sky" hyper-reflectivity. With immunosuppression, the ongoing disease process probably halted and the hyper-reflectivity may hence have decreased in the left eye. Another observation in our case was that the SFCT was increased. This leads us to another speculation – is AZOOR a part of pachychoroid spectrum? Higher SFCT in AZOOR eyes at baseline[6] has been demonstrated and impaired choroidal circulation with decreased blood flow velocity has been proven to be primarily affected in AZOOR.[7]

Conclusion

To the best of our knowledge, this is the first case where OCT angiography was performed at presentation in a case of AZOOR and en-face OCTA images demonstrated novel alterations in ellipsoid zone. OCTA may have a role for prognostication and response to treatment during the course of this disease. However, evidence in the form of prospective large-scale studies is needed.

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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