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Case report

Recovery of outer retina in acute idiopathic blind spot enlargement (AIBSE)

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ABSTRACT

Purpose: To report the anatomic and functional recovery of the ellipsoid zone in a case of acute idiopathic blind spot enlargement (AIBSE), which was documented by serial high-resolution optical coherence tomography imaging.

Observations: The patient's clinical presentation and follow up visits were documented via Humphrey's Visual Fields, fundus autofluorescence, and high resolution spectral domain optical coherence tomography (SD-OCT). At presentation, Humphrey's Visual Field testing showed an enlarged blind spot in the right eye. Fundus autofluorescence and optical coherence tomography showed an increased peripapillary autofluorescence and loss of the outer retinal layers, respectively. At 3 months a modest improvement in the visual field was observed. This improvement was stable at both the 7 and the 15 month follow up visits. SD-OCT corresponding to the areas of visual field improvement demonstrated recovery of the outer retina.

Conclusion and importance: Serial OCT imaging demonstrated anatomic evidence of ellipsoid zone recovery in isolated AIBSE. Anatomic recovery was consistent with the functional gain detected by visual field improvement.

1. Introduction

Acute zonal outer retinopathy (AZOOR) was first described by Gass, in his 1992 Donders Lecture to the Netherlands Ophthalmological Society [1]. Additionally, he and others, have suggested that AZOOR may represent a spectrum of diseases, including: multiple evanescent white dot syndrome (MEWDS), multifocal choroiditis (MFC), acute macular neuroretinopathy (AMN), presumed ocular histoplasmosis (POHS), punctate inner choroidopathy (PIC), acute annular outer retinopathy (AAOR), AZOOR and acute idiopathic blind spot enlargement (AIBSE) [1,2].

Collectively, both in clinical practice and in the literature, these diseases are referred to as the AZOOR Complex. Here we present a case of AIBSE in which we documented the recovery of the ellipsoid zone using serial multimodal imaging.

2. Case report

The patient presented with blurred vision in the right eye 1 month after experiencing superotemporal photopsias and metamorphopsia. The symptoms progressed over the course of a week, eventually stabilizing with resolution of the photopsias. The patient's past medical history was noncontributory and there was no history of a recent viral infection or autoimmune disease. Visual acuity was 20/25 and 20/20 in the right and left eye, respectively. Anterior segment examination was unremarkable with normal IOP in both eyes. Dilated fundus examination with biomicroscopy did not show retinal, choroidal or optic nerve abnormalities in either eye (Fig. 1). Humphrey Visual Fields (HVF) demonstrated enlargement of the blind spot in the right eye only with a mean deviation (MD) of −4.20 dB, $p < 0.5\%$ (Fig. 2). SD-OCT revealed loss of the ellipsoid zone (EZ) and outer nuclear layer (ONL), resulting in inner retinal collapse that extended from the peripapillary area to the parafoveal region (Fig. 1). Increased peripapillary autofluorescence was seen in the right eye (Fig. 3). A diagnosis of AIBSE was made based on symptoms, clinical exam, and imaging findings.

The patient was monitored without intervention for a period of 3 months.

Upon returning to the clinic a dilated fundus examination was unremarkable. Repeat HVF testing demonstrated improvement of the scotoma (MD = −3.55 dB, p< 1%; Fig. 2). However, increased peripapillary autofluorescence persisted (Fig. 3). Repeat SD-OCT imaging showed partial recovery of the parafoveal ellipsoid zone

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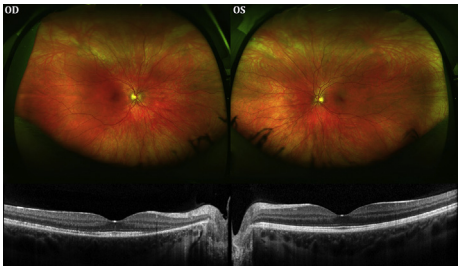

Fig. 1. Color fundus and horizontal optical coherence tomography (OCT) images were taken of the right and left eye at the time of presentation. OCT of the right eye displayed evidence of ellipsoid zone loss and outer retinal collapse in the area stretching from adjacent to the optic nerve to the parafoveal region. (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

(Fig. 4, 1162 μm of intact EZ from the foveal center at presentation vs. 1642 μm at 3 months). EZ recovery was limited to the part of the retina with relatively intact outer nuclear layer. After initial improvement, HVF and OCT findings remained stable at both the 7 and 15 month follow up visits although the MD continued to modestly improve (MD = −3.05 dB, $p < 2\%$ and MD = −2.92 dB, $p < 2\%$, respectively; Fig. 2). At 15 months, visual acuity in the right eye returned to 20/20 and the stable improvement in peripapillary hyper autofluorescence was noted (Fig. 3).

No consent was obtained from the patient to report this case. Consequently, we have omitted all identifiable information.

3. Discussion

There continues to be much debate regarding the classification of AZOOR and the AZOOR Complex diseases. The classification of AIBSE, which was originally described by Fletcher in 1988, is perhaps the best example of this disagreement [3]. In 1995, Jampol stated that AIBSE was likely a nonspecific finding associated with MEWDS, where the white dots were either very light or were not yet apparent [4]. Similarly, Yannuzzi in his 2013 Charles L. Schepens Lecture to the American Academy of Ophthalmology, suggested AIBSE was in fact a harbinger signifying the presence of either MEWDS, MFC or AZOOR [5]. He proposed that observing the clinical progression of individuals diagnosed with AIBSE would likely reveal the following: regression of the blind spot enlargement corresponds with the diagnosis of MEWDS, while persistence of the blind spot enlargement with peripapillary atrophy would lead to a diagnosis of MFC and finally persistence/progression of the blind spot enlargement was almost certainly AZOOR [5]. Alternatively, in a recent case series containing 27 patients diagnosed with AIBSE, Volpe suggested that AIBSE was a unique entity separate from MEWDS, MFC and AZOOR [6]. Volpe et al. described a characteristic clinical course in which positive visual symptoms resolve, yet a persistent and stable visual field defect remains [6].

The combination of our patient's symptomatology, clinical exam findings and imaging studies allowed us to make the diagnosis of AIBSE. Additionally, through our use of multimodal imaging techniques (specifically SD-OCT), we were able to gain surrogate histopathologic confirmation which explained this patient's clinical presentation. On the 3, 7 and 15 month follow up visits there were improvements in HVF (although complete resolution of the enlarged scotoma was not observed) and an eventual return of visual acuity back to 20/20 in the affected eye. Importantly, serial imaging studies showed that the patient's rescue of visual function corresponded with anatomic recovery of the EZ in an area of the diseased retina with relative sparing of the other outer retinal layers on SD-OCT. EZ recovery was not seen in the most nasal aspect of the macula where there was complete atrophy of the ONL. To our knowledge, this AIBSE case report's OCT findings provide the first account of virtual histology demonstrating that recovery of the outer retina resulted in improvement of the patient's visual field and visual acuity.

Lastly, this case report highlights the importance of multimodal imaging in assessing the AZOOR Complex diseases. If indeed AIBSE is a nonspecific, perhaps early finding, which suggests the presence of a more serious disease, such as MFC, then baseline images will allow the physician to more closely track their patient's clinical course and provide appropriate therapy in a timely manner. On the other hand, if the AZOOR complex actually represents related yet separate diseases, multimodal imaging could be critical in defining each diagnosis. For example, Mrejen et al. recently suggested that characteristic trizonal fluorescein angiography findings correlated with SD-OCT were sufficient to accurately diagnose AZOOR as a disease separate from the AZOOR complex [7]. Our findings indicate that anatomic recovery of the EZ and visual improvement is

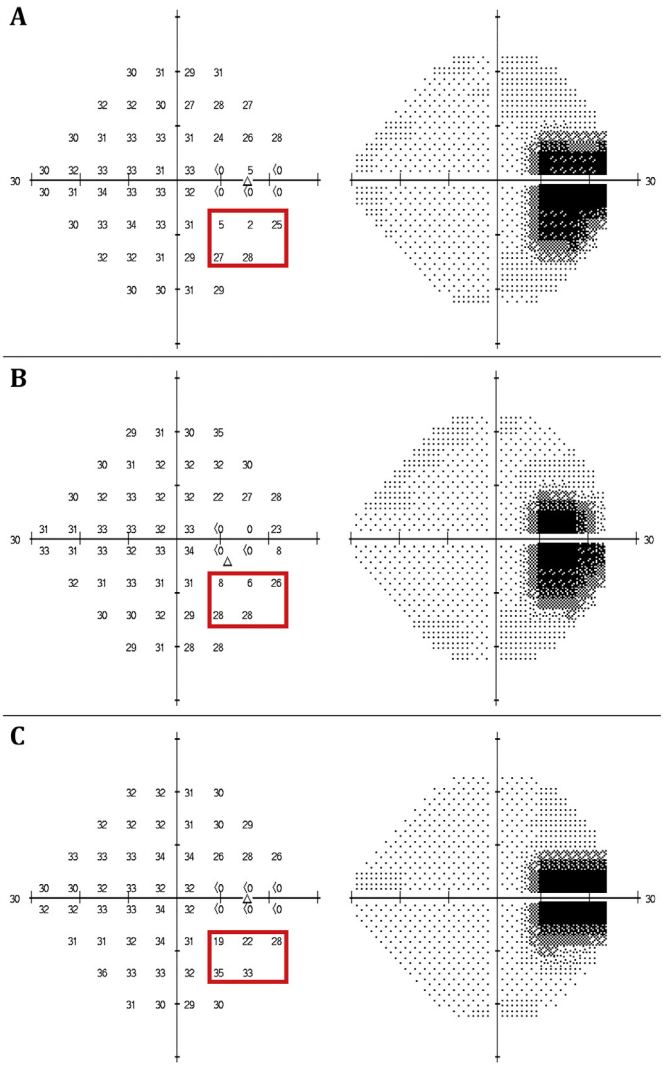

Fig. 2. When comparing the initial Humphrey visual field (HVF) (A), to the 3 month HVF (B) there was a modest improvement of the scotoma at the 3 month follow up visit. This improvement is evidenced by the reduction in the mean deviation and increase in the threshold decibel values (see red box). Subsequent HVF's remained stable as evidenced by the 15 month HVF (C) included in the lower panel. Mean Deviations were −4.20, −3.55, −3.05, and −2.92 dBs at presentation, 3, 7 and 15 month follow up visits. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

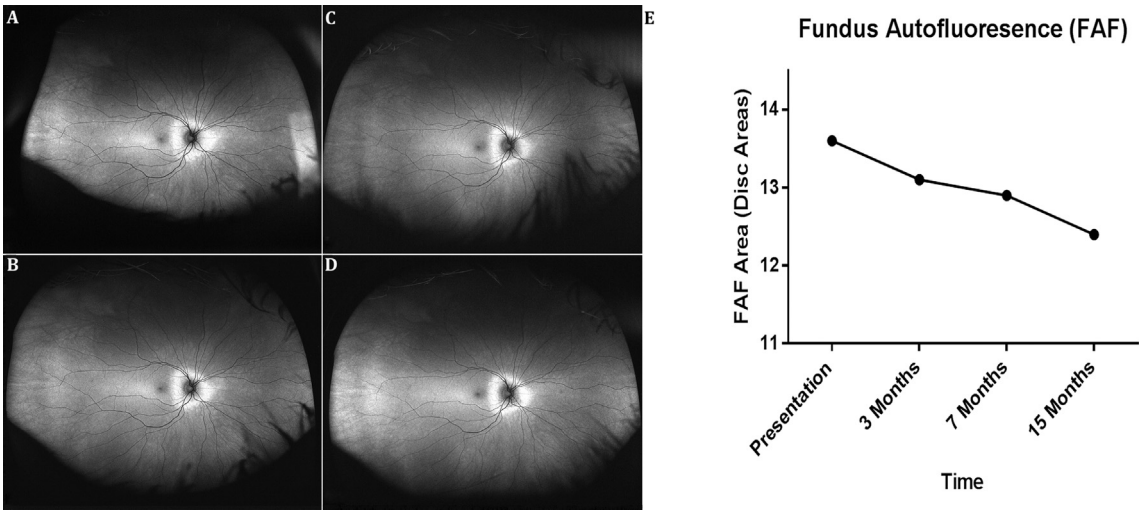

Fig. 3. Persistent peripapillary autofluorescence was observed in the right eye at all visits (A = Presentation, B = 3months, C = 7months, D = 15months). However, there is noticeable improvement in hyper autofluorescence as time progresses. Qualitatively, a reduction in fundus autofluorescence was observed at each of the follow up visits. We used the measurement and area tools in the Optos Review Software package to define the area of the optic nerve head and the area of the peripapillary autofluorescence. Next, we normalized the area of the peripapillary autofluorescence, to the measured area of the optic disc and plotted the total area of peripapillary autofluorescence as a ratio of optic disc areas (E) - Presentation = 13.6, 3 month = 13.1, 7 months = 12.9, 15 months = 12.4 disc areas.

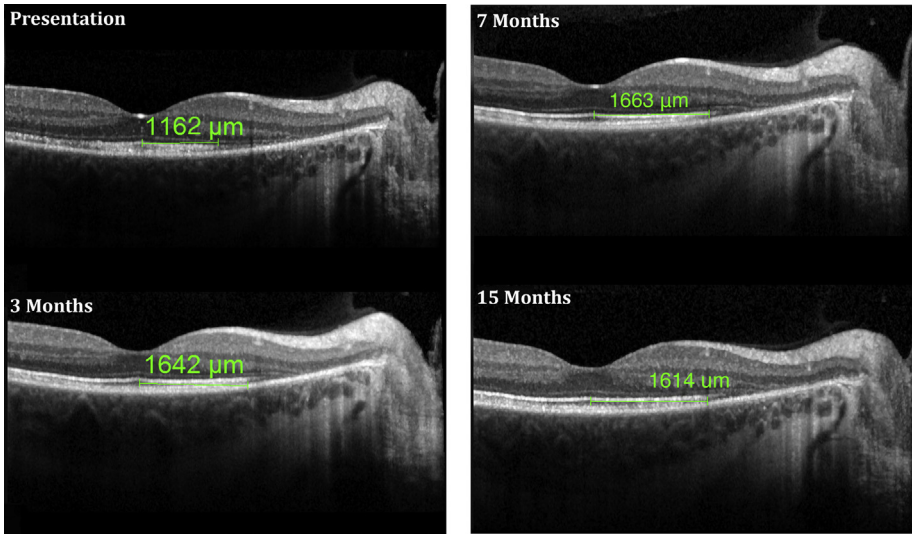

Fig. 4. Quantification of ellipsoid zone recovery was accomplished via serial optical coherence tomography (OCT) imaging and analysis was performed using the Heidelberg image analysis software. At presentation, we measured from the foveola to the loss of the ellipsoid zone (top panel = 1162 µm). There was a significant recovery of the ellipsoid zone, which measured 1642 microns at 3 months. This improvement remained stable at the 7 (1663 µm) and the 15 month (1614 µm) follow up visits.

possible in AIBSE cases where there is relative sparing of the ONL. This observation would be important in accurate counseling of the patient and serial imaging could be used to provide prognostic information.

4. Conclusions

The classification of AIBSE and the AZOOR Complex diseases will remain controversial until the disease(s) etiology becomes clear. Nonetheless, this case report provides evidence of the functional recovery of vision in a patient diagnosed with AIBSE. Through multimodal imaging we were able to document the dynamic anatomic relationship of the outer retina, which clearly demonstrates that recovery of the photoreceptor cell layer resulted in improved visual function.

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