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Changes in Optical Coherence Tomography and Fundus Autofluorescence Findings in Acute Zonal Occult Outer Retinopathy

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Abstract **Case Report**

We report a case of peripapillary acute zonal occult outer retinopathy (AZOOR) in a 43-year-old woman. Fundus photographs showed no specific abnormal findings, while blue-light fundus autofluorescence (FAF) imaging revealed well-defined hyperfluorescent areas in the peripapillary region, excluding the macula. The ellipsoid zone (EZ) was absent on optical coherence tomography (OCT) images of this area. Visual field testing with Goldmann perimetry showed blind spot enlargement and multifocal electroretinogram showed corresponding amplitude reductions. Three months after the initial visit, the EZ had reappeared and both FAF and visual field had improved. FAF together with OCT may be useful in the evaluation and monitoring of AZOOR.

Keywords: acute zonal occult outer retinopathy, optical coherence tomography, fundus autofluorescence, ellipsoid zone.

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INTRODUCTION

Acute zonal occult outer retinopathy (AZOOR) is characterised by an acute zonal loss of outer retinal function affecting one or both eyes. AZOOR occurs mainly in young women. Initially there are minimal or no fundus changes, but this slowly progressive form of retinal pigment epithelial (RPE) degeneration often leads to enlarged blind spots, multifocal electroretinogram (mfERG) abnormalities and permanent visual field loss [1, 2]. Optical coherence tomography (OCT) images of eyes with AZOOR show loss or irregularity of the ellipsoid zone (EZ) in areas corresponding to reduced mfERG responses and visual field defects [3-5]. Fundus autofluorescence (FAF) is a useful imaging modality for assessing the metabolic status of the outer retina and retinal pigment epithelium (RPE) [6-9]. To our knowledge, there are few reports in the literature describing changes in FAF findings in AZOOR [6-8]. Here, we report the changes in OCT and FAF findings in patients with AZOOR.

CASE REPORT

A 43-year-old Japanese woman presented with photopsia and a paracentral scotoma in her right eye. Best corrected visual acuity (BCVA) was 1.2 in both eyes and intraocular pressure measurements were normal. The patient had no significant medical history. No inflammatory cells were observed in the anterior segment or vitreous of either eye, and there were no specific abnormal findings on fundus examination (Figure 1A). Blue-light FAF images showed well-defined hyperfluorescent areas in the peripapillary regions, excluding the macula (Figure 1C). Visual field testing by Goldmann perimetry showed a blind spot enlargement in the right eye (Figure 1E). The size of the visual field defect corresponded to the size of the fundus lesions seen on FAF. OCT showed attenuation of the EZ with the exception of the macula in her right eye (Figure 1G). No specific abnormalities were found in her left eye (Figure 1B, D, F, H). Fundus photograph shows no specific abnormalities in the right eye (A). Blue-light fundus autofluorescence imaging shows hyperfluorescent areas excluding the macula in the right eye (C arrows). Goldmann perimetry shows blind-spot enlargement in the right eye (E). Optical coherence tomography shows attenuation of the ellipsoid zone excluding the macula in the right eye (G arrows). No specific abnormalities were detected in the left eye (B, D, F, H). Reduced mfERG amplitudes corresponding to the area of abnormal FAF were observed in the right eye. Based on the patient's history and the aforementioned examinations, peripapillary AZOOR was diagnosed. The patient was followed without treatment. Three months after the first visit, FAF (Figure 2A) and visual field (Figure 2B) both improved.

In addition, the reappearance of the EZ was detected by OCT (Figure 2C). Fluorescein angiography and indocyanine green angiography were not available in this case.

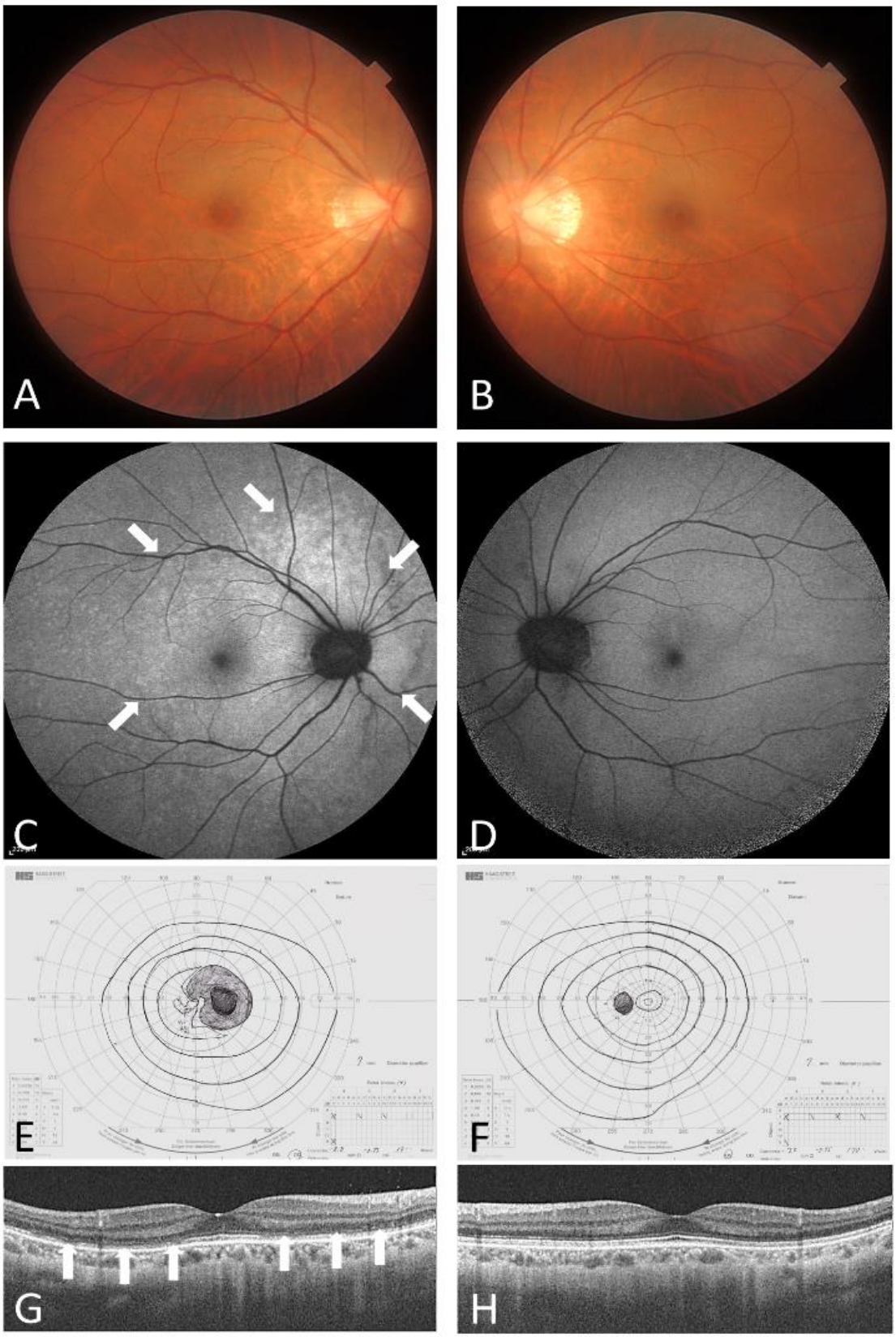

Figure 1: Examination findings at the initial visit

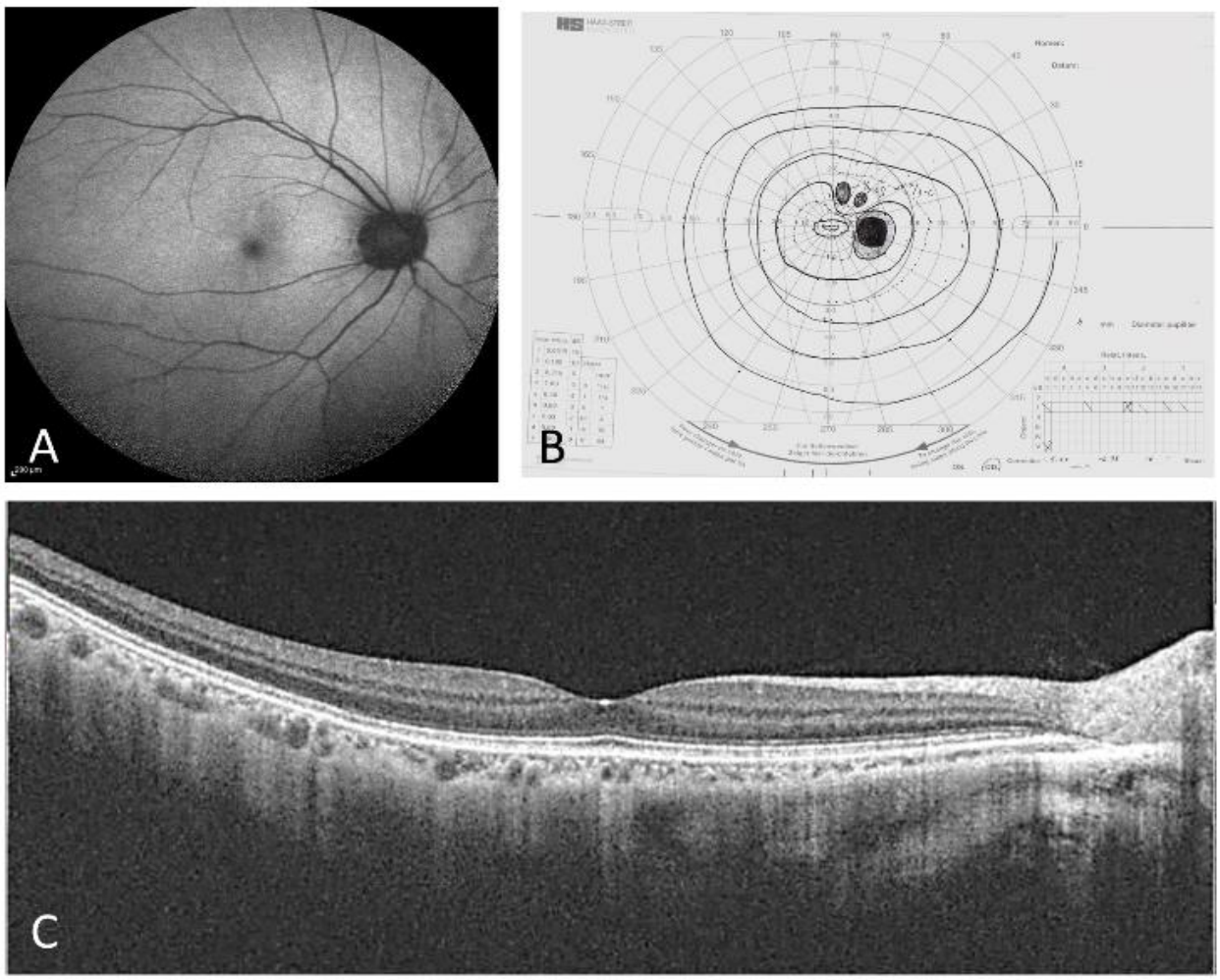

Figure 2: Examination findings three months after the initial visit

Blue-light fundus autofluorescence imaging shows normal autofluorescence (A). Goldmann perimetry shows improvement of blind-spot enlargement (B). Optical coherence tomography scan shows reappearance of the ellipsoid zone (C).

DISCUSSION

This present case highlighted that complete restoration of the EZ defect, which was associated with FAF recovery. The most common finding on FAF imaging in AZOOR is a zone of hypoautofluorescence surrounded by a border of hyperautofluorescence [8]. The hypoautofluorescence in the central portions of the AZOOR lesion is due to RPE atrophy, whereas the hyperautofluorescence of the border of the lesion is attributed to accumulation of lipofuscin-like material at the edges of the lesion. However, to our knowledge, there are few reports in the literature describing the changes of FAF in AZOOR [6-8]. Fujiwara et al. [6] investigated how AZOOR is represented on FAF and OCT in 19 eyes. According to their report, abnormalities on FAF imaging were detected in 17 (89%) of 19 eyes and they fell into the following three broad categories: only peripapillary involvement (53%), peripheral involvement (41%), and posterior pole involvement not primarily centered on the optic nerve (18%). Of the eyes with FAF abnormalities, 14 eyes (82%) had broad areas of hypoautofluorescence and 3 eyes (18%) had coarse granular regions of mixed hyper- and hypoautofluorescence. The boundary of autofluorescent abnormalities was well-defined in 11 eyes (65%), with 6 (35%) of these having a hyperautofluorescent border. Nine eyes (53%) showed progression of hypoautofluorescent areas during follow-up. Shifera et al. [8] reported the utility of FAF in monitoring the progression of AZOOR. According to their report, nine of the 10 subjects (14 eyes) had some degree of follow-up ranging from 1 month to 29 months with repeat FAF imaging at last follow-up. In 4 of these subjects, there was an improvement in symptoms and/or FAF findings.

In FAF images, hyperfluorescence was presumed to result from accumulated lipofuscin in RPE cells and to represent metabolic activity of RPE related to photoreceptor outer segment turnover [6-9]. Conversely, hypofluorescence is thought to represent areas of decreased RPE metabolism resulting from photoreceptor and/or RPE atrophy [6-9]. In particular,

blue-light FAF is a useful imaging modality for the evaluation of the metabolic status of the outer retina and the RPE [10-13]. However, the exact mechanism of hyperautofluorescence in acutely affected areas of the retina is not entirely clear. It is possible that this is due to photoreceptor outer segment inflammation resulting in loss of photopigment, and a subsequent relative increase in emission of autofluorescence from the underlying RPE [8]. The subjects who demonstrated return to normal autofluorescence seem to support this hypothesis rather than a primary RPE process for hyperautofluorescence, especially given the relatively low-intensity hyperautofluorescence this acute process produces [8]. Alternatively, increased production of autofluorophores from inflamed photoreceptor outer segments or infiltrating leukocytes could potentially contribute to the above finding. Similarly, in this present case, FAF in the formerly hyperfluorescent areas returned to normal within three months.

In conclusion, the formerly hyperfluorescent areas returned to normal FAF, and the improvement in FAF seemed to coincide with the improvement in OCT. Our findings were based on a single case of AZOOR and further studies with additional cases are necessary.

Disclosure: The author declares no conflict of interest.

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