Acute Idiopathic Blind Spot Enlargement Syndrome In A 63-Year-Old Man

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Citation

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Abstract

We present a case of acute idiopathic blind spot enlargement syndrome in a 63-year-old man. The enlarged blind spot was accompanied by no abnormal funduscopic findings. Optical coherence tomography showed attenuation of the photoreceptor inner segment/outer segment line between the disc and the nasal side of the macula corresponding to the visual field defect. Even in elderly patients with enlarged blind spots, it is important to consider the possibility of acute idiopathic blind spot enlargement syndrome.

INTRODUCTION

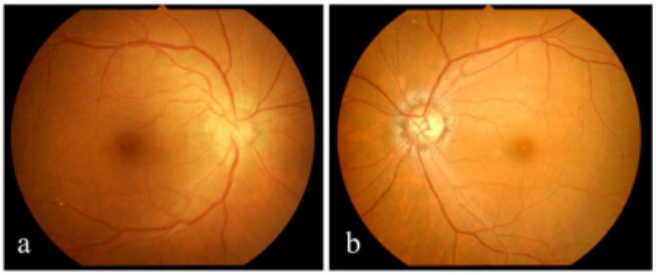
Acute idiopathic blind spot enlargement (AIBSE) syndrome was first reported in 1988 by Fletcher et al.1 as a clinical entity presenting with sudden scintillations and a temporal scotoma centered on the blind spot on an otherwise normal fundus. Later, AIBSE was reported to belong to a spectrum of conditions that include acute zonal occult outer retinopathy (AZOOR), acute macular neuroretinopathy, multiple evanescent white dot syndrome, presumed ocular histoplasmosis, punctate inner choroidopathy, and multifocal choroiditis and panuveitis, collectively called the AZOOR complex.2-6 All of these disorders are most common in young adult women, and each may be associated with visual field loss and abnormalities on electroretinograms. Optical coherence tomography (OCT) studies of eyes with AZOOR complex disorders reveal a loss or irregularity of the photoreceptor inner segment/outer segment (IS/OS) line in areas corresponding to the visual field defects.4-8 In this study, we report the findings for a 63-year-old patient with AIBSE.

CASE REPORT

A 63-year-old Japanese man presented with a 1-week history of a photopsias and paracentral scotoma in his right eye. Best corrected visual acuity was 0.7 and 1.2 in his right and left eyes, respectively. Intraocular pressure was 16 mmHg in each eye, and the patient had no specific 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 of the right eye (Figure 1a), but peripapillary discoloration was observed in the left eye (Figure 1b).

Figure 1

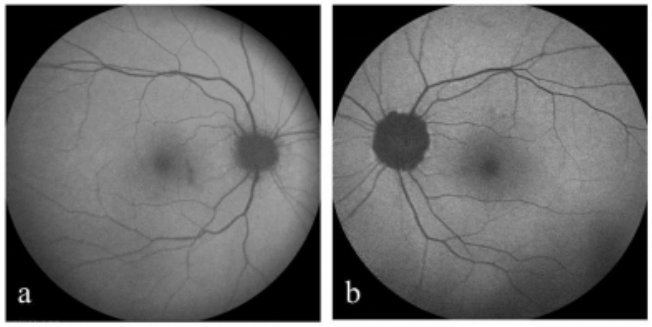
Fundus photographs show no specific abnormalities in the right eye (a), while peripapillary discoloration was evident in the left eye (b).



Fundus autofluorescent (FAF) imaging showed no specific abnormal findings in the right eye (Figure 2a), but clearly defined hypofluorescent areas in the peripapillary region were detected in the left eye (Figure 2b).

Figure 2

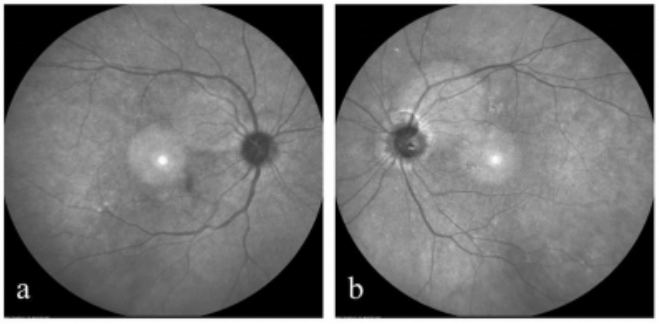
Fundus autofluorescence images show no specific abnormalities in the right eye (a), while clearly defined hypofluorescent areas in the peripapillary regions were detected in the left eye (b).



Near-infrared reflectance (NIR) imaging showed no specific abnormal findings in the right eye (Figure 3a), but mild hyperfluorescent areas in the peripapillary region were detected in the left eye (Figure 3b).

Figure 3

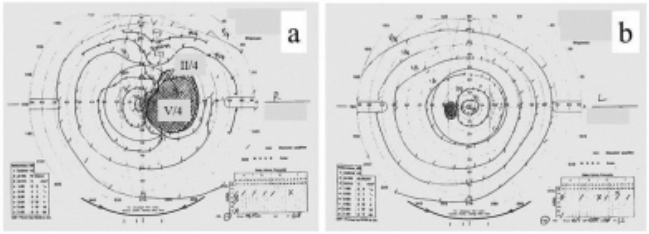
Near-infrared reflectance images show no specific abnormalities in the right eye (a), while mild hyperfluorescent areas in the peripapillary regions were detected in the left eye (b).



The Heidelberg Retina Angiograph 2 (Heidelberg Engineering, Heidelberg, Germany) was used to perform FAF and NIR imaging. Visual field testing by Goldmann perimetry showed blind spot enlargement in the right eye (Figure 4a), while the visual field was normal in the left eye (Figure 4a).

Figure 4

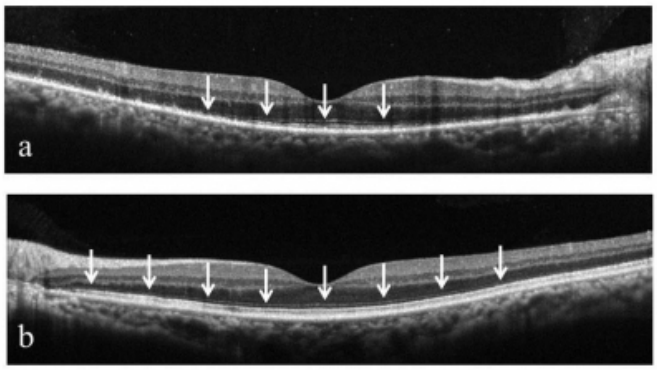
Goldmann perimetry showed blind spot enlargement in the right eye (a), while no specific abnormalities were detected in the left eye (b).



While OCT (RS-3000; Nidek, Gamagori, Japan) showed attenuation of the IS/OS line between the disc and the nasal side of the macula in the right eye (Figure 5a), no specific abnormalities were detected in the left eye (Figure 5b).

Figure 5

Optical coherence tomography horizontal scan revealed the absence of a photoreceptor inner segment/outer segment (IS/OS) line between the disc and the nasal side of the macula in the right eye (a). In the left eye, the IS/OS line was detected (b). Arrows indicate the IS/OS line.



Fluorescein angiography, indocyanine green angiography, and multifocal electroretinograms were not available in this case.

DISCUSSION

Due to the enlarged blind spot with no abnormal ophthalmoscopic findings, but with OCT abnormalities in the vicinity of the visual field defect, we diagnosed our patient with AIBSE syndrome.

Of the 70 published cases of AIBSE1-3, 9, 10 in which sex was specified, 57 (81%) involved women and 13 (19%) involved men. Additionally, most AIBSE patients are young adults, with an age range of 16 to 57 years.1-3, 9, 10 AZOOR also occurs in young women.5 Similar to AIBSE, of the 130 published cases of AZOOR in which sex was

specified, 99 (76%) involved women and 31 (24%) involved men.5 Most AZOOR patients are young adults, with an average age of 36.7 years (range: 13–79 years) in the 103 published cases.5

Fujiwara et al.8 investigated how AZOOR presents on OCT and FAF in 19 eyes. They described IS/OS boundary abnormalities in all 19 eyes studied: 14 eyes (74%) showed some absence of the IS/OS boundary and five eyes (26%) showed attenuation of the IS/OS boundary on OCT scans. The OCT findings in the present case were consistent with those previously reported for other AZOOR patients.4-8

In addition, in our case, hypofluorescent peripapillary areas on FAF were detected in the non-affected eye. Fujiwara et al.8 reported that FAF imaging abnormalities were detected in 17 (89%) of 19 eyes, which were grouped into the following three broad categories: peripapillary involvement only (53%), peripheral involvement (41%), and posterior pole involvement not primarily centered on the optic nerve (18%). Volpe et al.9 described focal areas of peripapillary deep pigmentary changes in some patients with AIBSE. Although no enlarged blind spot in the left eye was evident in our case, previous AZOOR complex disorder could not be completely ruled out.

Finally, our findings are based on a single AIBSE case; however, it is important to consider the possibility of AIBSE even in the elderly who have an enlarged blind spot.

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