Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com/journal/sjmcr/home

OPEN ACCESS

Acute Idiopathic Blind Spot Enlargement Syndrome

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| **Received:** 07.04.2019 | **Accepted:** 17.04.2019 | **Published:** 30.04.2019
DOI: 10.36347/sjmcr.2019.v07i04.006

Abstract **Case Report**

We present a case of acute idiopathic blind spot enlargement syndrome (AIBSE) in a 25-year-old woman. 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. The patient was followed without any treatment. Enlarged blind spot was gradually reduced during the 3-month follow-up period. Our findings may contribute to a better understanding of the natural course of AIBSE.
Keywords: acute idiopathic blind spot enlargement syndrome, acute zonal occult outer retinopathy.

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 syndromes, 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 25-year-old patient with AIBSE.

CASE REPORT

A 25-year-old Japanese woman presented with a 1-week history of visual field abnormality in her left eye. Best corrected visual acuity was 1.2 in both eyes. There were no specific abnormal findings on fundus examination (Figure 1).

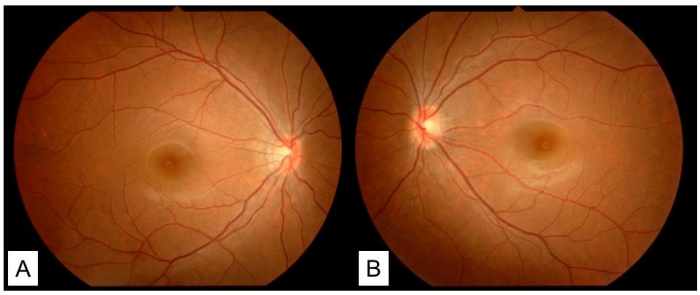

Fig-1: Fundus photographs of the (A) right and (B) left eyes at the initial visit
Note no specific abnormal findings.

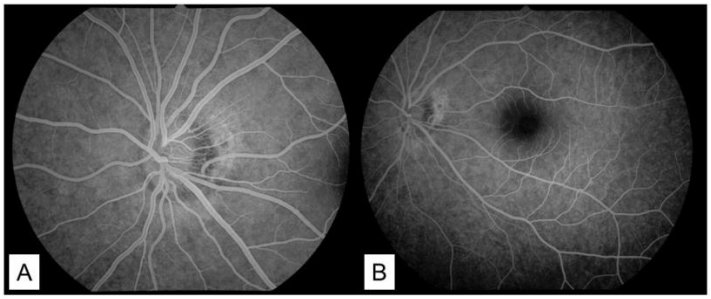

Fig-2: Fluorescein angiography of the (A) early and (B) late phase in the left eye
Note no specific abnormal findings.

Fluorescein angiography showed no specific abnormal findings in both eyes (Figure 2),

Indocyanine green angiography showed no specific abnormal findings in boht eyes (Figure 3).

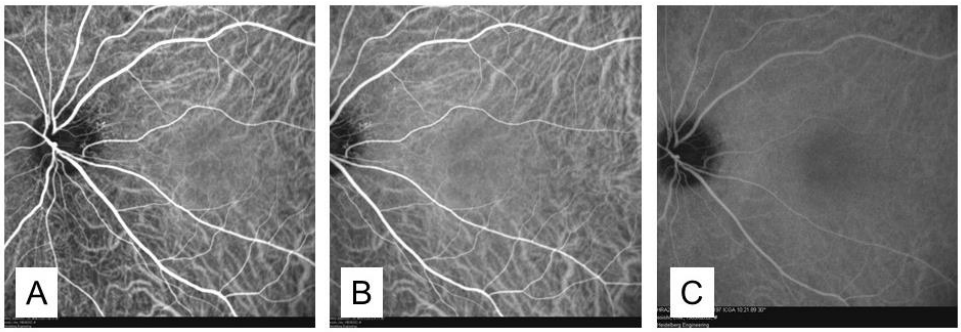

Fig-3: Indocyanine green angiography of the (A) early, (B) middle and (C) late phase
Note no specific abnormal findings.

Visual field testing by Goldmann perimetry showed blind spot enlargement in the left eye (Figure 4B), while the visual field was normal in the right eye (Figure 4A).

OCT showed attenuation of the IS/OS line between the disc and the nasal side of the macula in the left eye (Figure 5).

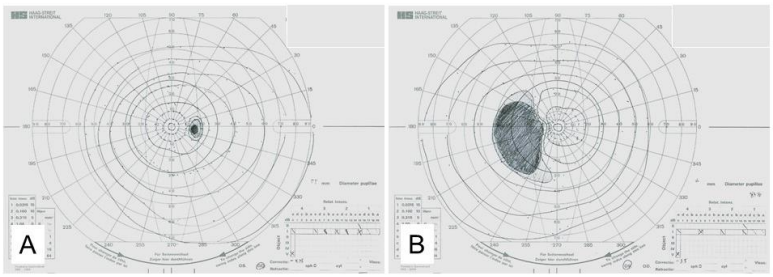

Fig-4: Goldmann perimetry of the (A) right and (B) left eyes
Note blind spot enlargement in the left eye.

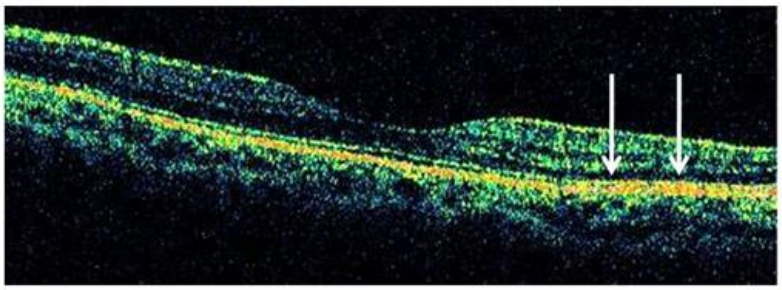

Fig-5: Optical coherence tomography horizontal scan of the left eye
Note attenuation of the IS/OS line (arrows).

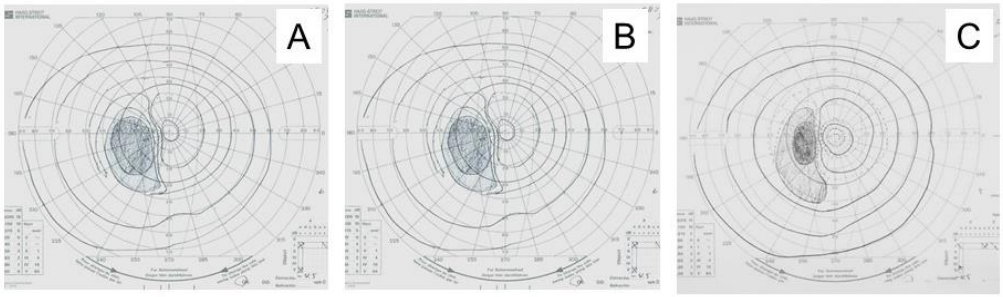

Fig-6: Goldmann perimetry of the left eye
A: initial visit; B: 1 month after initial visit; C: 3 months after initial visit

The patient was followed without any treatment. Enlarged blind spot was gradually reduced during the 3-month follow-up period (Figure 6).

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 AIBSE in which sex was specified, 57 (81%) involved women and 13 (19%) involved men [1-3, 9, 10]. Additionally, most AIBSE patients are young adults, with an age range of 16 to 57 years. 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].

In the largest series of AIBSE patients, Volpe *et al*. [9] evaluated 27 patients with AIBSE. According to their report, blind spot enlargement was highly variable in terms of size and density. In addition, there was no improvement of the enlarged blind spot in the 10 patients who were followed up. In this present case, enlarged blind spot was gradually improved in density rather than size.

CONCLUSION

Although our report is based solely on a patient and only a 3-months follow-up period, our observation may contribute to a better understanding of the natural course of AIBSE.

Disclosure

The author declares no conflict of interest.

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