Acute idiopathic blind spot enlargement syndrome mimicking optic neuropathy

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Optic neuropathy may be mimicked by retinal diseases like acute idiopathic blind spot enlargement (AIBSE) syndrome. We describe a 43-year-old woman with unilateral loss of vision associated with optic disc pallor. Following the clinical examination, the patient was submitted to magnetic resonance imaging to rule out structural changes and optic nerve enhancement. Multimodal retinal imaging revealed a pigmentary change adjacent to the optic nerve. Finally, AIBSE was confirmed by automated visual field testing, establishing the diagnosis.

Key words: Blind spot enlargement, optic neuropathy, uveitis

Optic neuropathy may be mimicked by retinal diseases like acute idiopathic blind spot enlargement (AIBSE) syndrome, a condition first described by Fletcher *et al*. in 1988.[1,2] In this article, we report a patient referred to the neuro-ophthalmologist with unilateral loss of vision associated with optic nerve pallor. Following clinical examination and multimodal retinal imaging, the patient was diagnosed with AIBSE syndrome.

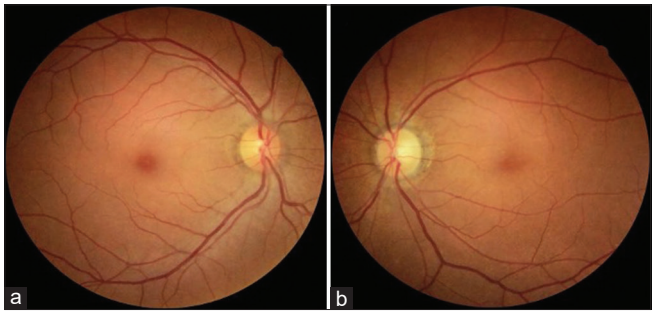

Figure 1: Retinography of OD: clear vitreous, pink optic disc, defined borders and cup, attached retina, vessels without changes (a). Retinography of OS: clear vitreous, optic disc with slight temporal pallor and adjacent pigmentary changes, attached retina, vessels without changes (b). OD: Right eye, OS: Left eye

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Case Report

A 43-year-old woman with loss of vision in the left eye (OS) was referred to the neuro-ophthalmologist. No comorbidities, use of medication, or ophthalmological history was reported. The clinical findings included myopia, visual acuity of 20/20 in the right eye (OD) and 20/25 in OS, and a positive afferent pupillary defect in OS. On biomicroscopy, no sign of inflammation was observed, and tonometry was normal. Fundoscopy revealed temporal optic nerve pallor in OS associated with hyperpigmentation of the adjacent retina [Fig. 1b]. No changes in OD [Fig. 1a] were visible. A magnetic resonance scan of the cranium and orbits also revealed no changes. Autofluorescence was normal in OD [Fig. 2a] but showed hyperautofluorescent changes around the disc in OS [Fig. 2b]. Optic coherence tomography (OCT) [Fig. 3 and b] identified no changes in OD but a disruption of the external retinal layers around the optic disc in OS. Finally, automated visual field testing confirmed the diagnostic hypothesis of AIBSE in OS [Fig. 4b]. No change was observed in visual acuity or on fundoscopy by the end of the follow-up period.

Discussion

AIBSE syndrome affects mostly middle-aged myopic women, with a preference for Caucasians or with no racial preference.[1,3]

The pathogenesis of AIBSE remains uncertain, but a series of stages have been hypothesized[4]: Following the acute inflammatory and involutionary stages, the disease becomes chronic. At this point, choriocapillaris lesions may be observed,

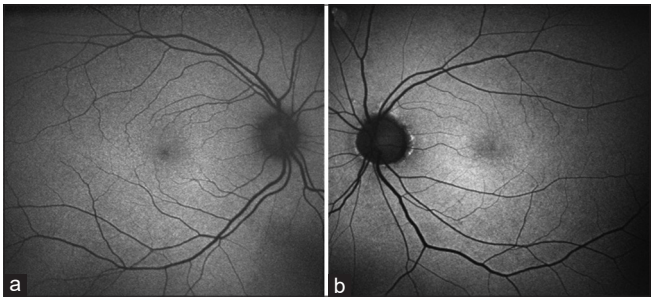

Figure 2: Autofluorescence of OD: Physiological hypoautofluorescence of the optic nerve, vessels, and the foveal region (a). Autofluorescence of OS: hypoautofluorescence associated with areas of punctiform hyperautofluorescence in the peripapillary region (b). OD: Right eye, OS: Left eye

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as in our patient.[4] This is followed months later by a stage of recovery during which OCT changes disappear and visual acuity tends to improve.[4,5]

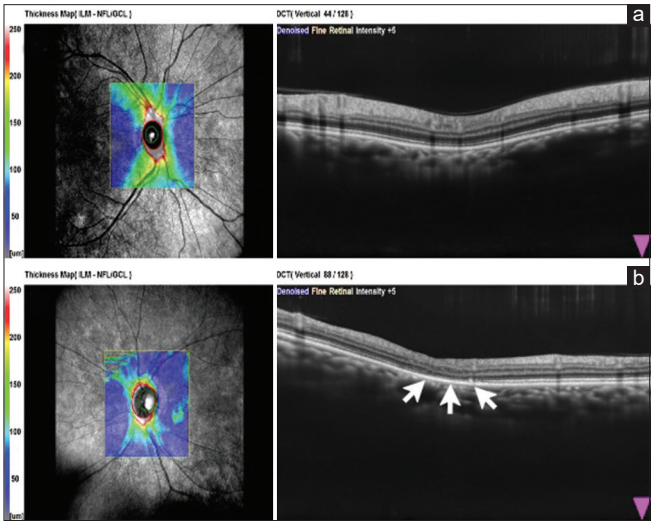

Figure 3: Optical coherence tomography of OD: retinal vitreous interface without changes, internal and external retinal layers preserved (a). Optical coherence tomography of OS: retinal vitreous interface without changes, internal and external retinal layers preserved, disruption of the external retinal layers in the peripapillary region (b). OD: Right eye, OS: Left eye

Diagnosis is established by multimodal retinal imaging. Autofluorescence exhibits patches of hyperautofluorescence in the affected region [Fig. 2b].[4] OCT allows to visualize microstructural changes in the external retina [Fig. 3b].[5] Multifocal electroretinography (mERG) can detect functional changes even in the absence of structural lesions or in minor lesions (as in our patient), though this modality was not employed.[4,5] The test from which the syndrome derives its name (the automated visual field test) detects enlargement of the blind spot [Fig. 4b], a finding that rarely reverses to normal [Fig. 4a]. The same is true for the peripapillary changes seen on retinography [Fig. 1b].[5]

To rule out optic neuropathy, the patient was submitted to magnetic resonance of the cranium and orbits, but no structural changes or optic nerve enhancement were observed.[4]

Our patient most likely experienced an isolated or chronic case of capillaropathy, considering the absence of inflammatory findings.[6]

No evidence-based treatment is available, and the list of differential diagnoses is long.[2,3] In any case, the condition is generally self-limited, followed by months of recovery.[5] Steroids may be used in some cases during the inflammatory stage.[5] Recurrence is rare.[4]

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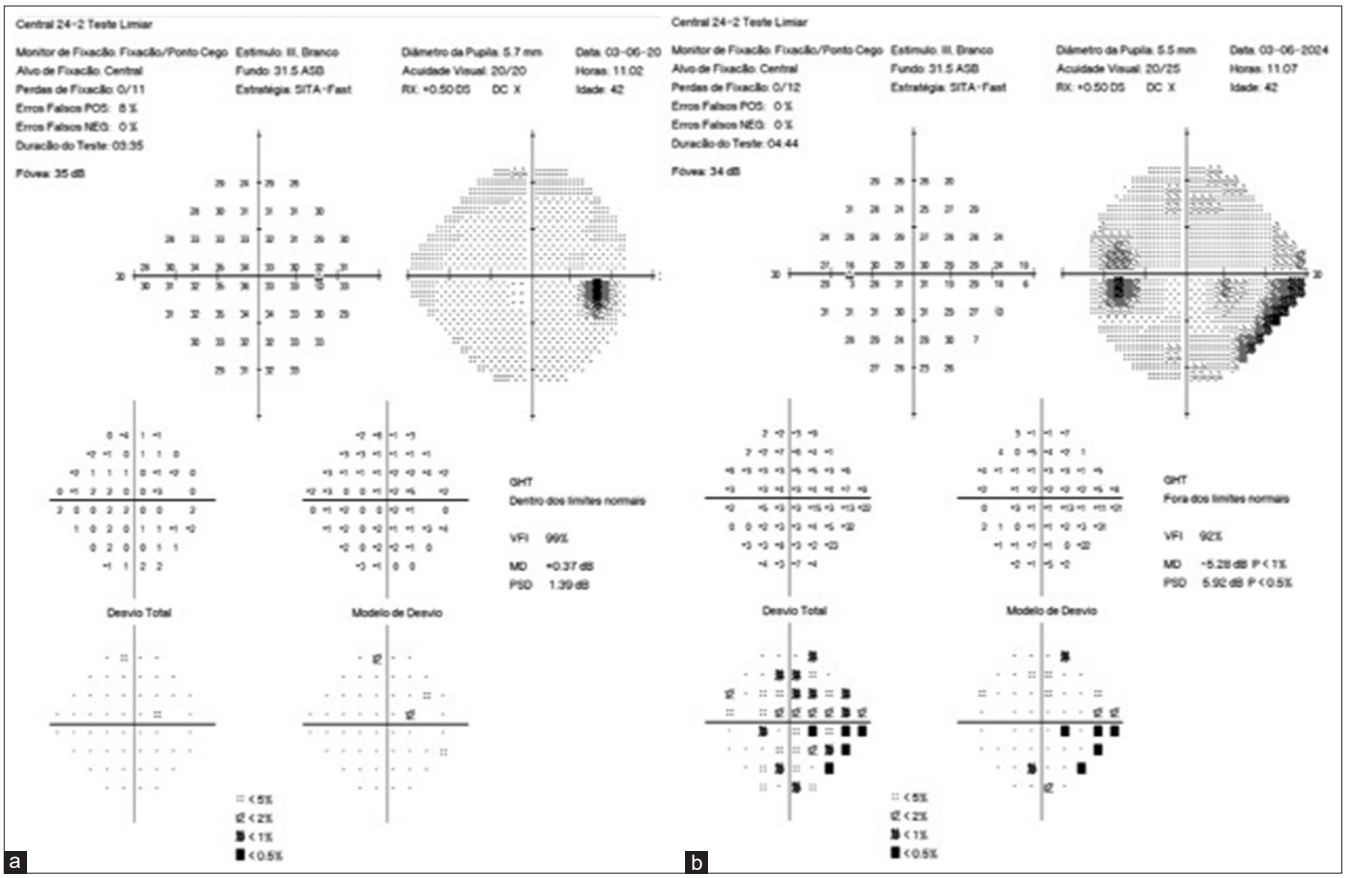

Figure 4: Automated perimetry: Testing in both eyes reliable. Normal findings in OD (a). Blind spot enlargement in OS (b). OD: Right eye, OS: Left eye

Authors contributions

RCSC was responsible for the literature search, data acquisition, data analysis, and preparation of the manuscript. AJDG and FCM contributed to the preparation and reviewing of the manuscript.

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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