Short communication

Acute idiopathic blind spot enlargement syndrome associated with choroidal neovascularization☆

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A B S T R A C T

Clinical case: A 17-year-old female presented with photopsia and a sudden loss of visual field in left eye (OS), with previous contralateral choroidal neovascularization. The examination suggested an acute idiopathic blind spot syndrome. The progress without treatment was favorable, with a reduction in the scotoma and without a worsening of her visual acuity.

Discussion: This case report is about an unusual and benign syndrome, typical of young women. Differential diagnosis must be made between the evanescent white dot syndrome and the acute zonal occult outer retinopathy. To our knowledge, this is the first published case associated with choroidal neovascularization, a fact that leads us to question its benignancy.

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Síndrome de aumento agudo idiopático de mancha ciega asociado a neovascularización coroidea

R E S U M E N

Caso clínico: Mujer de 17 años que consulta por fotopsias y escotoma en OI, con antecedente de neovascularización coroidea contralateral. La exploración sugiere un síndrome de aumento agudo idiopático de mancha ciega. La evolución sin tratamiento es favorable, con disminución del escotoma y sin empeoramiento de su agudeza visual.

Discusión: El síndrome de aumento agudo idiopático de mancha ciega es un síndrome raro y benigno, propio de mujeres jóvenes. Se debe hacer un diagnóstico diferencial con el síndrome de puntos blancos evanescentes y la retinopatía aguda zonal externa oculta. Tras revisar la bibliografía, no hemos hallado ningún caso descrito previamente en asociación con la neovascularización coroidea, hecho que nos obliga a cuestionarnos su benignidad.

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Introduction

The acute idiopathic blind spot enlargement (AIBSE) syndrome is an infrequent clinical condition affecting young women. It presents in acute form as photopsia and temporal unilateral scotoma condition, characterized by blind spots campimetric increase and late papillary hyperfluorescence in angiography (FA). It can involve relative afferent pupil defect and slight edema or papillary hyperemia, with peripapillary subretinal pigment changes in 50% of cases. Generally, full field electroretinogram (ERG) is normal although focal ERG is not. Visual acuity (VA) has good prognosis and improves spontaneously but the increased blind spots usually persist.

Clinical case

This case is of a 17-year-old female in follow-up by our service since age 11 due to idiopathic choroidal neovascularization (CNV) in the right eye (RE). In February 2011, the patient visited the emergency service due to photopsia and temporal black spots in vision in the left eye (LE). VA in RE was of 0.15 (−2.0 dioptries) and of 1.0 in LE (−0.75 dioptries). Anterior pole biomicroscopy and intraocular pressure were normal. The ocular fundus (OF) of RE exhibited old macular scars due to CNV, and no ophthalmoscopic alterations were detected in the LE (Fig. 1). Due to the non-specificity of the first exploration, a number of supplementary tests were performed. The visual field revealed increased blind spots in LE (Fig. 2). FA revealed late papillary hyperfluorescence in LE (Fig. 3), while FA with indocyanine green (ICG) in the LE was normal. Optic coherence tomography (OCT) of the LE macula was normal although the retina nerve fiber layer OCT revealed asymmetry with thickening in the LE nasal area (Fig. 4).

Fig. 1 – RE OF: sequels after choroidal neovascularization; and LE OF: without alterations.

Fig. 2 – RE CV: involvement due to previous macular scar and LE CV: increased blind spot.

Fig. 3 – FA showing macular scar in RE and late papillary hyperfluorescence in LE.

Multifocal ERG showed bilateral diffuse involvement which was more evident in the RE. Visual evoked potential (VEP) was requested and reported as delayed P100 latency more marked in RE. Other supplementary tests such as magnetic resonance, complete analysis with serology and chest X-ray were normal.

The patient evolved positively without treatment at 6 months follow-up with diminished visual field defect (Fig. 5) and no worsening of vision.

Discussion

In the presence of AIBSE, differential diagnostic should be considered with migraine, optic neuritis, multiple evanescent white dot syndrome (MEWDS) and acute zonal occult outer retinopathy (AZOOR). MEWDS is more likely due to being bilateral, with increased blind spots and ERG alteration in addition to exhibiting CNV as onset sign as described by Papadia. However, FA does not suggest the said syndrome. The case also fulfills some of the characteristics of AZOOR, but the typical retinal pigment epithelium alteration is absent.

Recently, some authors have demonstrated that idiopathic CNV appearing in young patients is generally of inflammatory origin and sometimes responds to systemic or pericocular steroids. Even though idiopathic CNV is generally a complication of inflammatory chorioretinopathy, it has been described as an onset sign in patients with MEWDS, multifocal choroiditis (MFC) or internal choroidopathy punctata (PIC) both ipsi- and contralateral. For this reason, Machida et al. proposed that idiopathic CNV could be a common initial stage for said
inflammatory diseases, particularly in young women with moderate-high myopia.

On the contrary, Volpe et al.\textsuperscript{4} suggested that diseases such as MEWDS, AIBSE, MCF, acute macular neuroretinopathy (AMN) and AZOOR could be the same entity with different presentations or even different stages of the same disease. This is supported by Gass\textsuperscript{5} when considering that these entities could share an identical and as yet unknown etiology and are secondary to the dysfunction of the photoreceptor external layer.

A review of references has not provided any described case of CNV as onset sign of AIBSE developed in the course of several years in the contralateral eye of the initial CNV.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES