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Case Report

Paracentral Acute Middle Maculopathy in Atrial Septal Defect

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Abstract

Paracentral acute middle maculopathy is an ischemic disease of the posterior pole of the retina characterised by the sudden appearance of a central scotoma caused by small infarcts in its inner layers. Optical coherence tomography highlights these lesions as typical hyperreflective bands involving the inner retinal layers. This report presents two cases of paracentral acute middle maculopathy associated with the presence of an atrial septal defect. The discovery of these retinal microinfarcts, especially in young patients with a history of migraine with aura, requires neurocardiological investigations to rule out the presence of an atrial septal defect and any cerebral ischemic lesions.

Keywords: Paracentral Acute Middle Maculopathy; Atrial Septal Defect; Patent Foramen Ovale; Migraine with Aura; Microembolism; Retinal Ischemia; Transcranial Color Doppler Ultrasound

Introduction

Paracentral Acute Middle Maculopathy (PAMM) is an ischemic disease of the posterior pole of the retina characterised by the sudden appearance of a central scotoma caused by small infarcts in its inner layers. In this report, I present two cases of PAMM in young subjects with Atrial Septal Defect (ASD).

Case Report

Case 1

A 35-year-old woman had been complaining for several days about the sudden appearance of a scotoma in the central visual field of her left eye. No other neurological symptoms were reported. She is in good general health and has not taken any medication except analgesics during episodes of Migraine with Aura (MA) from which she had been suffering for several years. Her Best Corrected Visual Acuity (BCVA) was 20/20 in both eyes. Bilateral intraocular pressure and anterior segment examination were normal. The fundus examination was normal in the right eye, while in the left eye there was a small orange lesion in the superotemporal paramacular area. Spectral Domain-Optical Coherent Tomography (SD-OCT) imaging of the left eye highlighted a hyperreflective band lesion at the level of the Inner Plexiform Layer (IPL) corresponding to the orange lesion observed during fundus examination (Fig. 1). The outer band of the retina (outer plexiform layer, outer nuclear layer and retinal pigment epithelium) appeared normal.

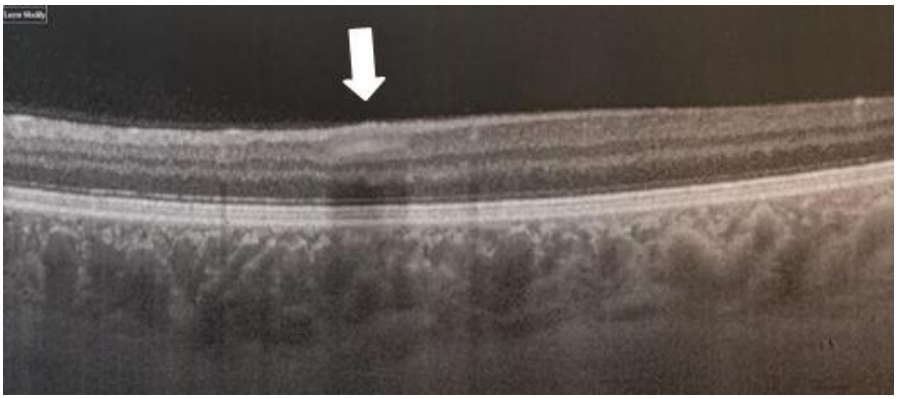
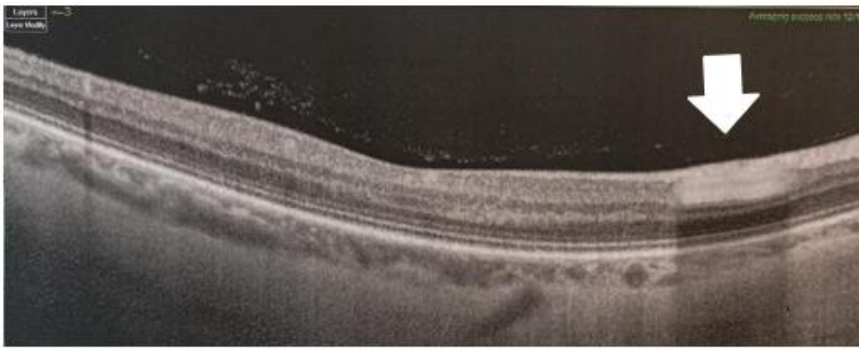


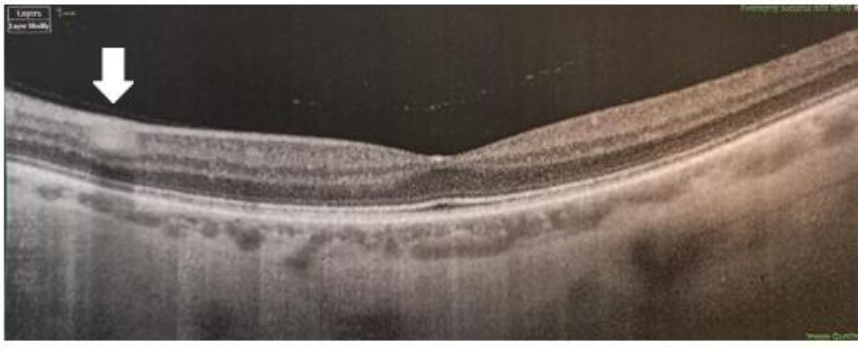
Figure 1: SD-OCT image of a typical hyperreflective band lesion at the IPL level in the left eye.

Case 2

A 38-year-old man complained of the sudden appearance of a scotoma in the central visual field of his right eye. A few days earlier, he had experienced a migraine preceded by an ophthalmic aura characterised by a scintillating scotoma lasting about 20 minutes. Again, the patient reported being in good general health. On ophthalmological examination his BCVA was 20/20 in both eyes. Bilateral intraocular pressure and anterior segment examination were normal. Fundus examination revealed a small orange lesion involving the superonasal macula of the right eye. At the level of the IPL, SD-OCT imaging demonstrated the typical hyperreflective band lesion in the right eye (Fig. 2) but also another small asymptomatic interpapillomacular lesion in the left eye (Fig. 2).



a



b

Figure 2: SD-OCT imagines of an hyperreflective band lesion (PAMM) at the IPL level in the right eye (a) and in the left eye (b). The "shadow cone" formed by the edematous thickening of the nerve fibers is clearly visible.

Management and Follow up

Haematochemical tests and instrumental investigations (carotid doppler ultrasound and encephalic magnetic resonance) revealed no abnormalities. On the contrary, echocardiogram revealed the presence of an ASD in both subjects, which was confirmed by Transcranial Colour Doppler Ultrasound with bubble test (TCDU). The retinal lesions were evaluated as PAMM and the two patients were followed without treatment. At 6-month follow-up, the black spots had completely disappeared and patient's visual acuity was normal. On examination of the fundus, the lesions were no longer evident, while SD-OCT revealed minimal retinal thinning in the areas corresponding to the lesions. The two patients consented to the publication of the images in this article.

Discussion

The retinal capillary vasculature serves the formidable role of supplying the metabolically active inner and middle retina. At the posterior pole, the retinal vasculature consists of three capillary plexuses: superficial, intermediate and deep [1]. The blood circulation of the retina is subject to self-regulation and, thanks to this mechanism, can compensate for oxygen deficiency. When this complex balance is disrupted, the retina can suffer ischemia which, if it occurs acutely, can cause infarcts that can take on the characteristics of PAMM. IPL, Outer Plexiform Layer (OPL) e Outer Nuclear Layer (ONL) are the ones with the highest metabolism. Furthermore, OPL and ONL constitute the so-called "watershed zone" at the border between retinal and choroidal circulation, thus representing the retinal layers most at risk of hypoxia. There are numerous factors that can predispose the onset of PAMM: central and branch retinal artery or vein occlusions, cilioretinal artery hypoperfusion, sickle cell retinopathy, hypertensive and diabetic retinopathy, dyslipidemia, pregnancy and migraine [2-4]. SD-OCT is the key tool for diagnosing PAMM [5]. Hyperreflective band lesions are the tomographic representation of post-infarction edema that thickens segments of nerve fibers in the retina (Fig. 1,2). Retinal lesions evolve into IPL thinning or atrophy and in the case of larger central lesions, visual impairment may persist [6]. Patent foramen ovale (PFO) and ASDs are two types of interatrial communications. The PFO is a normal part of fetal development that typically closes shortly after birth but may persist in as many as 25% to 30% of adults [7]. The communication between atria may result in paradoxic microembolism and embolic stroke [8]. PFO has also been proposed as a cardioembolic source in patients with MA [9]. PFOs are more prevalent in the MA (50%-60%) [9]. PFO and atrial fibrillation seem to be the most common sources of microembolism in the MA population [9]. In fact, during a migraine attack with aura, retina may undergo neurovascular decompensation characterised by spreading neuronal depression and vasoconstriction of the retinal capillaries, causing hypoxia and tissue necrosis especially at the posterior pole and in the deep retinal layers, where oxygen demand is greatest [6]. This phenomenon may therefore promote the onset of PAMM.

Conclusion

In the two cases described in this article, the cause of PAMM may be attributed to paradoxical microembolism aggravated by retinal hypoxia resulting from migraine attacks. The sudden appearance of a central scotoma in a subject with a history of MA should raise suspicion of PAMM. The diagnosis of PAMM requires careful examination of the fundus and SD-OCT, as both visual acuity and visual field examination may appear normal. The SD-OCT examination must be accurate, as small retinal lesions may not be detected by the operator. Finally, it is important to perform an echocardiogram and TDCU to check for the presence of a right- to-left shunt in ASD.

Conflict of Interest

The authors declare no potential conflicts of interest with respect to the research, authorship and/or publication of this article.

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