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Staphyloma-related chorioretinal folds

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ABSTRACT

Purpose: To report a case of bilateral idiopathic chorioretinal folds that seemed to be related to an atypical staphyloma.

Observations: A 49-year old man without medical history consulted for slight vision loss and metamorphopsia in the left eye. The ophthalmologic examination revealed moderate myopia and bilateral chorioretinal folds in the posterior pole, confirmed by multimodal imaging. Orbital and systemic examinations ruled out all the known etiologies of chorioretinal folds. 3-D optical coherence tomography and B-scan suggested that the folds were related to an atypical staphyloma that developed in the temporal part of the fundus, while sparing the peripapillary area. The peripapillary area, spared by the staphyloma process, appeared as a "dome-shaped disc" compared to the staphylomatous area.

Conclusion and importance: This case suggests that myopic patients with unusual staphyloma located outside the peripapillary area could develop chorioretinal folds.

1. Introduction

Chorioretinal folds (CRF) may be observed in many ophthalmic and orbital conditions, including malformative or compressive orbital disorders, thyroid-related orbitopathy, posterior scleritis, hypotonia, hyperopia, exudative macular degeneration, and optic nerve disorders.[1–6] We report the case of a patient with bilateral CRF and a very atypical staphyloma developed in the temporal part of the fundus, with relative sparing of the peripapillary area.

2. Case report

A 49-year old man was referred for blurred vision and mild metamorphopsia in his left eye (LE). His prior ophthalmologic examination had been performed 3 years earlier, the visual acuity was 20/20 in both eyes and the fundus examination was unremarkable according to his ophthalmologist. No imaging was performed at this time. He had no significant past medical history.

On examination, visual acuity was 20/20 in the right eye (RE) and 20/30 in the left eye (LE). He had moderate myopia with spherical equivalent of −4.25 D in the RE and −2.50 D in the LE. The axial length was of 24.5 mm in the RE and 23.6 mm in the LE. Intraocular pressure was normal in both eyes. Fundus examination revealed the presence of bilateral CRF (Fig. 1). Multimodal imaging was performed with a Topcon 50IA fundus camera (Topcon, Tokyo, Japan), Clarus fundus camera and PlexElite optical coherence tomography (OCT, Zeiss meditec, Dublin, Ca), Triton OCT (Topcon, Tokyo, Japan), and a HRA scanning laser ophthalmoscope (Heidelberg, Germany). The patient underwent OCT of the posterior pole (Fig. 2), autofluorescence imaging (Fig. 3) and fluorescein (Fig. 4) and indocyanine green (ICG) angiography. Multimodal imaging confirmed the presence of CRF, with horizontal folds, but did not show any associated condition. The orbital and systemic assessments, including thyroid hormones, brain and orbital MRI, and orbital B-scan, ruled out the usual disorders associated with CRF, including malformative or compressive orbital disorders, thyroid disorders, posterior scleritis, hypotonia, hyperopia, and optic nerve disorders. The 3D reconstruction of the posterior pole showed a marked slope between the peripapillary area and the temporal part of the fundus (Fig. 5). The B-scan also showed a change in curvature corresponding to a temporal atypical staphyloma (Fig. 6).

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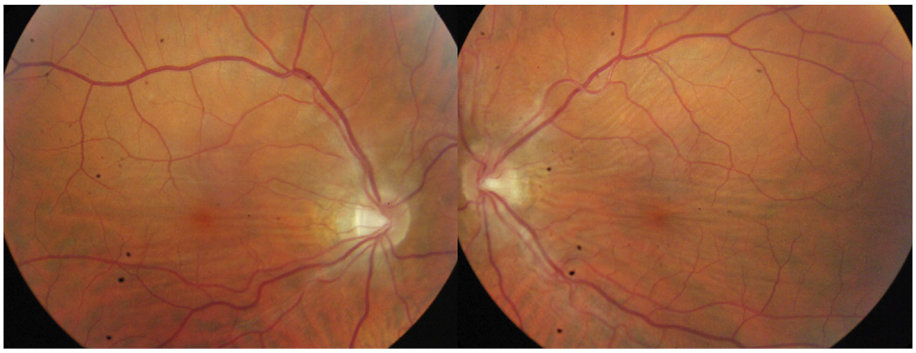

Fig. 1. 50° color fundus photography of the right and left eyes showing bilateral chorioretinal folds in the posterior pole.

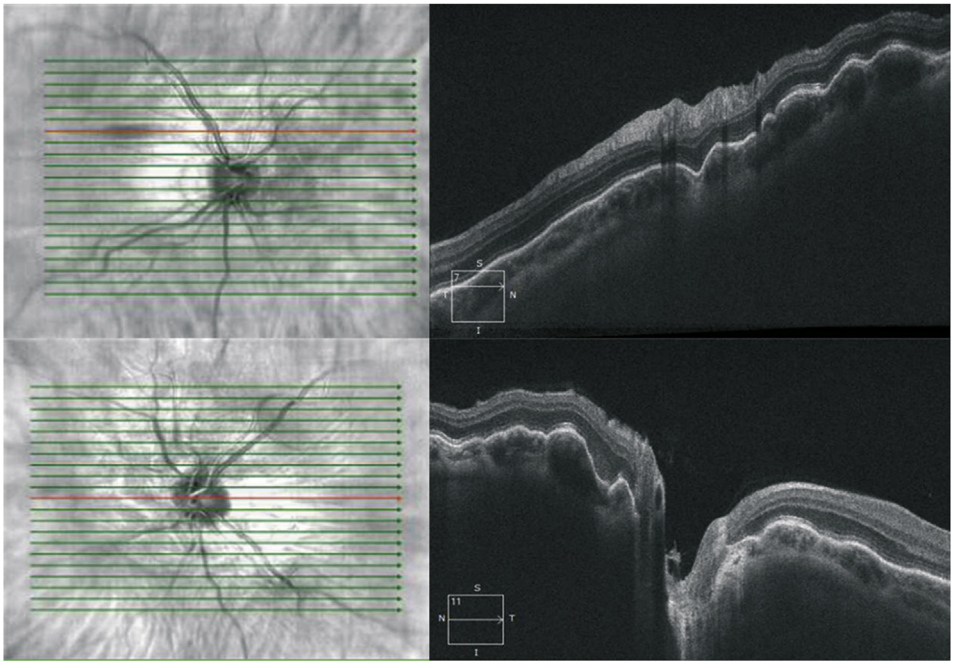

Fig. 2. Horizontal OCT B-scans of the right (top) and left (bottom) eyes showing bilateral chorioretinal folds, without obvious pachychoroid.

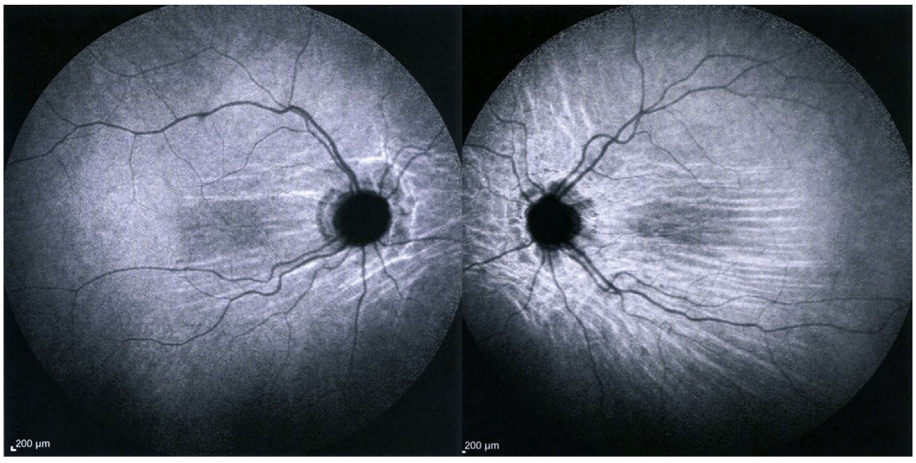

Fig. 3. Autofluorescence of the right and left eyes showing with a better contrast the bilateral horizontal chorioretinal folds as hypo- and hyper-autofluorescent lines in the posterior pole.

3. Discussion

We report a case of bilateral CRF responsible for metamorphopsia in one eye. Clinical examination ruled out hyperopia and hypotonia. Ultrasound and MRI ruled out posterior scleritis, and all local, compressive or malformative etiologies usually associated with CRF. Chorioretinal peripapillary folds have recently been described.[7] The authors have reported the folds as part of the pachychoroid disease

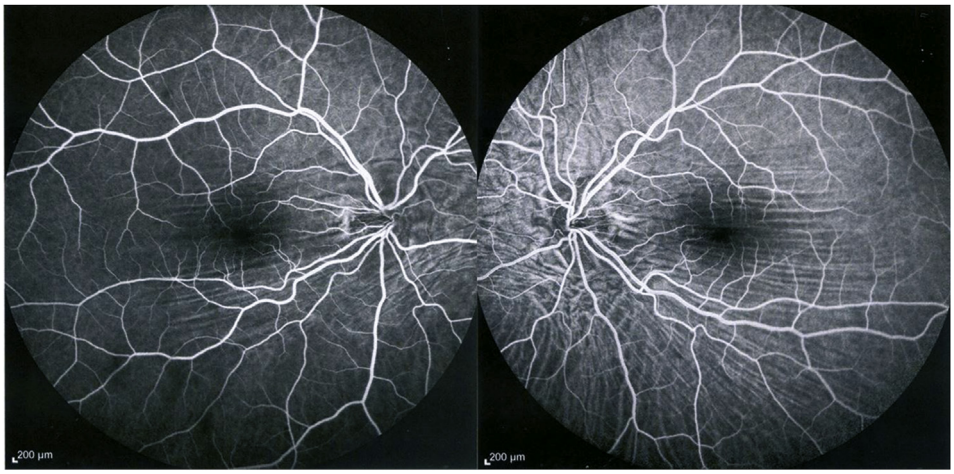

Fig. 4. Fluorescein angiograms of the right and left eyes showing an alternance of hypo- and hyperfluorescent lesions around the macula in both eyes and beneath the optic nerve on the left.

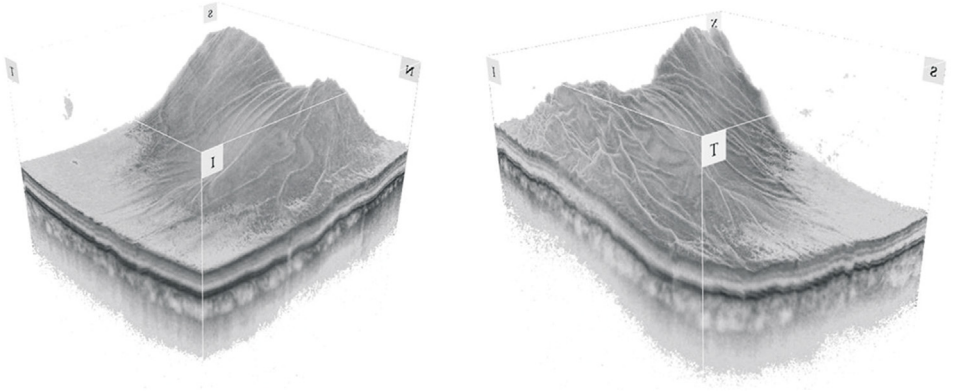

Fig. 5. 3-D reconstruction of the posterior pole of the right end left eyes showing a marked slope between the peripapillary area and the temporal part of the fundus.

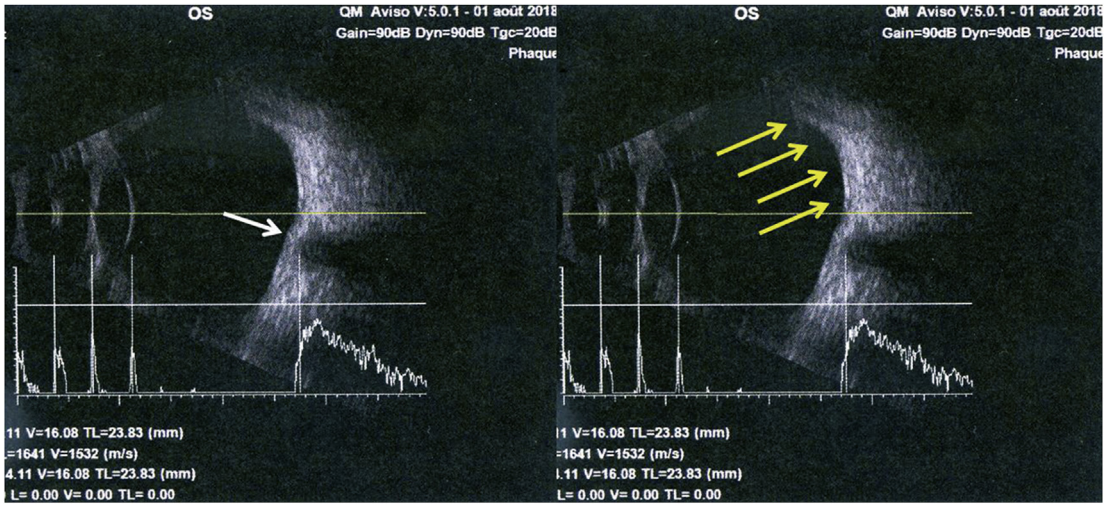

Fig. 6. B-scan of the left eye passing through the optic nerve showing that the optic nerve head is spared by the staphyloma (white arrow), while the staphyloma is located in the temporal part of the fundus, outside the peripapillary area (yellow arrows). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

spectrum, in a context of peripapillary pachychoroid syndrome (PPS). However, our case seemed different. Indeed, OCT did not show any thick choroid, and no dilated choroidal vessels were found in the Haller's layer. Furthermore, ICG angiography did not show any hyperpermeability of choroidal veins. In PPS, the folds seem to be related to irregularity of the RPE above dilated choroidal veins and above choroidal macrovessels.[8] On the contrary, in our case, horizontal and parallel folds were observed at the posterior pole.

Furthermore, this case does not match with any described cause of CRF, to the best of our knowledge. We could assume that these CRF

could be due to an atypical staphyloma. The progressive elongation of the posterior pole, without involvement of the peripapillary area, could have resulted in the development of CRF. A similar mechanism has been previously described in inferior staphylomas associated with tilted disc syndrome (TDS).[9] TDS is usually accompanied by an inferior staphyloma. An uneven growth of the eyeball, with progressive elongation of its inferior part has been suggested to explain the development of superior radial folds. Ishida T et al. have also reported choroidal folds radiating from the staphyloma edge in 6 out of 459 eyes (1.3%) with a posterior staphyloma on wide-field autofluorescence images. These CRF arose from the superior or supero-temporal staphyloma edge. In their study, 3D MRI images showed the presence of a notch along the superior or temporal edge of the outpouching, and the eye curvature flattened toward the steep edge of the outpouching.[10] This could explain why these folds may also be associated with T-shaped pigmentary changes in eyes with TDS.[11]

In our case, ultrasound examination (Fig. 5) confirmed the location of the staphyloma outside the peripapillary area. This atypical development of staphyloma could have changed the overall curvature of the eyeball, with relative preservation of the peripapillary area. This phenomenon could be close to what is observed in dome-shaped macula (DSM).[12–14] Indeed, in eyes with DSM, there is a relative sparing of the macular area, while staphyloma develop around it. The present case could thus be considered as a "dome-shaped disc". However, additional cases are needed to confirm our findings in order to describe a new condition, and to consider these folds as part of a new syndrome.

4. Patient consent

Written informed consent was obtained from the patient for publishing this case report and any accompanying images.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

The following authors have no financial disclosures related to this study: AGA, CL, AG, TG, SYC.

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