Short communication

Pseudo-vitelliform maculopathy and bilateral choroidal folds: Differential diagnosis

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A B S T R A C T

Clinical case: A 64-year-old woman. Best corrected acuity right eye (RE) 0.5 and 0.7 left eye (LE). Bilateral pseudophakia. No inflammatory signs. Normal IOP. RE fundus showed a rounded, yellow and excessive subfoveal deposit with positive autofluorescence. Multiple equatorial drusen and choroidal folds in both eyes. Fluorescein angiography of RE showed early foveal hypofluorescence and delayed hyperfluorescence. Optical coherence tomography revealed a hyperreflective deposit over the foveal epithelium pigment. Visual fields, ocular ultrasounds and electrooculograms (EOGs) were normal. Non-specific alterations in color tests.

Conclusion. Vitelliform maculopathy and choroidal folds are very rare diseases and, exceptionally, both appear together.

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Introduction

Vitelliform macular dystrophies are infrequent hereditary disorders, but their association to bilateral choroidal folds is highly unusual. A case report is presented in which an additional local or general underlying disorder was discarded.

Case report

Female, 64, without personal or familial antecedents of interest, who visited the practice because after bilateral cataract surgery one year back without surgical complications she did not notice significant improvement in the visual acuity (VA) of her RE. Best corrected VA in RE was of 0.5 and 0.7 in LE. Anterior pole exploration revealed bilateral pseudophakia, endoscopic intraocular lenses with transparent posterior capsule. Intraocular pressure (IOP) was of 14 mmHg without ocular or orbital inflammatory signs, ocular motility was normal and the patient did not refer ocular pain.

At the funduscopic level, the RE evidenced a yellowish-orange roundish and raised subfoveal deposit comprising 2/3 of the papillary diameter with abundant equatorial hard drusen in BE with choroidal folds in the posterior pole (Fig. 1).

Red-free light retinography revealed autofluorescence of the vitelliform macular deposit of the RE and the drusen in BE (Fig. 2). Fluorescein angiography (FA) showed hyperfluorescence of drusen in BE and of the choroidal folds. In the RE, the macula exhibited initial hypofluorescence without leaks with late contrast hyper-capture in the fovea (Fig. 3).

Optic coherence tomography (OCT) revealed a hyper-refringent deposit over the foveal retina pigment epithelium (RPE) of the RE (Fig. 4). Campimetric measurements were within normal ranges. The Farnsworth-Munsell 100 color test evidenced nonspecific alterations in the 3 axes, with 498 errors in RE and 396 in LE (Fig. 5). EOG revealed normal Arden indices in RE and LE of 3.08 and 3.36 respectively. The axial antero-posterior length was of 21.8 mm in RE and 22.17 mm in LE. Ocular echographies did not reveal pathological alterations (Fig. 6). After a follow-up of 4 years, the subfoveal roundish lesion of the RE progressively increased in size and reached the size of the papillary diameter without undergoing changes in color or thickness (378 µm). VA slightly diminished to 0.3 in RE while maintaining 0.7 in LE. The bilateral choroidal folds

Fig. 1 – Right eye fundus: raised yellow-orange subfoveal deposit, with abundant equatorial hard drusen. Left eye fundus exhibits abundant equatorial hard drusen. Choroidal folds in the posterior pole in both eyes.

Fig. 2 – Ocular fundus with red-free light: macular autofluorescence in right eye and of drusen in both eyes.
and drusen did not undergo changes. No associated systemic involvement occurred.

**Discussion**

Funduscopic findings lead to a differential diagnostic between the following disorders: adult vitelliform macular dystrophy, Best disease, confluent drusen, central serous chorioretinopathy (CSC) and the causes of the choroidal folds. Supplementary tests are essential for this diagnostic.

Adult vitelliform macular dystrophy generally debuts between the third and fifth decade of life, with a slight VA reduction. The funduscopic image can match that of the RE of our case and it is frequently associated to drusen. In addition, it exhibits autofluorescence compatible with lipofuscin of the vitelliform deposit, OCT hyper-refringence and normal EOG. In the early phase of FA, marked hypofluorescence of the late stages of the angiogram. Generally, adult vitelliform dystrophy involvement is bilateral although unilateral cases have been described. The color tests generally obtain scores below 400 errors.

This case could be a unilateral Best disease, although its expression in earlier periods of life and severely depressed EOG even in stages prior to macular involvement discard this possibility.

It could also be a deposit of confluent soft drusen in the context of ARMD. This diagnostic would be supported by the presence of drusen in both eyes. In addition, drusen are autofluorescent, but confluent drusen give a different image in OCT. FA discarded the presence of choroidal neovascular membranes.

Even though CSC is cloudy, even more so when it is liquid, it can give a macular image which is very similar to that observed in this case and it usually debuts in younger patients. In addition, in the OCT the liquid under the neurosensory retina or the RPE is usually hypo-refringent, in contrast with what was observed in this case.

Finally, we have discarded processes that could course with choroidal folds, such as ocular hypotony, hypermetropia, retrobulbar masses and globe compression, scleritis, thyroid disease, orbitary pseudotumor, orbitary cellulite or intracranial hypertension. The ocular fundus and oculor echography discarded the presence of tumor and/or choroidal detachment. OCT is able to differentiate between choroidal and

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**Fig. 3 –** Fluorescein angiography: hyperfluorescence of drusen and choroidal folds in both eyes without contrast leak although with late capture of right eye subfoveal deposit.

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**Fig. 4 –** Optic coherence tomography: hyper-refringent deposit over right eye foveal pigmented epithelium.
chorioretinal folds. The result was that we accepted the diagnostic of idiopathic or congenital choroidal folds.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES