VITELLIFORM MACULAR DYSTROPHY: ATROFIC FASE CASE REPORT

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PURPOSE

To describe a case of vitelliform macular dystrophy atrophic fase.

INTRODUCTION

Vitelliform macular dystrophy is aninherited macular dystrophy in which lipofuscin accumulates in the central macula causing progressive central vision loss. It characterized by bilateral yellow, yolk-like (vitelliform) macular lesions. While Best's presents during childhood, adult vitelliform typically presents later in life(1).

The material may persist, absorb, or break up and disperse at a late stage(2), developing a variety of subretinal and sub-RPE fibrosis, RPE atrophy, Geographic atrophy, choroidal neovascularization(3).

A good visual acuity (VA) is usually preserved and a more substantial visual loss occurs when there is complication(3).

OCT reveals that the vitelliform material appears as a dome-shaped, hyperreflective and homogenous lesion, located below the hyperreflective photoreceptor layer(1).

Fluorescein angiography (FA) shows central hypofluorescence surrounded by a small irregular hyperfluorescentring(2).

METHODS

Medical records review

CASE REPORT

A 57-years- old-woman referenced due to changes in the background of both eyes, low progressive vision for years. The best visual acuity is 20/200 in both eyes.

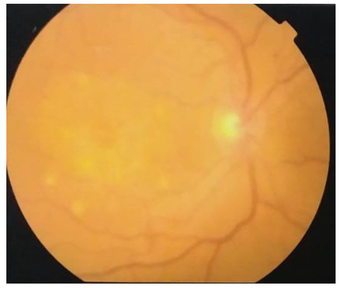
In fundus examination there is presence of yellow areas of viteliform content margining the area of macular atrophy in both eyes (Pictures 1A and 1B).

Complementary exams were requested for diagnosis

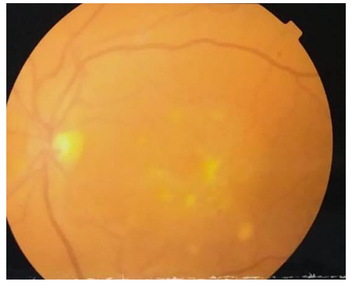
OCT with the presence of hyper reflective areas as sub RPE suggestive of vitelliform accumulation, subretinal hypo reflective area suggesting area atrophy after resorption vitelliform content and discontinuity of the photoreceptor layer (Pictures 2A and 2B). Autofluoresceinography with hyperautofluor in viteliform lesions associated with hipoautofluor in central atrophy area.

FA defecthy perfluor by defect in central window and hyperfluor by staining in paramacular viteliform injuries.

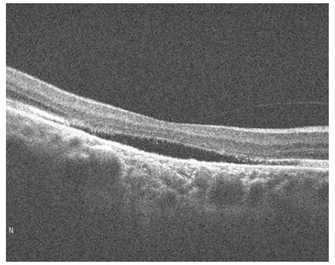
As a conduct, the patient's genes were dosed for diagnostic confirmation and semi-annual follow-up.



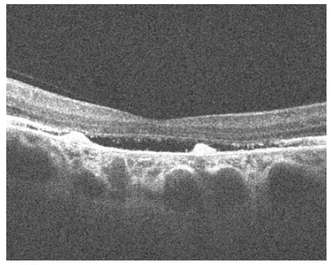
Pictures 1A



1B



Pictures 2A



2B

DISCUSSION

Vitelliform dystrophy is a relatively common retinal disorder with several differential diagnoses with variable prognosis and, in most cases, with treatment and improvement of the condition.

The need for genetic study assists in the diagnostic confirmation, however, regard less of confirmation, there is no therapy and clinical follow-up and guidance as to its prognosis and natural evolution of the disease are fundamental.

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