INTRODUCTION

Vitelliform macular dystrophy is a rare bilateral pathology with autosomal dominant mode of transmission. It has variable expression and penetrance, which affects the retinal pigmented epithelium (RPE). In electrophysiological studies, it shows changes in the electroculogram1. Among the atypical forms of the disease, those with asymptomatic presentation, whose diagnosis is made through electrophysiology and those with multiple lesions should be considered.

This rare entity with occasionally atypical presentation, as described in the report, makes its diagnosis more challenging. This demonstrates that it’s suspicion should always be remembered by ophthalmologists. In the Brazilian literature review, only two similar cases were registered3

CASE REPORT
E.E.L, 49 years old, male, from Olinda, attended the Emergency Service of the Altino Ventura Foundation with a complaint of progressive decrease in visual acuity in both eyes (AO) in 06 months, denied pain. He denied comorbidities. He presented better visual acuity (VA) of 20/200 in the right eye (RE) and 20/100 in the left eye (LE), but he had a medical record from 4 years prior with VA with better correction of 20/25 in the RE and 20/40 in the LE.

Fundoscopy showed multiple yellowish-white, rounded, subretinal lesions in the posterior pole and in the vascular arches with a finding of subretinal fluid (FSR) in a greater lesion in the AO foveal area (Figure 1). An optical coherence tomography (OCT) registered subretinal fluid in AO (Figure 2).

 Complete blood exam count was also requested to search for immunodeficiencies, which came up without abnormalities. Syphilis, tuberculosis, toxoplasmosis, cytomegalovirus and rapid HIV test were carried out.

About the ophthalmological findings, despite the non-typical lesions of Best's vitelliform macular dystrophy, this was introduced in the diagnostic hypotheses of the research arsenal. Therefore, the patient underwent an electroculogram (EOG) that presented a borderline pattern (1.7).

Currently, the patient is waiting for an electroretinogram to better elucidate the case, being already referred to the visual rehabilitation clinic to enhance the visual residue and the retina outpatient follow-up has been maintained.

DISCUSSION
Best's disease or Best’s vitelliform macular dystrophy is a rare bilateral pathology with an autosomal dominant mode of transmission. It presents itself with variable expression and penetrance. It is the second most common macular dystrophy, after Stargardt's disease. Onset may occur in childhood or decades later4.

It is generally characterized by the appearance of a single cystic macular lesion resulting from the accumulation of lipofuscin in the RPE cells, in the subretinal space and in the choroid5

The typical vitelliform lesion presents itself as a round, well-defined macular lesion (“egg yolk”) in the RPE, which varies in dimensions between 0.5-2.0 disk diameters; the size of the lesions and the stage of development in both eyes can be asymmetrical and sometimes only one eye is initially affected4. Occasionally, the condition may be extramacular and multiple, as described in this case report.

Currently, there is no treatment for this condition. The prognosis is generally good until middle age, from which point visual acuity declines in one or both eyes due to CNV (choroidal neovascularization), scarring or geographic atrophy4.

