

Unilateral Retinal Pigment Epithelium Dysgenesis (URPED): a very rare clinical disorder.



Beatriz Ribeiro¹, Ana Lúcia P. Peixoto², Ana Paula Couto¹, Emerson Monteiro¹, Adriana Muralha¹, Giovanna Provenzano¹, Elaine Castro¹, Raul N. G. Vianna¹.

- 1. Department of Ophthalmology, University Federal Fluminense (UFF), Niterói - RJ, Brazil.**
- 2. Instituto Brasileiro de Oftalmologia (IBOL), Rio de Janeiro - RJ, Brazil.**

PURPOSE

To report a very rare clinical disorder of a man with Unilateral Retinal Pigment Epithelium Dysgenesis (URPED) studied through a multimodal imaging approach including fundus photograph, fluorescein angiography (FA) and Fundus autofluorescence (FAF).

INTRODUCTION

URPED is a condition recently described in 2009 by Cohen et al after analyzing nine cases with a peculiar unilateral lesion located in posterior pole presents with central atrophy and peripheral fibrosis, along with hyperplastic changes in the retinal pigment epithelium. (1) Considering that, we showed a case of a asymptomatic man with URPED.

CASE REPORT

A 56-year-old man was referred to our Department. He did not complain of any visual symptoms and his past medical and ocular history were unremarkable. On examination, best corrected visual acuity (BCVA) was 20/20 in the right and 20/25 in the left eye. Slit-lamp examination findings were normal in both eyes. The right fundus appeared normal, but left fundoscopic examination revealed a peculiar well-circumscribed, unilateral, large yellowish-white lesion on the posterior pole that extended from the peripapillary region to the macula and inferior quadrant, including the inferior temporal vascular arcade. A multimodal evaluation of the retina was performed (Figure 1).

DISCUSSION

URPED is a unilateral condition that affects younger population. There are only about twenty cases reported in the literature. The lesion is in the RPE layer and gets its leopard-spot appearance. (2) fundus autofluorescence (FAF) and fluorescein angiography (FA) pictures provide an important contribution to URPED diagnosis, showing inverted images. Visual prognosis is still unclear and depends on development of complications, such as retinal folds, vitreoretinal disorders, macular atrophy and secondary choroidal neovascularization (CNV) (2). In this scenario, the almost pathognomonic differential features of URPED were best appreciate with multimodal evaluation.

BIBLIOGRAPHY

- Cohen SY, Fung AE, Tadayoni R, et al. Unilateral Retinal Pigment Epithelium Dysgenesis. *Am J Ophthalmol*; 148. Epub ahead of print 2009.
- Preziosa C, Staurenghi G, Pellegrini M. OPTICAL COHERENCE TOMOGRAPHY ANGIOGRAPHY FINDINGS IN A CASE OF CHOROIDAL NEOVASCULARIZATION SECONDARY TO UNILATERAL RETINAL PIGMENT EPITHELIUM DYSGENESIS TREATED WITH INTRAVITREAL BEVACIZUMAB THERAPY. *Retin Cases Brief Rep*. 2021 Sep 1;15(5):598-601.

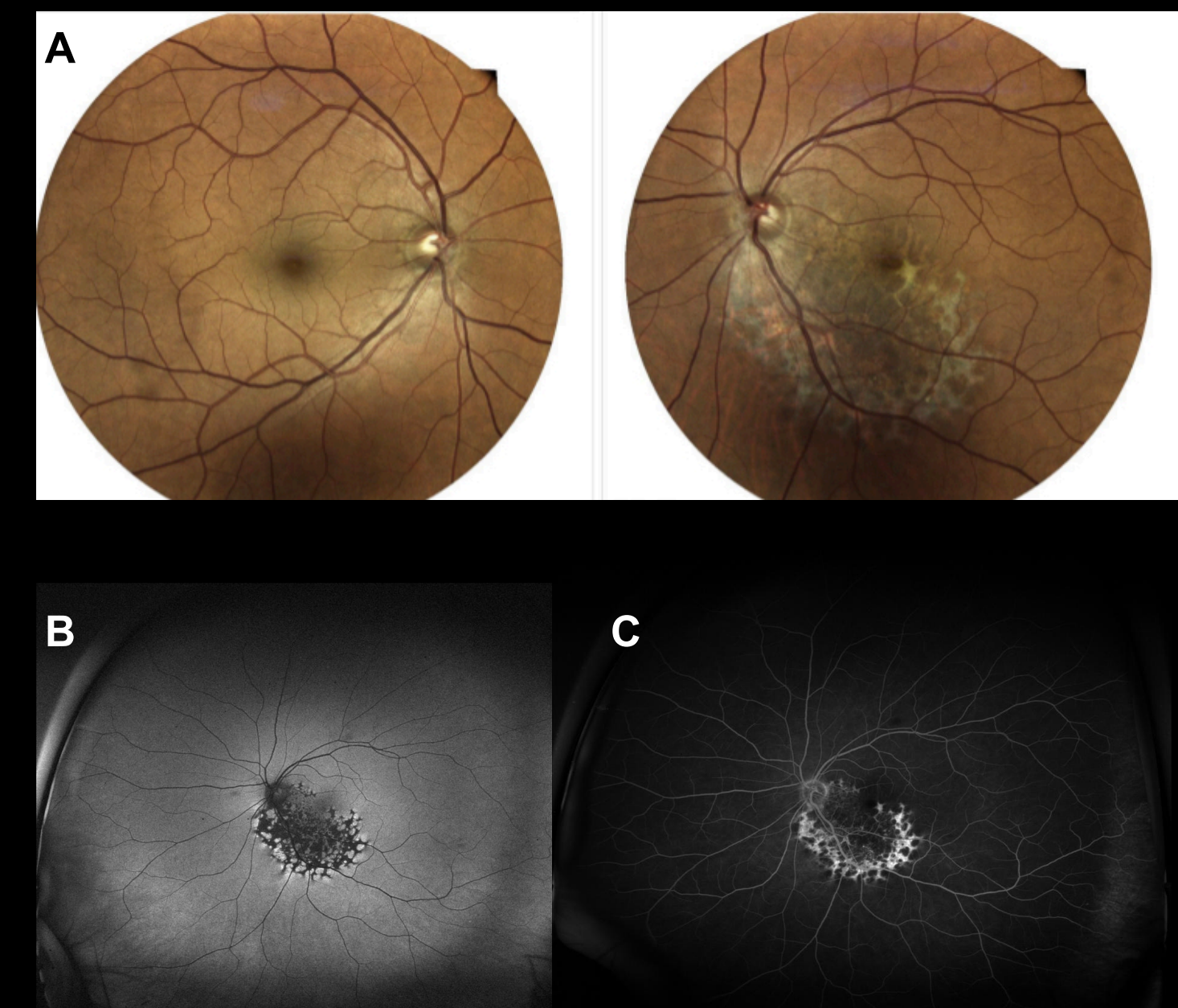


Figure 1. (A) Color retinography, (B) Fundus Autofluorescence (FAF) showing a hypoautofluorescent lesion with a hyperautofluorescent irregular margin that was inverted relative to (C) Fluorescein angiography.