Unilateral Retinal Pigment Epithelium Dysgenesis May Be a Bilateral Disease

Unilateral retinal pigment epithelium (RPE) dysgenesis is a rare entity with a poorly documented etiology, pathogenesis, and prognosis. It has a unique scalloped appearance on fluorescein angiography that is inverted on fundus autofluorescence (FAF) imaging. Associated epiretinal fibrosis, retinal detachment, or choroidal neovascularization can lead to vision loss. We report a case of RPE dysgenesis in which the patient had classic findings in only 1 eye but distinct, stellate RPE abnormalities in both eyes indicative of a bilateral, asymmetric disease. To our knowledge, bilateral RPE dysgenesis has not been previously described.

Report of a Case. A 35-year-old woman without significant medical history had a 6-year history of photopsias and dimmer vision in her right eye only. The patient's visual acuity was 20/25 OD and 20/20 OS. Anterior segment examination findings were normal in both eyes. Fundus examination of the right eye revealed a large, scalloped area of reticular RPE hyperplasia continuous with the optic nerve. Within the lesion, there was mottled atrophy of the RPE (Figure 1A and B). Humphrey examination findings of the left eye were normal.

Fluorescein angiography revealed patchy hyperfluorescence and hypofluorescence with an irregular margin of hyperfluorescence (Figure 1C and D). Imaging with FAF demonstrated a hypoautofluorescent lesion with a distinctive, reticular border that was inverted relative to fluorescein angiography. There was mild hyperautofluorescence at the edge of the lesion. Interestingly, there...
Figure 1. Fundus photographs, fluorescein angiograms, and fundus autofluorescence images. A, Fundus examination of the right eye shows a lesion with an appearance typical of unilateral retinal pigment epithelium dysgenesis. B, The left eye appears grossly normal on fundus examination. C, Fluorescein angiography of the right eye demonstrates a lesion with a hyperfluorescent irregular margin (arrow). D, Fluorescein angiography findings of the left eye are normal. E, Fundus autofluorescence imaging of the right eye highlights the distinctive appearance of the lesion on the right, with scattered multifocal areas of retinal pigment epithelium loss. Arrow indicates inversion of fluorescence compared with fluorescein angiography. F, In the left eye, the multifocal stellate lesions not visible on clinical examination are now appreciated on fundus autofluorescence imaging (arrows).
were also bilateral, multifocal, stellate areas of hypoautofluorescence that were not appreciated clinically in either eye (Figure 1E and F).

Optical coherence tomography through the lesion in the right eye demonstrated outer retinal thinning with associated attenuation of the inner segment–outer segment junction (Figure 2). Full-field electroretinography findings were normal in both eyes.

Comment. The first report of unilateral RPE dysgenesis was published in 2002.1 In this report, 3 young men and 1 woman aged 16 to 34 years were noted to have a round leopard-spot lesion contiguous with the optic nerve. In 2009, the typical characteristics of unilateral RPE dysgenesis were described in a set of 9 affected patients, 6 males and 3 females aged 14 to 42 years.2 The margin of the lesion is pathognomonic, with a scalloped reticular fringe of mild fibrosis and atrophy with inverted FAF imaging relative to fluorescein angiography, and is identical to the lesion in our patient.

The differential diagnosis of this lesion includes acute zonal occult outer retinopathy. However, acute zonal occult outer retinopathy typically has a smooth curvilinear border on autofluorescence3 and the electroretinographic findings are abnormal. Alternative differential diagnoses include traumatic retinal pigment epithelopathy and combined hamartoma of the retina and RPE.4

This case is unique in that it shows bilateral loss of the RPE in a multifocal, scattered, stellate fashion, best appreciated with FAF imaging. To our knowledge, this is the first reported case of unilateral RPE dysgenesis with abnormal findings on FAF imaging in the fellow eye. In the previously reported series of 9 patients by Cohen et al,1 only 3 cases were evaluated with FAF imaging and no evidence of bilateral disease was seen. However, as most patients in the case series did not have FAF imaging performed, some of these unilateral cases may have had subclinical bilateral disease.

The etiology of this condition remains elusive. It may be an RPE dysgenesis or dystrophy, or it could reflect previous inflammatory, infectious, or autoimmune insult to the RPE. It is intuitive that a dysgenesis should ultimately be bilateral, and this is the first report to our knowl-

edge illustrating this for RPE dysgenesis, which perhaps should no longer be termed unilateral.

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Levofloxacin-Associated Panuveitis With Chorioretinal Lesions

Drug-induced uveitis is a rare complication of many commonly prescribed medications, including bisphosphonates, sulfonamides, multiple vaccines, and topical β-blockers.1 Recently, a uveitis-like syndrome with iris transillumination defects and pupillary mydriasis associated with oral moxifloxacin use has been described.2 Fluoroquinolones are an increasingly recognized cause of bilateral uveitis,3 although the visual significance is often minimal. Herein, we describe a visually disabling but reversible manifestation of levofloxacin-associated panuveitis.

Figure 2. Optical coherence tomography. A, Optical coherence tomography through the right eye demonstrates outer retinal thinning with associated attenuation of the inner segment–outer segment junction with a corrugated appearance to the superficial aspect of the inner retina. B, Optical coherence tomography through the left eye shows normal findings in comparison. I indicates inferior; N, nasal; S, superior; and T, temporal.