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ORIGINAL ARTICLE

Spontaneous retinal pigment epithelium tear in geographic atrophy

Déchirures spontanées de l'épithélium pigmentaire rétinien (EPR) en atrophie géographique

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Age-related macular degeneration;
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Retinal pigment epithelium;
Tear

Summary
Purpose. — To report two cases of spontaneous retinal pigment epithelial (RPE) tears occurring in two patients affected with geographic atrophy (GA) due to non-exudative age-related macular degeneration (AMD).
Case report. — Two patients (a 79-year-old man and a 71-year-old woman) presented to our department with progressive visual loss. The man had a best-corrected visual acuity (BCVA) of 20/100 in the right eye (RE) and 20/50 in the left eye (LE); the woman had a BCVA of 20/200 in the RE and 20/160 in the LE. Upon complete ophthalmologic examination, revealing a large area of atrophy (> 175 μm in diameter) along with pigmentary changes, calcified drusen and no choroidal neovascularization (CNV) in either eye, the patients were diagnosed with GA due to non-exudative AMD. Interestingly, the imaging modalities performed, including fluorescein angiography (FA), indocyanine green angiography (ICGA) and spectral-domain optical coherence tomography (SD-OCT), clearly highlighted the presence of spontaneous RPE tears in the context of non-exudative AMD, while in general, RPE tears are a well-recognized complication of exudative AMD.
Conclusions. — To our knowledge, this is the first description of spontaneous RPE tears as a possible complication of GA due to non-exudative AMD.

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MOTS CLÉS
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Décollement de l'épithélium pigmentaire (rétinien) ;
Épithélium pigmentaire rétinien ;
Déchirure

Résumé
Objectif. — Nous présentons deux cas de déchirures spontanées de l'épithélium pigmentaire rétinien (EPR) chez deux patients affectés d'atrophie géographique due à une dégénérescence maculaire liée à l'âge (DMLA) sèche.
Case clinique. — Deux patients (un homme de 79 ans et une femme de 71 ans) ont été adressés à notre service avec une baisse progressive de vision. L'homme avait une meilleure acuité visuelle corrigée (MAVC) de 20/100 à l'œil droit (OD) et de 20/50 à l'œil gauche (OG) ; la femme avait une MAVC de 20/200 à l'OD et de 20/160 à l'OG. Lors d'un examen ophtalmologique complet, montrant une grande zone d'atrophie (>175 μm de diamètre) avec de changements pigmentaires, drusen calcifiés et pas de néovascularisation choroïdienne (NVC) dans les deux yeux, les patients ont été diagnostiqués d'atrophie géographique due à une DMLA sèche. Les examens d'imagerie effectués (angiographie en fluorescence, angiographie au vert d'indocyanine, tomographie en cohérence optique) ont clairement mis en évidence la présence des déchirures spontanées de l'EPR dans le contexte de la DMLA sèche, alors que généralement les déchirures de l'EPR sont une complication reconnue de la DMLA humide.
Conclusion. — À notre connaissance, il s'agit de la première description de déchirures spontanées de l'EPR comme complication d'une atrophie géographique due à une DMLA sèche.

Introduction

Retinal pigment epithelium (RPE) tears are well-recognized complications of exudative age-related macular degeneration (AMD). In most cases, the RPE tear is part of the natural history of pigment epithelium detachment (PED) that has developed as a result of occult choroidal neovascularization (CNV). Spontaneous RPE tears were first described as a complication of PEDs by Hoskin et al. [1]. Since then, RPE tears have been described spontaneously in association with central serous retinopathy, angioid streaks and trauma [2]. RPE tears also represent a complication that develops either spontaneously or after various treatments for exudative AMD, such as laser photocoagulation, photodynamic therapy (PDT) and intravitreal anti-vascular endothelial growth factor (VEGF) drugs [3—5]. Clinically, a well-demarcated area of bare choroid is visible immediately adjacent to a hyperpigmented area, which represents the redundant and retracted RPE. Patients usually report sudden and severe loss of vision, and in most cases, RPE tear is associated with a poor visual prognosis (often 20/200 or less) even if it does not involve the subfoveal region [6,7].

Geographic atrophy (GA) as the result of non-exudative form of AMD is defined by any sharply delineated round or oval area of at least 175 μm in diameter, characterized by hypopigmentation or apparent absence of RPE. We report two cases of spontaneous RPE tears occurring in two patients affected with GA due to non-exudative AMD. To our knowledge, spontaneous RPE tears have never been described in this form of non-exudative AMD.

Report of cases

Case 1

A 79-year-old man presented to our department with progressive visual loss. Best-corrected visual acuity (BCVA) was 20/100 in the right eye (RE) and 20/50 in the left eye (LE). Upon a complete ophthalmologic examination, showing a large area of atrophy (>175 μm in diameter) along with pigmentary changes, calcified drusen, and no choroidal neovascularization (CNV) in both eyes, the patient was diagnosed with GA due to non-exudative AMD.

Interestingly, fluorescein angiography (FA) and indocyanine green angiography (ICGA) of the LE showed at the posterior pole a large area of early hyperfluorescence (Fig. 1A and C) that became more intense in the late phases (Fig. 1B and D) due to a window defect, characterized by a lozenge of central inhomogeneous hypofluorescence, suggesting a ripped, corrugated and retracted RPE with no signs of CNV (Fig. 1A—D, arrows). Spectral-domain optical coherence tomography (SD-OCT) scan clearly documented the presence of GA and of a RPE tear in the context of non-exudative AMD (Fig. 1E).

Case 2

A 71-year-old woman presented to our department with progressive visual loss in both eyes. BCVA was 20/200 in the RE and 20/160 in the LE. Upon a complete ophthalmologic examination, showing a large area of atrophy (>175 μm in diameter) along with pigmentary changes, calcified drusen, and no CNV in both eyes, the patient was diagnosed with GA due to non-exudative AMD.

Interestingly, fundus autofluorescence showed at the posterior pole a large area of hypoautofluorescence located superiorly to the macula, whose inferior border was characterized by variations in the degree of autofluorescence, like from a curled or redundant RPE layer (Fig. 2B). FA and ICGA of the LE showed superiorly to the macula a large area of early hyperfluorescence (Fig. 2C and E) that became more intense in the late phases (Fig. 2D and F) due to a window defect, and a central inhomogeneous hypofluorescence, suggesting a ripped, corrugated and retracted RPE with no signs of CNV (Fig. 2C—F, arrows). SD-OCT scan clearly