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SHORT COMMUNICATION

Curvilinear pigmentary lesions in a rod-cone dystrophy

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PURPOSE. To report a peculiar curvilinear pigmentary lesion in the peripheral fundus in a rod-cone dystrophy.
METHODS. Observational case report. Fundus examination of a 57-year-old woman who was known to have a generalized rod-cone dystrophy since she was 8 years old.
RESULTS. The peripheral fundus examination revealed a curvilinear lesion which resembles a well-known finding associated with a presumed ocular histoplasmosis syndrome or multifocal choroiditis.
CONCLUSIONS. The differential diagnosis of a peculiar curvilinear pigmentary lesion in the peripheral fundus may be expanded to include a generalized rod-cone dystrophy. (Eur J Ophthalmol 2005; 15: 641-2)

KEY WORDS. Curvilinear change, Retinal pigment epithelium, Rod-cone dystrophy

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INTRODUCTION

A peculiar curvilinear pigmentary lesion in the peripheral fundus is a well-known finding associated with a presumed ocular histoplasmosis syndrome or multifocal choroiditis (1-4). We show a case of a rod-cone dystrophy with similar changes.

Case report

A 57-year-old woman who was known to have a generalized rod-cone dystrophy since she was 8 years old was noted on clinical examination to have a peculiar curvilinear pigmentary lesion in the fundus of both eyes, characteristic of an antecedent inflammatory disease. Electrophysiologic examination in 1997 showed a severely reduced cone function and an extinguished rod response in each eye consistent with a rod-cone dystrophy. Her best-corrected visual acuity was 10/400 in each eye in 2004 when she was referred to our clinic. The peripheral fundus examination revealed widespread and zonular areas of retinal pigment epithelial and atrophic and pigmentary disturbances (Fig. 1). The curvilinear pigmentary lesion was found inferotemporal quadrant in both eyes (Fig. 1), more prominently evident in the right eye (Fig. 2). There was no remarkable vitreous adhesion, pigment dust, or fibrillary degeneration. Nuclear cataract was found in both eyes but no posterior subcapsular opacification. The curvilinear lesion resembles a well-known finding associated with a presumed ocular histoplasmosis syndrome or multifocal choroiditis. However, there was no acute or previous history of intraocular inflammation in this patient.

DISCUSSION

Peripheral curvilinear pigmentary lesions are known to occur in the presumed ocular histoplasmosis syndrome (1, 2) and multifocal choroiditis (3, 4). However, to our knowledge, this entity has not been previously described

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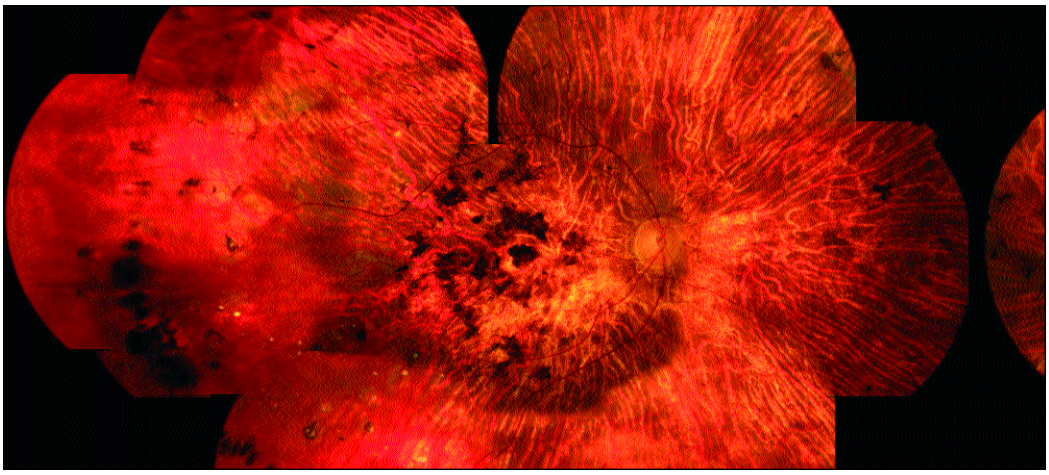

Fig. 1 - Fundus color photographs of the right eye (left) and the left eye (right) reveal zonular areas of retinal pigment epithelial loss in both eyes.

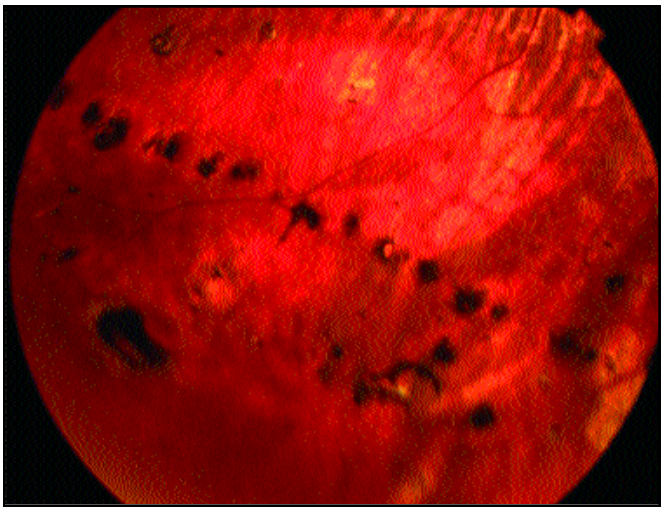

Fig. 2 - Fundus photograph in the periphery of the right eye shows peculiar pigment epithelial clumping in a curvilinear fashion.

in a patient with a known generalized rod-cone dystrophy. Our patient developed progressive retinal changes and these hyperpigmented curvilinear tracks without evidence of previous ocular inflammation (5). The mechanism for the production of these hyperpigmented spots is not known. Some possible explanations, although purely speculative, include the following: stimulation of the cells secondary to interactions between the retina and the vitreous base, serous or rhegmatogenous detachments that can cause pigmentary formation, and finally, reactive hyperpigmentation due to inflammation within the retina. Even if this pigmentary manifestation in the fundus took decades to evolve, recognition of the lesion is relevant to clinical ophthalmology since it may lead to a better recognition of the association in a rod-cone dystrophy. Based on our report, the differential diagnosis of peripheral curvilinear pigmentary lesions may be expanded to include a generalized rod-cone dystrophy.

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