Correlation of Functional Impairment and Morphological Alterations in Patients With Group 2A Idiopathic Juxtafoveal Retinal Telangiectasia

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Objective: To correlate functional impairment with morphological alterations in patients with group 2A idiopathic juxtafoveal retinal telangiectasia.

Methods: As part of the Macular Telangiectasia Project, a cohort of 10 patients underwent additional functional testing and imaging studies including photopic and scotopic fine matrix mapping, microperimetry, reflectance, and autofluorescence imaging with scanning laser ophthalmoscopy.

Results: From clinical stage 2 to 5, scotopic central function was reduced, which corresponded to depletion of macular pigment density. From clinical stage 3 onward, severe photopic and scotopic scotomata with up to 30 dB of loss were found next to fixation and were not totally confined to abnormalities seen with standard imaging modalities. The number of test points with loss of 10 dB or more was significantly greater for scotopic testing than for photopic testing ($P = .007$, Wilcoxon signed rank test).

Conclusions: Rod function may be more severely affected than cone function in patients with group 2A idiopathic juxtafoveal retinal telangiectasia, and this may occur early in the disease progression. Severe reduction in retinal sensitivity is spatially confined to morphological alterations seen with scanning laser ophthalmoscopy imaging. The findings imply that idiopathic juxtafoveal retinal telangiectasia is not solely a vascular disease and that early neuronal involvement may be implicated in the pathogenesis of the disease.

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IDIOPATHIC JUXTAFOveal RETINAL telangiectasia (IJT) is a rare condition causing progressive visual loss. It usually manifests with a slow decrease in vision, metamorphopsia, positive scotoma, and reading difficulties.1-7

According to the classification introduced by Gass and Blodi2 in 1993, group 2A is the most common type of IJT. Occult, bilateral involvement with juxtafoveal telangiectasis, minimal exudation, superficial retinal crystalline deposits, and right-angle venules characterize this manifestation of IJT. Late in the course of the disease, hyperplastic pigment plaques and retinal neovascularization may occur. Based on fundus photographs and fluorescein angiography findings, Gass and Blodi divided group 2A IJT into 5 clinical stages according to the disease development. More recent studies of patients with group 2A IJT using optical coherence tomography (OCT) examination have revealed intraretinal cystoid spaces without foveal thickening and disruption of the photoreceptor inner segment–outer segment junction early in disease evolution, whereas later stages showed foveal thinning and outer retinal atrophy.1,8-13 Preliminary data using 2-wavelength autofluorescence imaging indicate that macular pigment density (MPD) is significantly reduced in the central retina.14

These recent findings have provided increasing evidence that group 2A IJT is not a disease limited to the retinal vasculature. However, to date, little is known of the functional consequences of cystoid spaces or the irregular distribution of macular pigment and their relationship with telangiectatic retinal vessels. It is also still unknown why telangiectatic vessels are usually first found in the temporal parafoveal area and which retinal layers and cells are primarily affected. A gradual decrease in central visual acuity has been reported, but no systematic studies on the spatial correlation of morphological alterations and functional impairment are available to our knowledge. Overall, the pathogenesis of group 2A IJT remains unclear and there is no treatment available to halt or alter the progression of this disease.15

In this study, we performed functional testing and imaging studies on a cohort of patients with group 2A IJT to better understand the functional implications of morphological changes with regard to both cone and rod photoreceptor system involvement.