Optical coherence tomography angiography in Leber hereditary optic neuropathy

Figure Characterizing the progression of Leber hereditary optic neuropathy using optical coherence tomography angiography and fundus photography of a patient's left eye

(A, B, D) Note acute phase optic disc hyperemia and circumpapillary telangiectasias. (C, E, F) Three months after presentation, note significant capillary dropout in the papillomacular bundle and obvious disc cupping.

A 17-year-old man had acute vision loss OS (visual acuity 20/40), having lost vision OD months before (20/400). There were cecocentral scotomas OU. Mitochondrial DNA testing revealed a T14484C mutation, suggestive of Leber hereditary optic neuropathy (LHON), which results in painless sequential vision loss. Acutely, there is hyperemia of the optic nerve, circumpapillary microangiopathy, and dilated, tortuous vasculature¹; the pathognomonic finding is nerve swelling without leakage on fluorescein angiography. Chronically, the optic nerve develops atrophy, sometimes with cupping.² Optical coherence tomography angiography provides rapid, high-resolution visualization of retinal vasculature without dye injection, and is a novel imaging modality in LHON (figure).

Joaquin O. De Rojas, MD, Nailyn Rasool, MD, Royce W.S. Chen, MD, Jason Horowitz, MD, Jeffrey G. Odel, MD
From Columbia University Medical Center, New York, NY.

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Correspondence to Dr. Chen: rc2631@cumc.columbia.edu

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