Teaching *NeuroImages*: Chiasmal enlargement and enhancement in Leber hereditary optic neuropathy

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Figure Retinal photography and MRI of the chiasm

(A) Funduscopy reveals bilateral elevation and hyperemia (pseudoedema) of the optic discs. (B) T2 fluid-attenuated inversion recovery images show asymmetrical chiasmal hyperintensities and enlargement. (C, D) Axial and sagittal gadolinium-enhanced T1-weighted images show chiasmal enhancement (arrows).

A 19-year-old man was admitted for subacute severe painless bilateral loss of vision. The patient was known to be a healthy carrier of the mitochondrial G11778A mutation, with a family history of Leber hereditary optic neuropathy. Funduscopy revealed bilateral optic disc elevation and hyperemia (figure). Goldmann visual fields demonstrated large central scotomas in both eyes. CSF disclosed normal opening pressure and cytochemical examination. Unexpectedly, brain MRI revealed an asymmetrical chiasmal enlargement and enhancement (figure). Similar cases have rarely been reported.^{1,2} These findings would suggest disruption of the blood–brain barrier at the early

stage of the disease,² likely due to an inflammatory-like mechanism.

AUTHOR CONTRIBUTIONS

Elodie Ong: drafting/revising the manuscript, analysis or interpretation of data, accepts responsibility for conduct of research and final approval, acquisition of data. Damien Biotti: drafting/revising the manuscript, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and final approval, acquisition of data, study supervision. Lucie Abouaf: drafting/revising the manuscript, accepts responsibility for conduct of research and final approval, acquisition of data. Guy Louis-Tisserand: analysis or interpretation of data, accepts responsibility for conduct of research and final approval, acquisition of data. Caroline Tilikete: drafting/revising the manuscript, study concept or design, accepts responsibility for conduct of research and final approval, study supervision. Alain Vighetto: drafting/revising the manuscript, analysis

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