Teaching NeuroImages: Scleral thickening and optic disc edema from glycosaminoglycan deposition in Hunter syndrome

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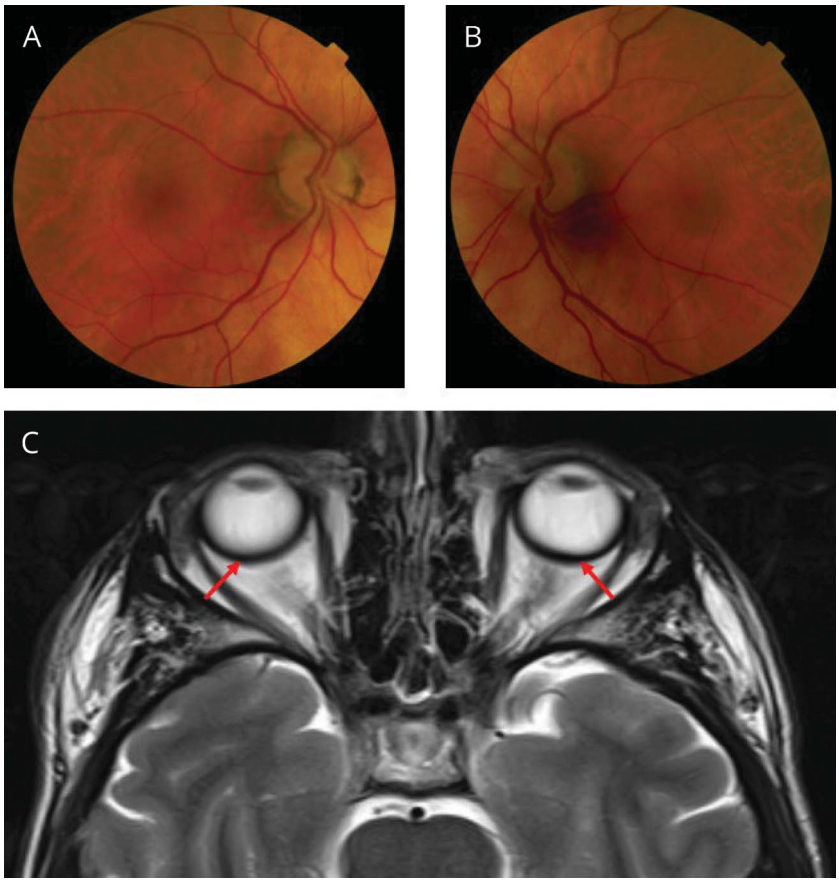
Figure Fundus photographs and MRI



Fundus photographs demonstrate waxy pallor of the right optic nerve (A) and optic disc edema of the left eye (B). (C) Axial T2-weighted MRI with symmetric hypointense signal circumferentially along the globes most notable along the posterior margins indicates thickened sclera from glycosaminoglycan deposition.

A 43-year-old man with a history of Hunter syndrome presented to the neuro-ophthalmology clinic with decreased peripheral vision. Computerized visual field testing revealed bilateral ring scotomas. The right optic disc was flat and there was left optic disc edema. Optical coherence tomography revealed disc edema of the left nerve, inner microcystic changes in the nasal fovea of the left eye, and parafoveal atrophy of the outer retinal layers in both eyes. MRI demonstrated posterior ocular globe thickening likely secondary to glycosaminoglycan deposition in the sclera (figure), known to occur in Hunter syndrome.[1] The mechanism of the optic disc edema in our patient could have been from compression of the optic nerve at the scleral opening.

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Author contributions

Meagan D. Seay: drafting/revising the manuscript, data acquisition, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and final approval. Heather Lau: drafting/revising the manuscript, data acquisition, study concept or design, analysis or interpretation of data, accepts responsibility for conduct of research and final approval, acquisition of data. Steven L. Galetta: drafting/revising the manuscript, accepts responsibility for conduct of research and final approval.

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Disclosure

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Reference

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