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. 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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Reference
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