Teaching NeuroImages: Superior segmental optic nerve hypoplasia confirmed by optical coherence tomography

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A 36-year-old woman without complaints was referred for abnormal visual fields (figure 1A). She had 20/20 vision in both eyes and a right afferent pupillary defect. Fundus examination suggested superior segmental optic nerve hypoplasia (SSONH), a congenital optic nerve disorder (figure 1B). Optical coherence tomography (OCT) of the optic nerve, which measures nerve fiber layer thickness using interferometric techniques,1 showed decreased thickness of the superior segment of both nerves (figure 2). In this case, OCT provided a noninvasive and reliable method for confirmation of SSONH suspected by inferior arcuate visual field defects and optic nerve appearance.2

REFERENCES

Figure 1 Humphrey visual field of the patient’s right and left eye

(A) Visual fields for the right eye (top) and left eye (bottom) revealed inferior arcuate defects consistent with superior segmental optic nerve hypoplasia. (B) Optic nerve photographs of the right eye (top) and left eye (bottom) showed evidence of superior segmental optic nerve hypoplasia.
Optical coherence tomography of the patient’s retinal nerve fiber layer of the right eye (A) and left eye (B) revealed superior thinning compared to normal distribution. Right and left eye comparison (C) demonstrated more superior nerve fiber layer thinning in the right eye compared to the left eye.
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