

# Retinal Hemangioblastoma and Syrxinx in von Hippel–Lindau Disease

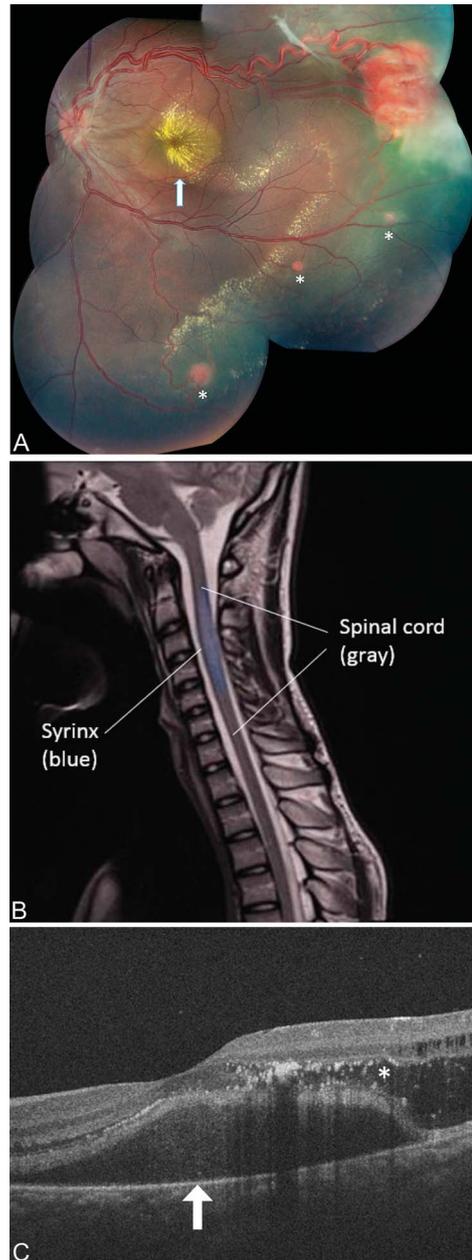
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A 14-year-old boy presented with decreased vision in his left eye for 4 weeks. Review of systems was otherwise negative.

Best-corrected visual acuity was 20/20 on the right eye and 20/100 on the left eye. Ophthalmoscopy of the right eye revealed a normal fundus and optic nerve; evaluation of the left eye revealed an exudative retinal detachment with a macular star (arrow, Figure 1A). A reddish-orange vascular tumor emanated from the tortuous superior arcade of the left eye (Figure 1A). The inferotemporal retina contained 3 smaller lesions (asterisks, Figure 1A) with associated exudation. Magnetic resonance imaging revealed several enhancing spinal cord nodules and a cervical spine syrinx (blue shading, Figure 1B). Spectral domain optical coherence tomography of the macula showed macular exudation (asterisk, Figure 1C) and retinal detachment (arrow, Figure 1C). Von Hippel–Lindau disease can present with profound visual disturbance secondary to macular exudation and retinal detachment because of retinal hemangioblastoma.

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In a prospective study describing retinal manifestations of von Hippel–Lindau disease, Dollfus et al<sup>1</sup> found 22% of eyes with retinal detachment and 15% with macular exudation. Kim, et al<sup>2</sup> describes treatments for retinal hemangioblastomas, smaller tumors (<0.5 mm) treated with laser photocoagulation, moderate tumors (0.5–3 mm) treated with transpupillary thermotherapy, and large tumors (>3 mm) treated with cryotherapy and transpupillary thermotherapy. Sequelae of central nervous system hemangioblastoma can include pseudocyst, syrinx, and increased intracranial pressure. Syrinx may present



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**Fig. 1.** **A.** Color fundus photograph of the left eye showing exudative retinal detachment (arrow), a vascular tumor along the superior arcade, and three lesions located inferotemporally with associated exudation (asterisks). **B.** Magnetic resonance imaging showing a syrinx in the cervical spine (blue shading). **C.** Optical coherence tomography showing exudative retinal detachment (arrow) and macular exudation (asterisk).

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None of the authors have any financial/conflicting interests to disclose.

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with deficits in pain and temperature sensation in a cape-like distribution.

Cyst formation has been associated with central nervous system hemangioblastoma,<sup>3</sup> and this may play a role in the exudative retinal complications in retinal hemangioblastoma. A complete systemic evaluation is needed to rule out tumors of the retina, brain and spine, kidney, pancreas, and adrenal glands, which were later ruled out in our patient. Laser photocoagulation was applied to the retinal hemangioblastomas with subsequent tumor regression.

**Key words:** retinal hemangioblastoma, von Hippel-  
**AU2** Lindau disease.

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### References

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