# A 25-Year-Old Man with Asymptomatic Retinal Capillary Hemangioma

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## Introduction:

An apparently healthy man underwent a dilated fundus exam during a screening for a family history of von Hippel-Lindau disease. The patient's father was treated for a renal cell tumor, his brother a pheochromocytoma.

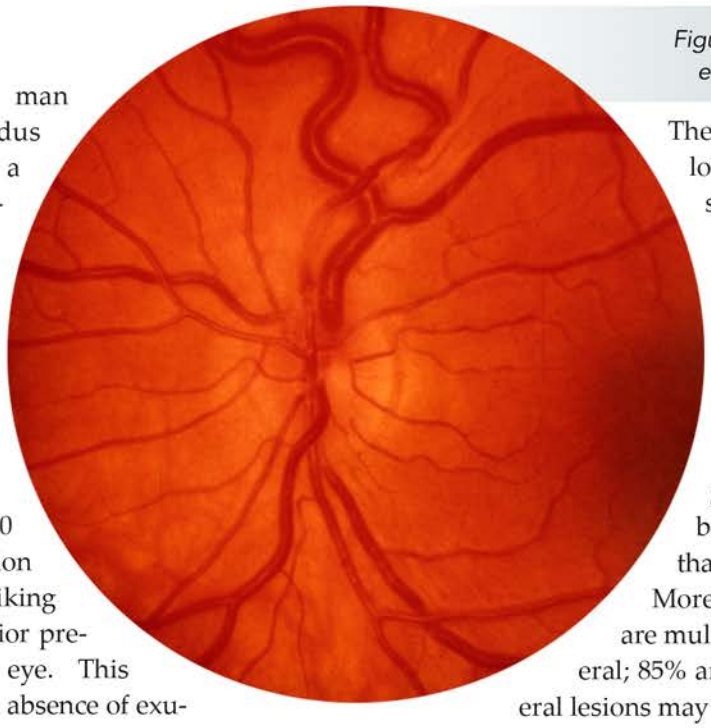

*Figure 1: Angioma with dilated efferent and afferent vessels.*

## Examination:

The patient's acuity was 20/20 in both eyes. His examination was normal except for a striking vascular tumor in the superior pre-equatorial retina of the left eye. This lesion was remarkable for an absence of exudation and dilated afferent and efferent vessels that extended posteriorly to the optic nerve.

## Discussion:

Von Hippel-Lindau disease (VHL) is a dominantly inherited familial cancer syndrome predisposing to a variety of malignant and benign neoplasms, most frequently retinal, cerebellar, renal cell carcinoma, pheochromocytoma, spinal hemangioblastoma, and pancreatic tumors. The incidence of VHL is about 1:36,000. Autosomal dominantly inherited with penetrance almost complete by the age 65 is the rule. The germline mutation has been localized to a tumor suppressor gene mapped to chromosome 3p25. Retinal tumors, present in about 50% of patients with VHL, are frequently the first presentation.

Retinal capillary hemangioma is a lesion that often begins as a red dot, no larger than a diabetic microaneurysm. More than 2/3 of hemangiomas are multiple. Sixty percent are bilateral; 85% are peripheral. Small peripheral lesions may be difficult to detect, but are readily identified on fluorescein angiography. These lesions may remain stable for years, but these high flow shunts may lead to vision loss from progressive intraretinal and subretinal exudation. In untreated eyes, rhegmatogenous and exudative retinal detachments are possible.

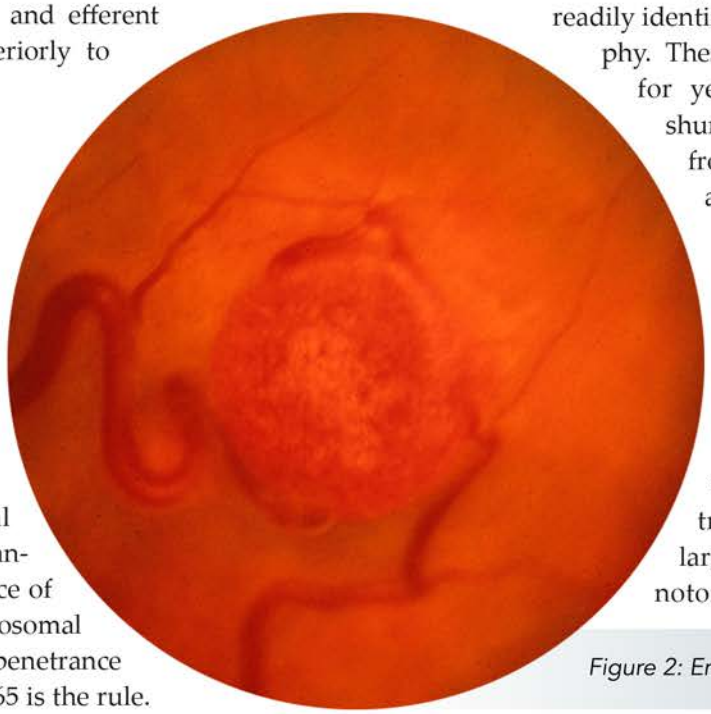

*Figure 2: Engorged superior disc vessels.*

## Treatment:

Small lesions are easy to treat successfully; whereas, large hemangioblastomas are notoriously difficult. Lesion

size, location and presence of exudate determine the choice of treatment. Lesions less than one disc diameter in size are most suitable for photocoagulation. Transcleral cryotherapy is more suitable for larger lesions. Two or three freeze-thaw cycles are recommended making certain the freeze encompasses the entire tumor.

Our patient underwent a triple freeze-thaw cryopexy in a single session under a general anesthetic. A post-treatment angiogram several weeks later determined what appears to be complete closure of the angioma.

## Conclusion:

The clinical features of VHL are diverse. The diagnosis of VHL can be made when multiple tumors are present or when an isolated tumor is identified in an individual with a positive family history. A multidisciplinary approach to systemic screening of families is invaluable to detect asymptomatic lesions requiring treatment.

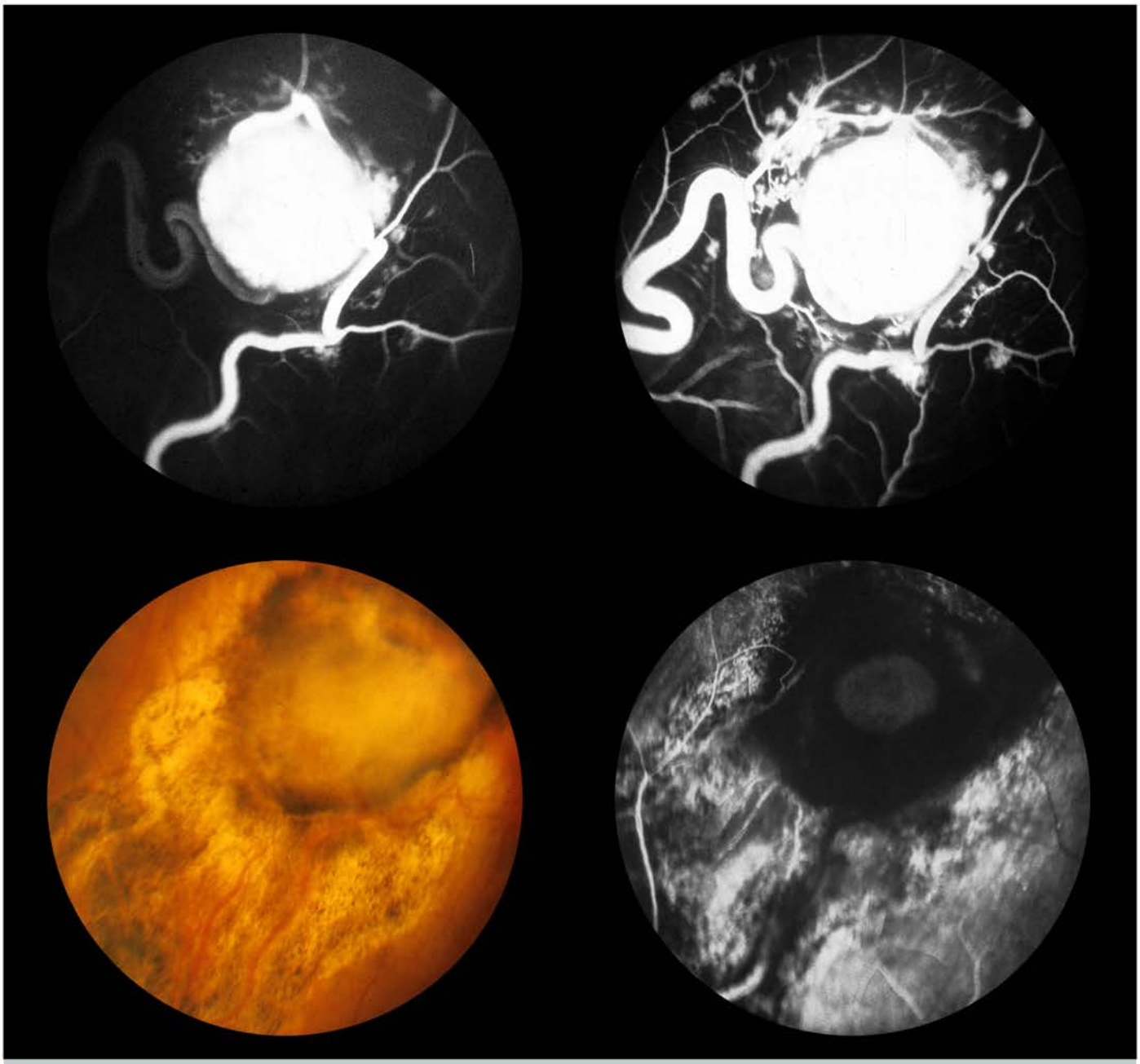

*Figure 3: (top left) Telangiectasia surrounding angioma. Figure 4: (top right) No obvious leakage. Figure 5: (bottom left) Post-cryo. Figure 6: (bottom right) Satisfactory ablation.*

## References:


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Wong WT, Agrón E, Coleman HR, et al. Clinical characteristics of retinal capillary hemangioblastomas in a large population with von Hippel-Lindau disease. Ophthalmology 2998; 115:181-186.

Palmer JD, Gragoudas ES. Advances in treatment of retinal angiomas. Int Ophthalmol Clin. 37(1997); pp: 150-170.


Eugen
von Hippel
1867 – 1939

Arvid
Lindau
1892 – 1958

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