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Retinal hemangioblastoma

Retinal hemangioblastomas tend to be located peripherally, and may hemorrhage and cause retinal detachment. Erythrocytosis may be due to erythropoietin liberated by the tumor.

- a) Retinal hemangioblastomas occur in > 50% of Von Hippel-Lindau disease patients. Mean age of presentation: 25 years
- b) frequently bilateral, multifocal, and recurrent
- c) often asymptomatic. Visual symptoms occur with progressive growth, edema, retinal detachments, and hard exudates
- d) typically located in the periphery and near or on the optic disc
- e) microangiomas measuring a few hundred microns without dilated feeding vessels maybe located in the periphery
- f) retrobulbar HGB are rare (5.3% in NIH cohort)
- g) severity of optic disease correlates with CNS and renal involvement
- h) early diagnosis and treatment with laser photocoagulation, and cryotherapy can prevent visual loss. Low dose external XRT may be an option for refractory cases

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Last update: 2024/06/07 02:51

