JOURNAL OF CLINICAL CASE STUDIES, REVIEWS AND REPORTS

Case Report
Volume 5 | issue1

Juxtapapillary Capillary Hemangioma in A Patient With Suspected Von Hippel-Lindau Disease - Clinical Case and Treatment Review

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Submitted: 16 February 2026 **Accepted:** 20 February 2026 **Published:** 26 Febraury 2026

Citation: Casas Camila (2026) Juxtapapillary Capillary Hemangioma in A Patient With Suspected Von Hippel-Lindau Disease - Clinical Case and Treatment Review. J of Clin Case Stu, Reviews & Reports 5(2), 01- 06.

Abstract

Background

Retinal capillary hemangiomas (also known as hemangioblastomas or retinal angiomas) are proliferations of capillaries in the retina, which can be found in extrapapillary or juxtapapillary locations. They may or may not be part of the autosomal dominant neoplastic syndrome known as Von Hippel-Lindau. We present the case of a 19-year-old patient with a juxtapapillary retinal capillary hemangioma in her right eye, for whom a systemic diagnosis was sought. The therapeutic challenge arose due to the tumor's location and the associated exudation with serous foveal detachment. Among the available therapies in this case, we considered initiating treatment with oral propranolol and anti-VEGF and/or corticosteroid therapies to reduce tumor size and/or the exudation.

Clinical Case

A 19-year-old female patient presents with blurred vision in her right eye (OD) of two years' duration. Her past medical history includes a six-month episode of tachycardia, interpreted as an anxiety attack, which went untreated. She has no other relevant medical history. On ophthalmological examination, her visual acuity was 20/20 in the right eye (OD) and 20/20 in the left eye (OS).

Biomicroscopy was unremarkable. Intraocular pressure was 10 mmHg in the right eye and 11 mmHg in the left eye (OS). Fundus examination in the left eye revealed an orange mass with feeding vessels in the right eye (OD), juxtapapillary, with serous retinal detachment at the macular and peripapillary levels, delimited by lipid exudation and surrounded by whitish mottling. The left eye (OS) showed similar peripheral characteristics, associated with pigment clumps.

Fluorescein angiography was performed, revealing early hyperfluorescence with late filtration. Ultrasound and optical coherence tomography (OCT) confirmed the tumor with associated serous drainage.

The diagnosis was juxtapapillary capillary hemangioma. Due to the patient's medical history and age-related risk factors, a consultation with internal medicine was requested to rule out other tumors associated with Von Hippel-Lindau disease.

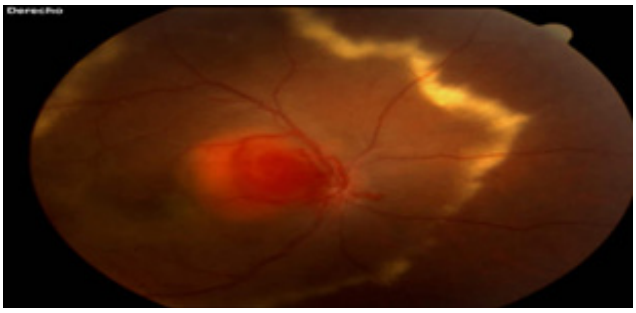

Figure 1: Retinography of the right eye: orange tumor with dilated vessels, corresponding to the juxtapapillary retinal capillary hemangioma (RCH) with exudation that involves the entire posterior pole, surrounded by a corona of lipid exudation

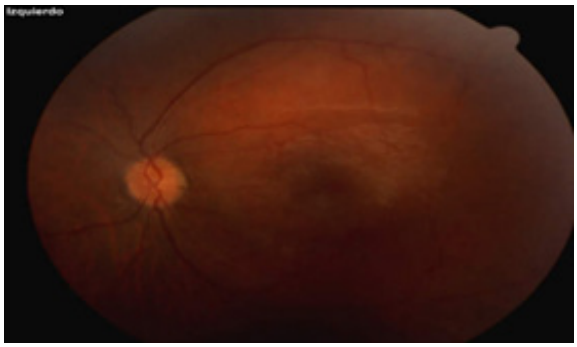

Figure 2: Retinography of the left eye: Posterior Pole Without Particularities

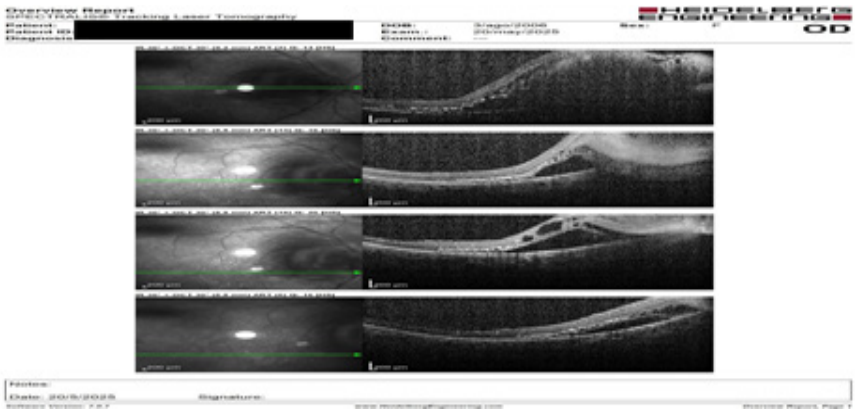

Figure 3: Macular OCT Demonstrates Retinal Capillary Hemangioma (RCH) Tumor Associated with Neuroepithelial Detachment along with hard Exudates and Cystoid Macular Edema.

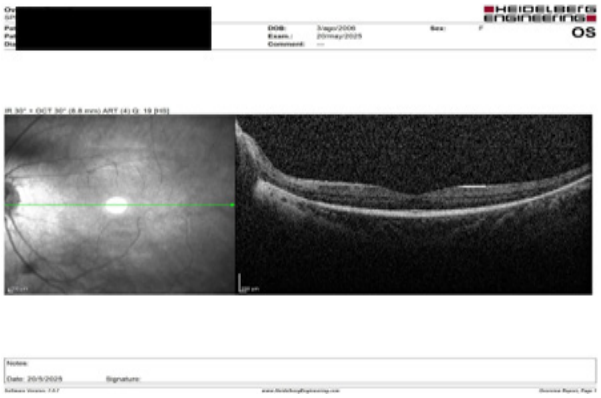

Figure 4: Macular OCT OI without Alteration of Retinal Layers with Preserved Anterior and Posterior Profile Next to the Foveolar Contour.

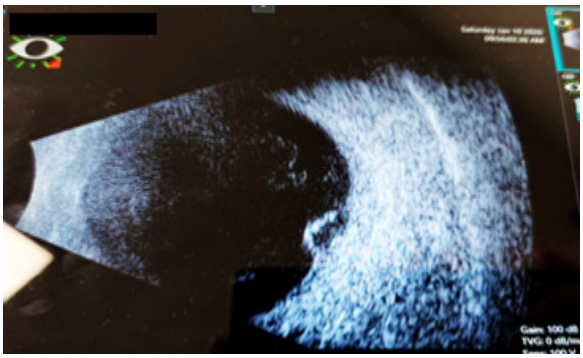

Figure 5: Ocular Ultrasound Showing a Tumor with Hypoechoic Content Compatible with Serous Detachment

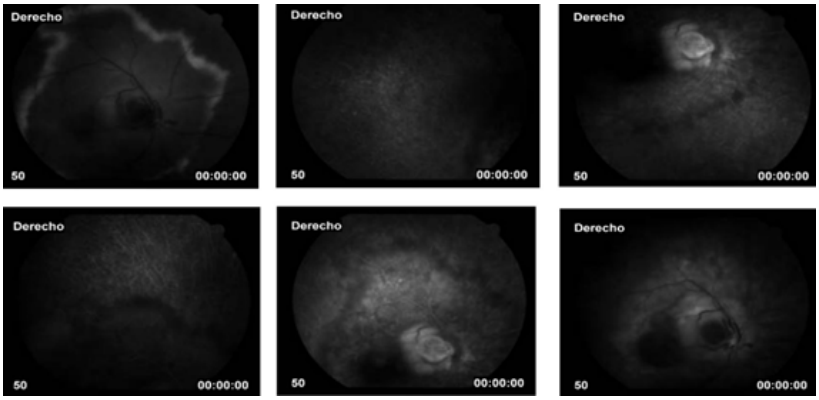

Figure 6: Fluorescein Angiography Showing Early Hyperfluorescence at the Site of Retinal Capillary Hemangioma (RCH) with late Leakage.

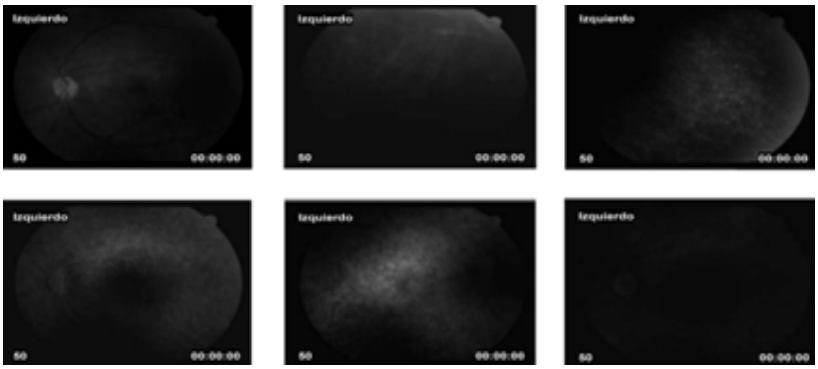

Figure 7: Fluorescein Angiography of the Left Eye Without any Particularities

Discussion

Von Hippel-Lindau disease is an autosomal dominant neoplastic syndrome resulting from a germline mutation in the VHL gene, leading to the development of various benign or malignant tumors and/or cysts in several organ systems. Affected individuals may develop lesions in the central nervous system (CNS), including cerebellar, spinal cord, brainstem, nerve root, and supratentorial hemangioblastomas, as well as retinal hemangioblastomas and endolymphatic sac tumors. In other organ systems, lesions include renal cysts and carcinomas, pheochromocytomas, pancreatic cysts, and neuroendocrine tumors, as well as epididymal and broad ligament cystadenomas [1].

Its incidence is approximately one in 36,000 live births, and it has a penetrance rate greater than 90% by age 65. The main causes of death are usually complications related to renal cell carcinomas and hemangioblastomas of the central nervous system [2,3].

Retinal capillary hemangioma (RCH) is an angiomatous hamartoma that histopathologically corresponds to the proliferation of capillaries that alters the architecture of the retina. These lesions most frequently exhibit endophytic growth and are predominantly located peripherally and superotemporally, although juxtapapillary hemangioma is described in 11-15% of VHL cases [3]. RCHs are also called retinal angiomatosis, retinal hemangioblastoma, von Hippel disease, and von Hippel tumor [4].

The various screening protocols agree on beginning ophthalmological screening annually from age 5 in those with suspected VHL disease and every 6 months in patients diagnosed with it, generally up to age 60. They also recommend routinely performing systemic studies to minimize the risk of missing new tumors [8].

Crypto-retinal hemangiomas (CRHs) can grow in an endophytic or exophytic pattern. The endophytic type is the most common. Visual loss can be due to several causes, including exudative or tractional phenomena, the development of epiretinal membranes, and vitreous hemorrhage. If a CRH is located far from the optic nerve and exudation is prominent, the differential diagnosis should include telangiectasias, Coats' disease, Leber's granulomatosis, an arterial macroaneurysm, a cavernous hemangioma, familial exudative vitreoretinopathy (FEVR), or a vasoproliferative tumor [4]. If it is obscured by a vitreous hemorrhage or retinal detachment, the dilation of the feeding vessels can be mistaken for a racemose hemangioma or for anastomoses between tumor and retinal vessels, as in retinoblastoma [4].

If it is located in the juxtapapillary area, it must be differentiated from papilledema, papillitis, juxtapapillary choroiditis, juxtapapillary subretinal neovascularization, or a choroidal hemangioma [5].

Ocular complications include exudative retinal detachment, secondary angiomatosis, intraretinal exudation, epiretinal membrane, vitreous hemorrhage, retinal tear, rhegmatogenous retinal detachment, and neovascularization of the retinal disc or peripheral retina [10,22].

The lesions typically increase in size progressively and rarely regress spontaneously, making early treatment advisable. They usually begin as a small red or gray intraretinal spot a few microns in diameter, resembling a microaneurysm or a punctate intraretinal hemorrhage. Wide-field fluorescein angiography (FA) allows for the identification of these lesions, which exhibit early and late hyperfluorescence [6]. As the size increases, there is evidence of increasing nodularity, irrigation and drainage blood vessels that dilate and become tortuous, exudation with retinal edema, subretinal fluid and/or lipid, with early hyperfluorescence and late leakage in AF [7,8].

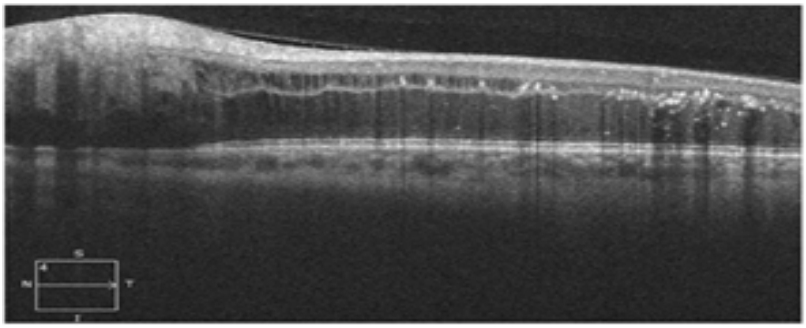

Figure 8: Incipient RCH is Observed as an Intraretinal Hyperreflective Mass on OCT.

Laser photocoagulation is the method of choice for small lesions, from less than 3 mm up to a maximum size of 4.5 mm. Extrapapillary HCR up to 1.5 mm can be reliably removed with laser photocoagulation in one or more treatment sessions. Several types of lasers, including argon green, diode, yellow, and krypton, have proven effective [9]. Vascular lesions may absorb yellow light more readily than other laser wavelengths, based on the absorption spectrum of oxyhemoglobin [21]. Furthermore, the use of a blue-green laser immediately after intravenous infusion of sodium fluorescein may allow for better uptake of laser energy by these vascular lesions [10].

The prognosis is often uncertain in larger tumors, requiring several sessions and carrying a high risk of failure.

Lesions between 1.5 and 4.5 mm in diameter are more difficult to destroy with laser photocoagulation. Several sessions are almost always required, and some lesions persist after several treatment sessions. Large burns are performed at low intensity and for a long duration [11,12]. It is recommended to begin by delineating the lesion, then targeting the vessels, requiring several sessions, and finally targeting the tumor surface. The effects are often not permanent. Cryotherapy is reserved for larger tumors (generally greater than 4.5 mm), anterior to the equator, or when opacity of the vitreous humor prevents photocoagulation. It can be applied to lesions refractory to previous laser photocoagulation or as a first-line therapy in general for lesions with very prominent feeding and draining vessels. Exudation may often increase temporarily after the procedure, and vitreous contraction may also occur.

Photodynamic therapy with verteporfin, or transpupillary thermotherapy, with or without anti-VEGF, is primarily used for juxtapapillary hemangiomas.

Other options include brachytherapy, external beam radiation therapy (reserved for highly selected cases), and vitrectomy, indicated in cases of vitreoretinal hemangiomas and massive tractional or exudative retinal detachment, with or without endocryotherapy or with tumor excision [8].

Some use prednisone up to 1 mg/kg/day for one to three days, starting on the same day as treatment, with a gradual reduction until discontinuation four to seven days after treatment. Surgical treatment of large extrapapillary retinal hemangiomas with vitrectomy and/or cerclage may be performed when there is rhegmatogenous or tractional retinal detachment, epiretinal or vascular proliferation affecting central vision, or associated vitreous hemorrhage.

In these cases, the risk of vitreoretinal proliferation is higher due to compromise of the blood-ocular barrier. Excision of a CRH requires attention to intraocular hemostasis, which may include treatment of the lesion with endolaser photocoagulation, followed by diathermy of the feeding vessels under high intraocular pressure before excision, or ligation of the feeding vessels with intraocular suture before excision [13].

Transretinal ligation of feeding vessels is a valid option, but revascularization must be considered due to the potential opening of adjacent vessels [14].

In one case series, patients who underwent retinectomy and endovenous laser ablation for retinal hyperplasia (RH) were left with silicone oil, which was removed after 6 months; however, the long-term prognosis was poor. Eyes were enucleated due to neovascular glaucoma, reflecting the aggressive reproliferation of RH in these eyes.

However, in a series by Overdam, he reported that extrapapillary RH treated with retinectomy, endovenous laser ablation, complete vitreous excision, and epiretinal membrane removal had a good prognosis, with only one case of RH recurrence [18-19].

Through vitrectomy, endodiathermy of feeding and draining vessels, endoresectal resection of the optic nerve head (ORH), and silicone oil injection, Karacorlu et al. (20) achieved visual improvement and a 92% anatomical success rate in 13 eyes.

None of the tumors in their series were located less than 10 mm from the optic nerve or fovea.

The management of juxtapapillary ORH, however, is complicated by the adverse visual effects that can result from attempted tumor ablation, particularly for tumors located within or near the papillomacular bundle. Decreased visual acuity, altitudinal loss of the visual field, and the development of central scotomas have been associated with laser photocoagulation treatment.

For some juxtapapillary retinal hyperplasias (RCHs) that do not cause significant exudation or that spare the macula, observation is recommended. Even in cases where macular exudation affects central vision, observation may be the best option, often with an increase or decrease in intraretinal fluid and sometimes with very slow tumor growth. In the absence of safe ablative options, treatment is usually limited to attempting to minimize exudation that affects central vision. Photodynamic therapy, proton beam therapy, or the use of anti-VEGF or intravitreal corticosteroids can be used individually or in combination to reduce exudation in some cases, although the benefit is usually limited [15].

In these patients, retinal vascular proliferation is common in both juxtapapillary and extrapapillary RCHs, which, in these situations, can be treated surgically when their location is epiretinal [14].

HCR can induce retinal vascular proliferation on its surface, but the tumor component resides in the retina and cannot be removed without retinectomy (16-17). Hypoxia-induced factor (HIF) inhibitors have recently emerged as a potential treatment option for HCR. These agents suppress the expression of angiogenic factors such as VEGF and platelet-derived growth factor (PDGF) by acting on the HIF pathway. A case series of 7 patients with VHL (12 eyes) treated with belzutifan (an HIF inhibitor) revealed that all eyes with active HCR achieved ocular tumor control during a mean follow-up of 13 months [21]. Currently, it is only approved for the treatment of renal cell carcinoma in VHL.

Tyrosine kinase inhibitors (e.g., sunitinib, pazopanib) target VEGF receptors and other pro-angiogenic pathways, inhibiting tumor growth and reducing exudation. In a pilot study, 3 patients with VHL-related juxtapapillary CRH were treated with sunitinib, resulting in a reduction of macular edema and stabilization of vision in all cases, despite no reduction in tumor size being observed [22].

Furthermore, gene therapies aimed at restoring functional VHL protein or correcting mutations mediated by CRISPR/Cas9 are under investigation.

Finally, the effect of oral propranolol in the treatment of infantile hemangiomas is well established, and there are currently case reports and clinical trials demonstrating its effectiveness in reducing tumor size and exudation in the treatment of both retinal capillary hemangiomas and choroidal hemangiomas.

A pilot clinical trial with 120 mg/day of propranolol to delay/stop the growth of retinal hemangioblastomas in patients with VHL, with a 3-year follow-up, concluded that there was both tumor disappearance with a decrease in exudation, as well as a decrease in plasma levels of VEGF and HIF. It was not studied at doses of 1 to 3 mg/kg/day, which is the treatment dose for infantile hemangiomas [23].

In a clinical case where a patient received laser photocoagulation as treatment for a choroidal hemangioma, visual acuity (VA) improved to 20/100 three months later, but foveal serous detachment persisted. Treatment with daily oral propranolol (120 mg) was initiated. One month later, VA improved to 20/20 and the foveal serous detachment resolved. After eight months of follow-up, both VA and macular thickness remained stable [25].

The commonly used pharmaceutical presentation is propranolol 40 mg tablets, one every eight hours, up to a total dose of 120 mg/day, reached in seven to ten days [26]. In a case report of a 55-year-old patient with choroidal hemangioma associated with serous retinal detachment, treatment was initiated with oral propranolol (80 mg twice daily), after which the dose was reduced to 40 mg twice daily due to adverse effects described by the patient, such as dizziness and weakness. Six weeks after starting propranolol treatment, the retina was adhered, with complete reabsorption of the subretinal fluid [27].

It is important that an experienced cardiologist manage propranolol, establishing the dosage regimen and evaluating any associated adverse effects, the most frequent of which are hypotension, bradycardia, and hypoglycemia [28,29].

Conclusion

We can conclude that our patient's case presents a treatment challenge due to its juxtapapillary retinopathy of prematurity (ROP), which compromises visual acuity due to the exudative phenomena present. Therefore, in conjunction with the internal medicine and cardiology departments, we decided to initiate oral treatment with propranolol, increasing the dose to 120 mg/day, and to monitor her progress to assess the need for anti-VEGF and/or corticosteroid therapy for her foveal serous detachment.

Furthermore, given her age, which is at risk for the development of other systemic tumors and/or cysts, and her history of tachycardia, we initiated screening, in conjunction with the internal medicine department, for a possible diagnosis of Von Hippel-Lindau syndrome. This screening included MRI of the brain, brainstem, and spinal cord, urine catecholamine testing, and abdominal ultrasound and CT scans, with results pending.

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