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# Double peeling and endolaser ablation for retinal detachment in von Hippel-Lindau disease

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ABSTRACT

*Purpose:* To describe a successful surgical approach to macula-off retinal detachment in von Hippel-Lindau disease.

*Observations:* A 28-year-old male with a history of von Hippel-Lindau disease presented to us with significant worsening of vision in his single functional eye after undergoing a cryotherapy and laser session for multiple retinal capillary hemangioblastomas. Given a tractional and exudative retinal detachment involving macula, we performed a vitrectomy, epiretinal membrane peeling, internal limiting membrane peeling, endophotocoagulation of all hemangioblastomas, and fluid-air exchange. Over 30 days, there was total resolution of retinal detachment and improvement of his vision. At 13-month follow-up, the patient exhibited anatomical and functional stability.

*Conclusions and Importance:* Double peeling and endolaser ablation may be an alternative treatment approach for patients with tractional and exudative retinal detachment in von-Hippel-Lindau disease.

## 1. Introduction

Von Hippel-Lindau disease (VHLD) is a rare hereditary syndrome caused by germline mutations in the *VHL* gene, characterized by an increased risk of Retinal Hemangioblastomas (RHs) among other several benign and malignant tumors.[1–3]

Regarding ocular involvement, RHs are observed up to 60% of VHLD patients, and the mean age at detection is 25 years.[2] These lesions can be identified by routine screening examinations or after development of visual impairment from macular edema, hemorrhages, or exudative retinal detachment.[1,3,4] RHs can be located on the optic disc (juxtapapillary/epipapillary) or in other parts of the retina (extrapapillary).[1,3,4] Extrapapillary RHs are more common (85% of cases) and are most often located in the temporal region, ranging from small, red-grayish dots to reddish-white tumors with nourishing vessels and dilated and tortuous drains.[1,3,4] Growth of RHs occurs over months to years, leading to exudative events like macular edema or exudative retinal detachment.[1,3,4] Recurrent episodes of RHs activity can lead to growth of epiretinal membranes (ERM) and tractional retinal detachment.[1,3,4]

The most common ocular treatment for RHs includes laser photocoagulation, cryotherapy, or radiotherapy.[1,3,4] Other therapeutic options include transpupillary thermotherapy, photodynamic therapy, and vitrectomy endorecection. Intravitreal injections of anti-vascular endothelial growth factor agents and corticosteroids have also been used as adjuvant treatments, with the aim of reducing vascular permeability and inflammation but with little effect on tumor involution.[1,3,4] The therapeutic approach generally depends on the size and location of the RH, and the presence of complications.

Here, we report successful treatment of complicated RHs using endolaser ablation, ERM and internal limiting membrane (ILM) peeling for tractional and exudative macula off retinal detachment (T&ERD) in a patient with VHLD.

## 2. Case report

In May 2019, a 28-year-old man with a previous diagnosis of VHLD was referred for a retinal evaluation after significant worsening of vision in his left eye (OS). Over the last two years, OS RHs had been treated with laser photocoagulation sessions, showing refractory macular exudates and a best correct visual acuity (BCVA) of 20/30. Seven days before his presentation to us, one last cryotherapy and laser photocoagulation session was done in OS, followed by an acute and dramatic worsening of BCVA to 20/400. His right eye (OD), had previously been

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**List of abbreviations**

| | |
|---|---|
| BCVA | Best Correct Visual Acuity |
| ERM | Epiretinal Membranes |
| HCPA | Hospital de Clínicas de Porto Alegre |
| ILM | Internal Limiting Membrane |
| MD | Medical Doctor |
| OCT | Optical Coherence Tomography |
| OD | Right eye (ocular dexter) |
| OS | Left eye (ocular sinister) |
| PhD | Post-Doctoral Degree |
| RD | Retinal Detachment |
| RH(s) | Retinal Capillary Hemangioblastoma(s) |
| T&ERD | Tractional and Exudative Retinal Detachment |
| VHL | von Hippel-Lindau |
| VHLD | von Hippel-Lindau Disease |

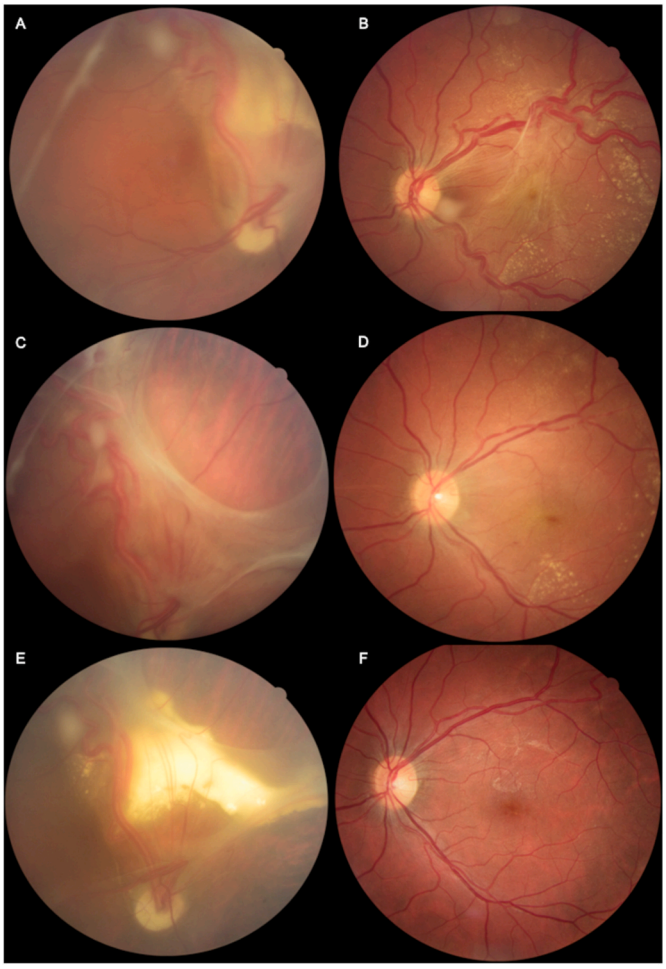


**Fig. 1.** Fundus photography during patient follow-up. Baseline evaluation: a) right eye (OD) with media opacity and juxtapapillary anatomical disorganization and b) left eye (OS) with glial tissue, macular exudates, vascular dilation, and tortuosity of the temporal vessels. One-month evaluation after vitrectomy with membrane peeling and endolaser in OS: c) OD was stable and d) OS with vascular tortuosity and exudate reduction. Five-month postoperative evaluation: e) OD with subretinal exudative material concentrated in the region above the disc and f) OS with significant improvement in macular anatomy.

diagnosed with multiple RHs and a retrobulbar lesion involving the optic nerve (Fig. 1) and superior orbit. Besides poor visual prognosis in OD (hand motions), he had undergone to external radiation therapy in order to control the orbital and intraocular tumors.

OS fundus examination revealed mild vitreous opacities and a retinal detachment involving the posterior pole and the superotemporal region associated with hard exudates and macular epiretinal membranes. Numerous RHs associated with tortuosity and vascular dilatation were observed in the temporal region, both posterior and anterior to the equator. The optic disc was normal (Figs. 1 and 2). Optical coherence tomography (OCT) images of OS showed an epiretinal band diffusely adherent to the macular region and a neurosensory retinal detachment (Fig. 3).

Given the tractional and exudative retinal detachment (T&ERD) with visual loss in his single functional eye, the patient agree to be submitted to an OS vitrectomy, ERM peeling, ILM peeling, endophotocoagulation of all RHs, and fluid-air exchange (Fig. 2). During the first week of follow-up, there was a significant improvement in OS macular anatomy and vision (BCVA = 20/50; Figs. 2 and 3). Over 30 days, there was total resolution of retinal detachment and improvement of his vision to BCVA = 20/20. In the last evaluation at 13-month follow-up, the patient exhibited anatomical and functional stability (BCVA = 20/20) in OS and all RHs were stable or decreased in size (Figs. 2 and 3).

## 3. Discussion

We describe a successful surgical approach to T&ERD in a patient with VHLD. In these patients, the treatment of choice is generally laser photocoagulation ablation, especially for extrapapillary RHs up to 1.5

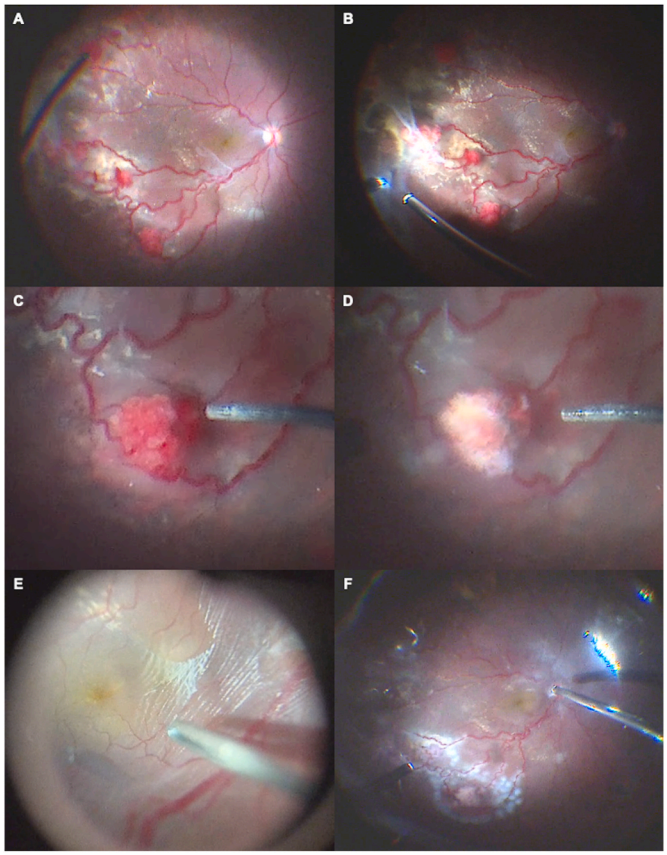


**Fig. 2.** Surgical steps. A: initial aspect during vitrectomy; B: releasing fibrous tractions; C: retinal capillary hemangioblastoma (RH) before endophotocoagulation; D: RH aspect after endophotocoagulation; E: internal limiting membrane peeling; F: final aspect after fluid-air exchange.

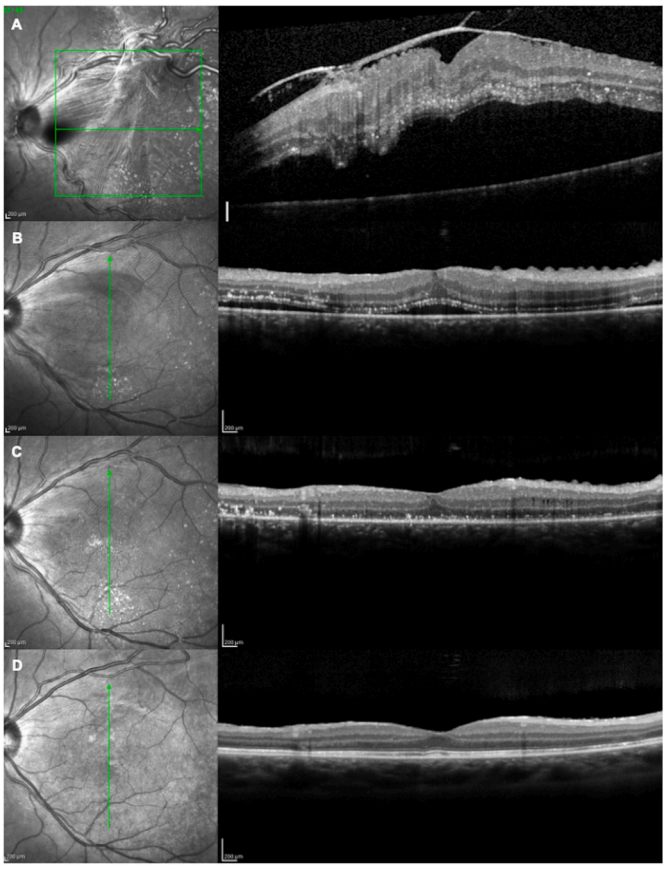


**Fig. 3.** Optical coherence tomography (OCT) findings. A: baseline image showing exudative and tractional retinal detachment (E&TRD), retinal thickening, and epiretinal membrane (ERM) formation; B: One week after surgery with great improvement but still some subretinal fluid and hard exudates; C: One month after surgery with absorption of subretinal fluid and faint, hard exudates; D: Stability of macular anatomy at 13-month follow-up.

mm in diameter located posterior to the equator.[1,3,4] RHs between 1.5 mm and 4.5 mm in diameter may require numerous treatment sessions over months and years to achieve involution, often associated with hemorrhagic and exudative events.[1,3,4] Cryotherapy is usually reserved for treatment of RHs located anterior to the equator, associated with exudative retinal detachment or not responsive to laser photocoagulation.[1,3,4] RHs larger than 4.5 mm are challenging to manage because they do not respond well to ablative treatments, often requiring a combination of therapeutic strategies, including surgical excision and radiotherapy.[1,3,4] Although vitreoretinal surgery has been described by some authors for excision or ablation of RHs in patients with VHLD, this procedure is associated with high rates of complications, including hemorrhages, recurrent retinal detachments, and neovascular glaucoma.[1–3,5] In the Gaudric and Krzystolik series, many individuals experienced persistent, significant visual loss and few achieved a visual acuity better than 20/40.[5–7] These results may have been related to recurrent loss of the blood-retinal barrier leading to scarring at the vitreoretinal interface. Many of these patients had a posterior hyaloid strongly adherent to the retina, as well as ERM and vitreoretinal proliferations, making the procedure more challenging.[5–7] Retinotomies and retinectomies were also frequently needed for adequate anatomical success, elevating inflammatory markers and cell proliferation, with more interventions required.[5–8]

In our case, a previous cryotherapy procedure probably triggered the T&ERD some days later, which is a known complication of this treatment for vasoproliferative retinal lesions. Given this patient's single functional eye, we opted for the most conservative surgical approach possible, which was corroborated by the successful detachment of the posterior hyaloid/ERM peeling and the release of traction. We did not perform excision of RHs because there was satisfactory regression of tumors after endophotocoagulation. After peeling posterior hyaloid/ERM assisted by triamcinolone, we re-stained with brilliant blue G and peeled ILM to achieve a more definitive relief of the traction and to reduce the chance of recurrent ERM formation.[9,10] Since there was total re-attachment of the retina and no bleeding points after fluid-air exchange, we left the vitreous cavity with air. The patient progressed very well during the postoperative period, quickly returning to his daily activities and remained stable over more than one year of follow-up.

## 4. Conclusion

The management of VHLD can be challenging due to its rarity and the great individual variability in its phenotypic presentation. We believe that sharing clinical experiences between specialists in the field may help to achieve increasingly positive results for control of the vitreoretinal complications related to VHLD. Although this report has described only one case, our technique involving double (MER/ILM) peeling and endolaser ablation may be an alternative treatment approach for patients with similar presentations.

## Patient consent

The patient provided informed consent for publication of this report.

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None.

## Authorship

Felipe Mallmann: surgery, retinal images, writing-original draft preparation, reviewing and editing. Marcelo Maestri: Writing-Reviewing and Editing.

## Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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