• Letter to the Editor •

Vitreous surgery in treating complex retinal capillary hemangioma

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Dear Editor,

Retinal capillary hemangioma (RCH) is a benign vascular tumor that poses significant challenges due to its potential to cause vision impairment and retinal detachment (RD). We present a comprehensive case report of a large-diameter RCH treated with vitreous surgery, underscoring the importance of selecting appropriate treatment strategies based on the individual characteristics of RCH patients to minimize their impact on vision and future well-being.

RCH is most often found in the periphery of the retina (85%) and less frequently in the optic disc area (15%)[1]. This condition can manifest as either a sporadic unifocal lesion or as an initial and common symptom of von Hippel-Lindau (VHL)[2], which results from mutations in the VHL tumor suppressor gene, promoting tumor growth in several organs, including the retina, central nervous system, and kidneys[3].

RCH fundus observations present with a nearly round, orange-red mass, typically located in the superior or inferior temporal periphery of the retina, with dilated feeding artery and tortuous draining vein extending from the optic disc[4]. RCH may be asymptomatic in the early stages, however as capillary function gradually deteriorates, typical symptoms will become evident, including persistent exudation, continuous leakage within and beneath the retina, leading to macular exudation and exudative RD. Proliferation of glial cells can also lead to tractional-rhegmatogenous RD. Treatment strategies for RCH are based on the size, location, and extent of complications of the tumor[5]. Pars plana vitrectomy (PPV) is commonly reported as a clinically indicated treatment, achieving long-term improved anatomical results and minimal vision-threatening side effects[6].

A 31-year-old woman sought consultation due to a progressive decrease in visual acuity in her right eye (RE) accompanied by floating dark shadows for three days. She had no medical disorders, was not using any ongoing medications, and there was no family history of neurological or urinary issues. Examination revealed her corrected visual acuity to be 14/20 in the RE and 20/20 in the left eye with normal intraocular pressure and anterior segment findings in both eyes. Dilated fundus examination in the RE using a 90 D lens disclosed a large orange-red mass in the lower peripheral region, measuring about 7.5 mm in basal diameter. Extensive yellow-white subretinal exudation and subretinal exudative RD were noted surrounding the mass (Figure 1A). The left eye showed no signs of similar peripheral vascular anomalies.

Both B-scan (Figure 1B) and color Doppler ultrasounds (Figure 1C) of the RE revealed an elevated hyperechogenic area on the inferior wall, which remained in a fixed position. Fundus fluorescein angiography highlighted a prominent hyperfluorescent mass, with intensity enhancement in the early staining phases of feeder arteries and draining veins, followed by progressive leakage in the later stages (Figure 1D-1F).

The systemic examination was unremarkable. Comprehensive genetic testing for the patient and her parents did not identify any pathogenic variants linked to VHL. Based on these findings, the case is classified as an isolated RCH, with complication of exudative RD in RE.

Considering the large tumor exhibiting active exudation and RD, our hospital performed a primary vitreoretinal surgery on the patient's RE. Surgery utilized a standard three-port, 23-gauge PPV setup, enhanced with a microscope equipped with an illumination system for precise tumor endoresection.

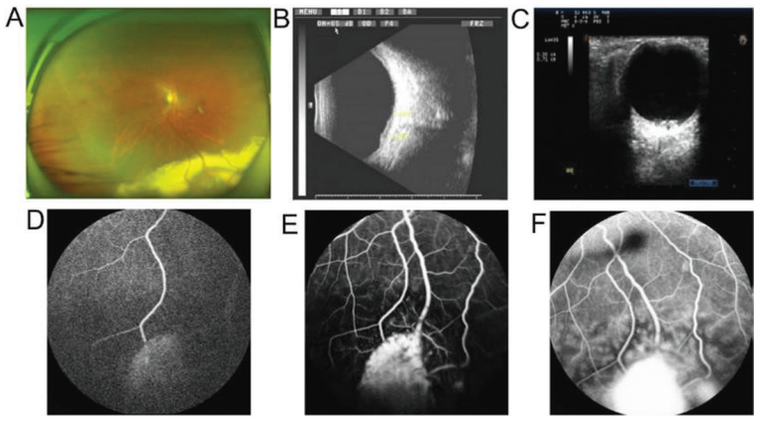

Figure 1 Ocular examination of the patient with retinal capillary hemangioma A: Wide-field fundus photography of RE displays a tumor in the peripheral area with prominent feeding vessels, surrounded by exudative retinal detachment, not involving the macula; B: B-scan ultrasound reveals a moderately echogenic lesion in the lower wall of the eyeball, measuring 8.08 mm in basal diameter and 2.02 mm in thickness. The lesion has clear boundaries and no choroidal excavation. C: Color Doppler ultrasound shows thickening and elevation in the lower scleral wall, with a maximum elevation of 0.20 cm and a base diameter of 0.71 cm, without evident blood flow signals. D–F: Fundus fluorescein angiography demonstrates the following in RE: arterial phase (D, prominent feeding arteries connected to a high-fluorescence mass rapidly filling), venous phase (E, tortuous draining veins beginning to fill), late phase (F, significant leakage from the vascular tumor). RE: Right eye.

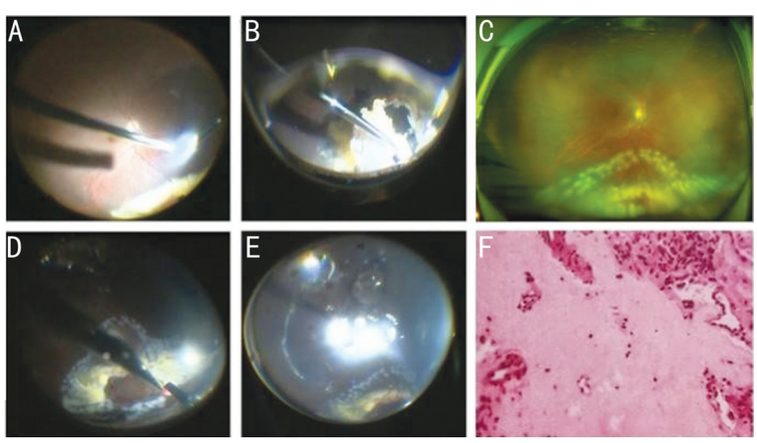

Figure 2 Retinal capillary hemangioma patient undergoing pars plana vitrectomy surgery A: After vitreous removal, the fundus examination reveals an orange-red vascular tumor in the lower peripheral retina, surrounded by yellowish-white exudates; B: Electrocoagulation of feeding artery and draining vein, hemangioma excision, and specimen retrieval for pathology; C: Repositioning of the retina and application of laser photocoagulation; D: Examination shows retinal attachment, and intraocular silicone oil tamponade is applied; E: On the fourth postoperative day, the tumor is excised, and peripheral laser closure is evident; F: Pathology result—gray-white tissue from the right eye retina shows angiomatous proliferation with stromal loosening degeneration.

After the vitreous was fully detached, the critical step involved targeted electrocoagulation of the tumor's feeding artery, draining vein, and around hemangioma, aiming to mitigate further blood supply to the affected area. We excised the hemangioma with the vitrectomy probe, then removed the 23 G cannula from the superior temporal region. Using a paracentesis knife, our team performed an inverted L-shaped sclerotomy approximately 6 mm in length and 2 mm in width at the port site to facilitate the removal of the hemangioma. Next, laser photocoagulation was applied to the margins of the retinal excision. The final step of the surgery involved the application of silicone oil tamponade, to facilitate the repositioning of the retina and supporting its attachment (Figure 2A-2D).

Four days following the surgery, corrected visual acuity in the RE was 20/50, and intraocular pressure was normal. The presence of silicone oil filling the vitreous cavity of the RE was a positive indicator of the successful excision of the tumor, complemented by the application of fresh peripheral laser treatment (Figure 2E). The pathological examination of the resected tumor specimen postoperatively identified a proliferation similar to retinal vascular tissue (Figure 2F). Three months after the operation, the patient underwent a procedure to remove the silicone oil. Furthermore, at the three-year follow-up, there was no evidence of any new tumor growth.

In our case, the RCH young patient has a relatively large diameter mass, which is a rarity in both clinical practice and

reported literature. To determine the most suitable treatment strategy for this unique case, we conducted a thorough review of both domestic and international research. Laser photocoagulation shows particular promise in treating peripheral tumors that are 1.5 mm in diameter or smaller. However, its effectiveness diminishes with increasing tumor size[7]. Cryotherapy is favored for managing mid-sized tumors (those larger than 1.5 mm). Nevertheless, its effectiveness is limited for RCH patients with significant complications, including vitreous hemorrhage and RD[8]. For tumors larger than 4.0 mm or those resistant to laser photocoagulation and cryotherapy, alternative interventions such as plaque brachytherapy, external beam radiotherapy, or proton beam therapy may be viable options. It's important to note that these treatments come with the potential risk of RD. Photodynamic therapy (PDT) with verteporfin is an effective treatment, especially for juxtapapillary lesions, which can occlude tumor vessels and eradicate tumors. Nonetheless, the prohibitive cost of PDT makes repeated sessions unfeasible. Anti-vascular endothelial growth factor injections combined with PDT treatments are successful and effective for managing RCH[9].

PPV is considered the preferred treatment for complex RCH or larger RCH cases accompanied by exudative or tractional RD[8]. After finalizing the treatment plan for the patient, we evaluated the timing of the surgery and considered the necessity of preceding photocoagulation or cryotherapy.

Recent findings advocate for the early implementation of PPV to ensure optimal local tumor control and superior anatomical restoration. First, nearly half of the patients treated with laser, cryotherapy, or a combination of both do not achieve tumor destruction and subsequently require PPV[8]. Additionally, early intervention with PPV is advantageous as it alleviates vitreous traction, eliminates the foundation for neovascular growth, purges inflammatory growth factors, and facilitates the enhanced application of anti-vascular endothelial growth factor and steroid medications[10]. Thirdly, employing 23- or 25-gauge miniature vitrectomy tools, combined with the advancements in observation technologies and surgical techniques, significantly lowers the risks associated with intraoperative bleeding, exudation, and local tumor recurrence[11-12].

Vitrectomy also carries some risks of complications such as intraoperative bleeding, recurrent RD due to proliferative vitreoretinopathy, and the potential emergence of new tumors. To minimize these risks, we have adopted the strategy of effectively obstructing the tumor's blood supply during the operation. Furthermore, we carefully and safely induce posterior vitreous detachment, gradually extending towards the mid-periphery using 23-gauge instruments. During the surgery, we thoroughly removed the vitreous at the base and surrounding areas. The postoperative systemic treatment with oral glucocorticoids was administered.

In conclusion, this case report provides an in-depth exploration of the strategic considerations for managing complex RCH. The absence of disease progression post-surgery underscores the potential of vitreous surgery as a viable option for handling such intricate cases, offering crucial insights for ophthalmologists and reinforcing the importance of tailored surgical interventions in ophthalmic oncology.

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REFERENCES

1 Wong WT, Agrón E, Coleman HR, *et al*. Clinical characterization of retinal capillary hemangioblastomas in a large population of patients with von Hippel-Lindau disease. *Ophthalmology* 2008;115(1):181-188.

2 Guo J, Du LP, Zhou PY, *et al*. Combined therapy guided by multimodal imaging of fifteen retinal capillary hemangioblastomas in a monocular Von Hippel- Lindau syndrome case report. *BMC Ophthalmol* 2022;22(1):205.

3 Tyagi S, Kumar KC, Saluja K. How 'eye' helped in von hippel-lindau syndrome. *Eur J Ophthalmol* 2023;33(1):NP5-NP9.

4 Gossage L, Eisen T, Maher ER. VHL, the story of a tumour suppressor gene. *Nat Rev Cancer* 2015;15(1):55-64.

5 Singh AD, Nouri M, Shields CL, *et al*. Treatment of retinal capillary hemangioma. *Ophthalmology* 2002;109(10):1799-1806.

6 Karacorlu M, Hocaoglu M, Sayman Muslubas I, *et al*. Therapeutic outcomes after endoresection of complex retinal capillary hemangioblastoma. *Retina* 2018;38(3):569-577.

7 Krivosic V, Kamami-Levy C, Jacob J, *et al*. Laser photocoagulation for peripheral retinal capillary hemangioblastoma in von hippel-lindau disease. *Ophthalmol Retina* 2017;1(1):59-67.

8 Hajjaj A, van Overdam KA, Gishti O, *et al*. Efficacy and safety of current treatment options for peripheral retinal haemangioblastomas: a systematic review. *Acta Ophthalmol* 2022;100(1):e38-e46.

9 Zubicoa A, Andonegui J, Compains E, *et al*. Combined treatment with intravitreal bevacizumab injection and photodynamic therapy for juxtapapillary retinal capillary haemangioma. *J Fr Ophtalmol* 2020;43(4):e139-e141.

10 Zubair T, Callaway NF, Ludwig CA, *et al*. Von hippel-lindau syndrome phenotype with prominent vitreoretinal neovascularization treated with early PPV: a case series and literature review. *Ophthalmic Surg Lasers Imaging Retina* 2020;51(2):109-115.

11 Cohen MN, Jumper JM, Duker JS. Treatment and excision of retinal capillary hemangioblastoma. *Ophthalmic Surg Lasers Imaging Retina* 2021;52(3):165-167.

12 Barca F, Vicini G, Nicolosi C, *et al*. Combined brachytherapy and vitreoretinal surgery for a large retinal capillary hemangioma with exudative retinal detachment. *Eur J Ophthalmol* 2021;31(6):NP75-NP80.