# EN BLOC RESECTION OF RETINAL VASOPROLIFERATIVE TUMOR USING 23G VITRECTOMY. A CASE REPORT

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## SUMMARY

**Purpose:** Retinal vasoproliferative tumor is one of the benign vascular tumors which in advanced stages leads to exudative retinal detachment with the formation of epiretinal and subretinal membranes. In such advanced stages, one of the therapeutic options is pars plana vitrectomy. This article presents the case of a patient on whom was performed 23-gauge pars plana phacovitrectomy with en bloc resection of the tumor followed by histological confirmation.

**Case report:** A 70-year-old patient with a one-year history of unilateral loss of vision in his left eye was admitted to our clinic for examination in February 2018. At admission, the best corrected visual acuity in the right eye was 1.0, and in the left eye was light perception. Based on the clinical picture, sonographic examination of the eye, and fluorescein angiography, the patient was diagnosed with a retinal vasoproliferative tumor. Due to the advanced stage of disease, we proceeded with surgical intervention. We performed 23-gauge phacovitrectomy with a bloc resection of the tumor. Subsequent histological examination confirmed the presence of the presumed tumor. The follow-up exam a few months later showed a completely attached retina with silicone oil tamponade, without exudative retinopathy. However, the best corrected visual acuity improved only slightly to the ability to count fingers at one meter.

**Conclusion:** Pars plana vitrectomy with en bloc resection of retinal vasoproliferative tumor is one of the therapeutic modalities in advanced stages.

**Key words:** retinal vasoproliferative tumor, pars plana vitrektomy, en bloc resection, exudative retinal detachment

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## INTRODUCTION

In 1983 Shields et al. described the condition of unusual retinal vascular tumor forming exudative retinopathy and called this tumor "presumed acquired nonfamilial retinal haemangioma". Later it was found, that this vascular mass involved not only the retina, but also retinal pigment epithelium and choroid. Therefore, the tumor was renamed vasoproliferative tumor of the ocular fundus [1].

Vasoproliferative tumors of the retina (VPTR) are benign lesions occuring in otherwise healthy patients between 40 and 60 years of age [2]. VPTR can be divide into primary (idiopathic) and secondary (if associated with other ocular disease like retinitis pigmentosa, pars planitis, sickle cell retinopathy, Coat´s disease, retinopathy of prematurity, toxoplasmosis, toxocariasis, or retinal detachment surgery). Primary VPTR accounts for 53 to 80% of the cases. The sex ratio is 1:1. Most patients seek treatment from an ophthalmologist complaining of a decrease in visual acuity, floaters, photopsias, or metamorphopsia, although some cases are discovered during routine evaluation.

Primary VPTR appears as a solitary intraretinal vacular mass in the retinal periphery and is typically located in the inferotemporal periphery (42%) or inferior (21%) portion of the fundus between the globe equator and the ora serrata [1]. Bilateral and multifocal lesions are typical in secondary VPTR. Shields et al. report multiple tumors in 6% of primary and 41% of secondary VPTR [4].

With fundoscopy examination VPTR exhibits a red to

orange color and can present with the following features: hard exudates surrounding the tumor, subretinal fluid, subretinal or intraretinal hemorrhage, vitreous hemorrhage, cystoid macular edema, epiretinal and subretinal membrane, exudative retinal detachment, hypertrophy of retinal pigment epithelium [5].

Further tests for VPTR, in addition to ophthalmoscopic examination, include ultrasound examination (USG), optical coherence tomography (OCT), and fluorescein angiography (FA).

Ultrasound examination can show solid tumors with medium to high internal reflectivity without choroidal shadowing. USG can also measure and follow VPTR response to treatment [4].

Fluorescein angiography is of limited value due to the peripheral location of tumors and the difficulty of capturing the initial phases of vasular filling. When it is successful, FA can reveal early filling of tumor in the arterial phase with increasing hyperfluorescence and leakage in the later phase [6].

Optical coherence tomography, like FA, has a limited role, but is used to document secondary retinal changes like macular edema and membrane formation [6].

## CASE REPORT

A 70-year-old patient with a one-year history of unilateral loss of vision in the left eye was admitted for examination to the Department of Ophthalmology in Faculty Hospital of Nitra in February 2018. Upon admission, the best corrected visual acuity (BCVA) in the right eye (RE) was 1.0, and in the left eye (LE) was light perception. Intralocular pressure (IOP) in RE was 20 mmHg, LE 17 mmHg. Upon examination with slit lamp, the anterior segment of both eyes was quiet, optical media were transparent without uveal irritation, only incipient corticonuclear cataract was present. Fundoscopy of the LE revealed posterior vitreous detachment, exudative retinal detachment in all quadrants including the macula. Hard exudates were present in the macula, and on the upper periphery there was a subretinal fibrous band. In the inferotemporal quadrant was a large subretinal hemorrhage on which anterior edge, between equator and ora serrata, was found a red vascularised tumor. Feeder vessels were only slightly tortuous and dilated (Fig. 1). The finding on the ocular fundus of the RE was physiological. The patient did not have any prior medical conditions and did not take any prescriptions medication.

Ultrasound examination (Eyecubed, Ellex, Australia) of the LE, B scan, revealed flat exudative retinal detachment with a tumorous mass in the inferotemporal quadrant with a base of 7.26 mm and height 2.07 mm (Fig. 2).

OCT (Spectralis, Heidelberg Engeneering, Germany) showed epiretinal membrane (ERM) with detachment of neurosensory retina (Fig. 2 B-D).

We had no success with FA (Canon CX-1, Digital hybrid retinal camera, USA) due to the peripheral location of

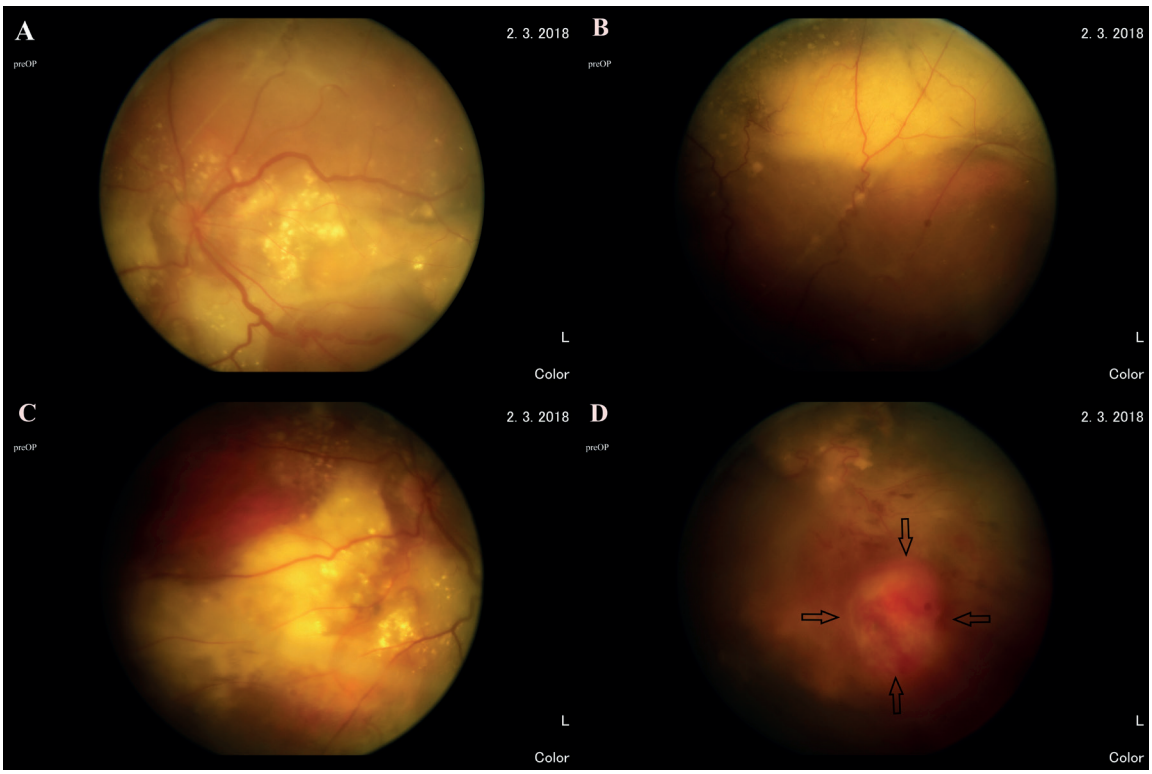


**Figure 1.** Exudative retinal detachment of left eye with hard exudates **(A, C),** subretinal fibrous band **(B)** and retinal tumor – black arrows **(D)**

the tumor and also failed to capture the arterial phases of angiography. Middle phase of FA showed a retinal aneurysm and telangiectasias with increasing hyperfluorescence. The late phases revealed leakage in the foveal avascular zone and around the VPTR site as well as staining of the tumor (Fig. 3).

Due to the advanced stage of disease and the presence of complete exudative retinal detachment and thickened epiretinal membrane, we proceeded with surgical intervention. We performed 23-gauge phacovitrectomy with membrane peeling and en bloc resection of the tumor. After insertion of the 23-gauge trocars we performed phacoemulsification without intraocular lens implantation. After making an incision in the detached thickened posterior vitreous membrane, yellow opaque fluid began to flow from the retrohyaloid space. Consequently, we proceeded to peel a markedly adhering ERM over the tumor itself, and we continued peeling from almost the entire surface of the retina. After circular barrage by endodiathermy and endolaser photocoagulation around the tumor, we gradually loosened the tumor from the base, and once it was mobile, we performed en bloc resection with a part of the adjacent retina using vitrector and

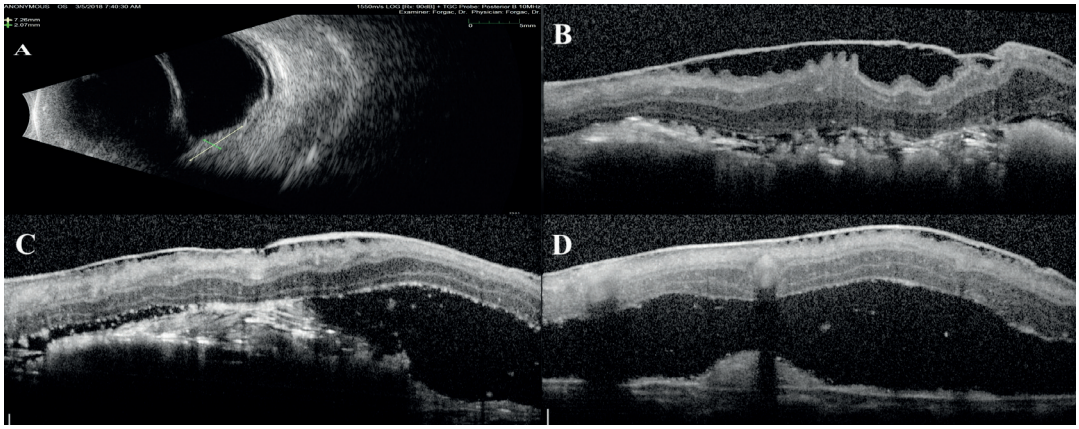


**Figure 2.** Ultrasound sonography of left bulb **(B scan).** Retinal tumor size 7.26 x 2.07 mm with exudative retinal detachment **(A),** Optical coherence tomography of left eye with epiretinal membrane and detachment of neurosensory retina **(B–D)**

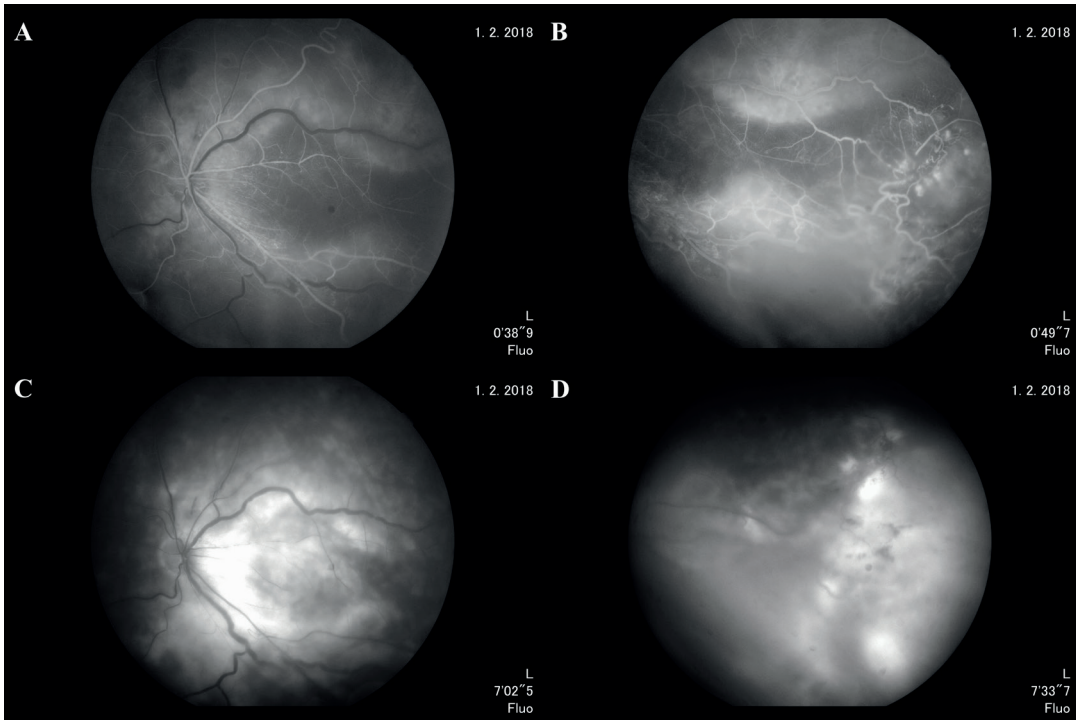


**Figure 3.** Fluorescence angiography of left eye. **(A)** arteriovenous phase with laminar vein filling, **(B)** hyperfluorescence due to telangiectasia around tumor, **(C)** leakage of fluorescein dye in foveal avascular zone, **(D)** leakage and staining of fluorescein dye around tumor

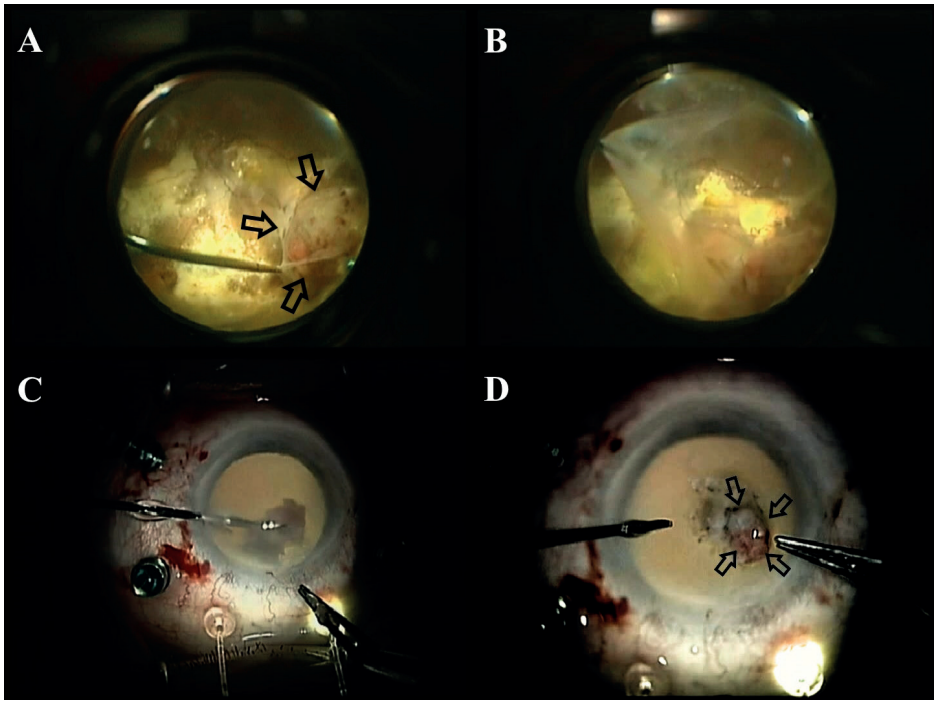


**Figure 4.** Images from surgery. **(A)** epiretinal membrane (ERM) peeling above tumor – black arrows, **(B)** ongoing ERM peeling, **(C)** extraction of tumor through the posterior capulectomy and corneal incision, **(D)** tumor with adjacent retina

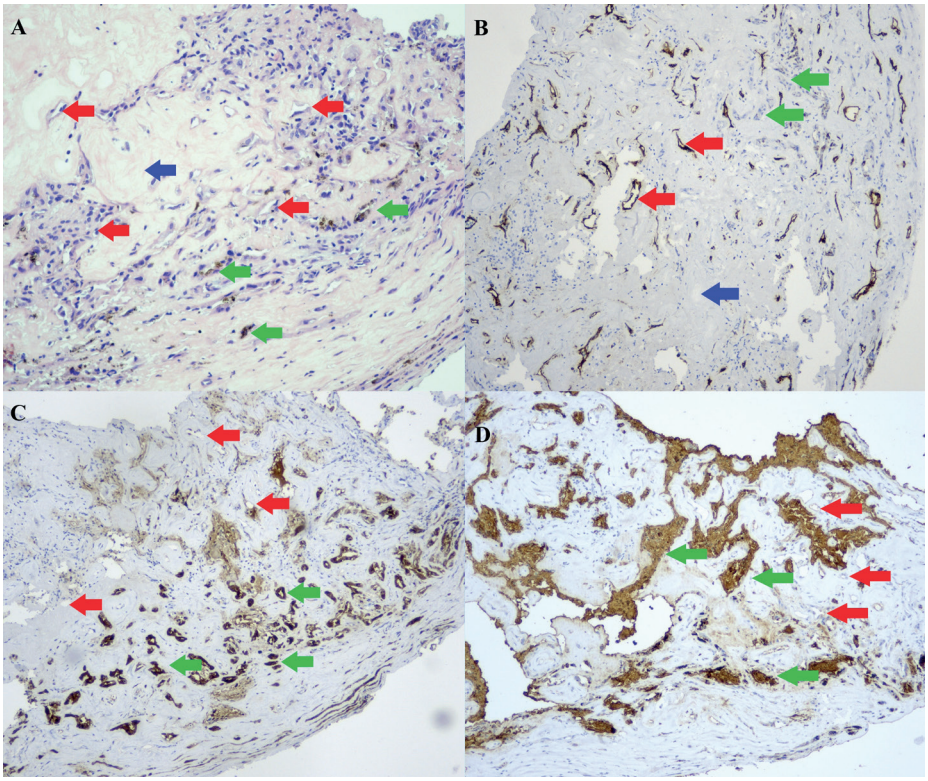


**Figure 5.** Histopathology and immunohistochemical staining. **(A)** hematoxylin-eosin, original magnification×20, **(B)** CD-34 Antibody, original magnification×10, **(C) )** S-100 Antibody, original magnification×10, **(D)** GFAP (glial fibrillary acid protein) Antibody, original magnification×10, red arrows – vasoproliferations, blue arrows – hyalinized stroma, green arrows – reactionary glial proliferation with melanin

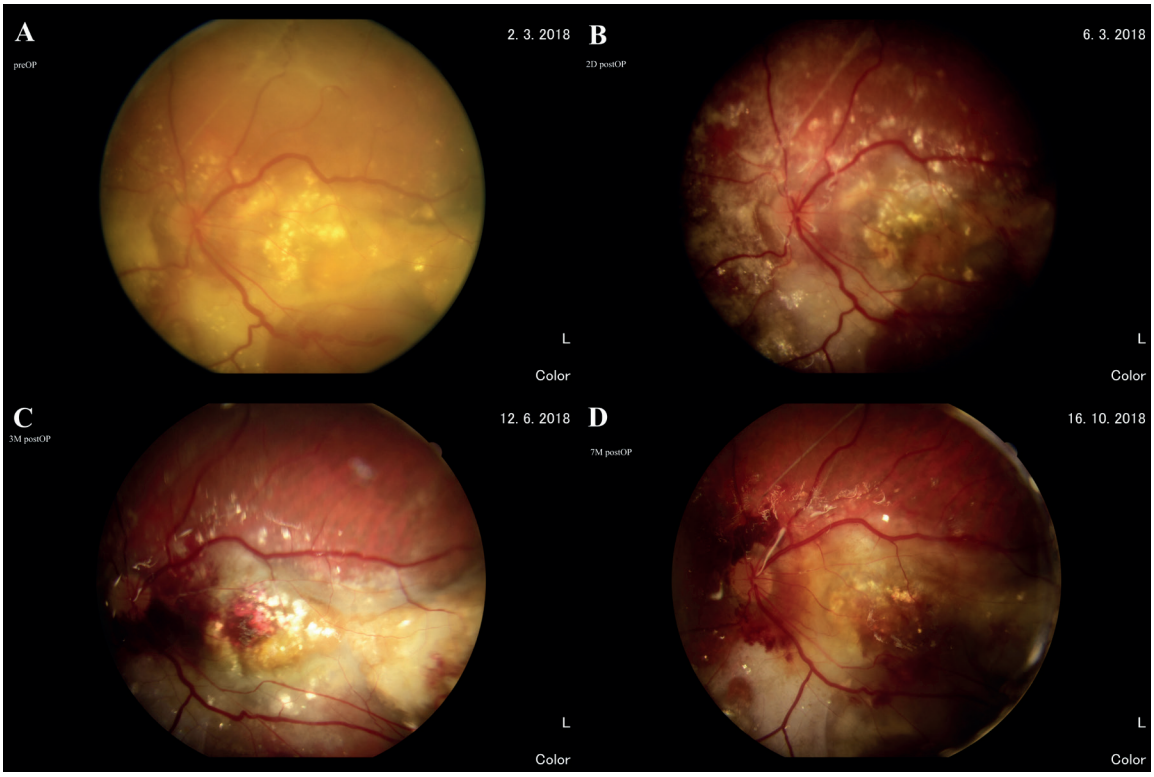


**Figure 6.** Macula. **(A)** before surgery, **(B)** 2 days after surgery, **(C)** 3 months after surgery, **(D)** 7 months after surgery

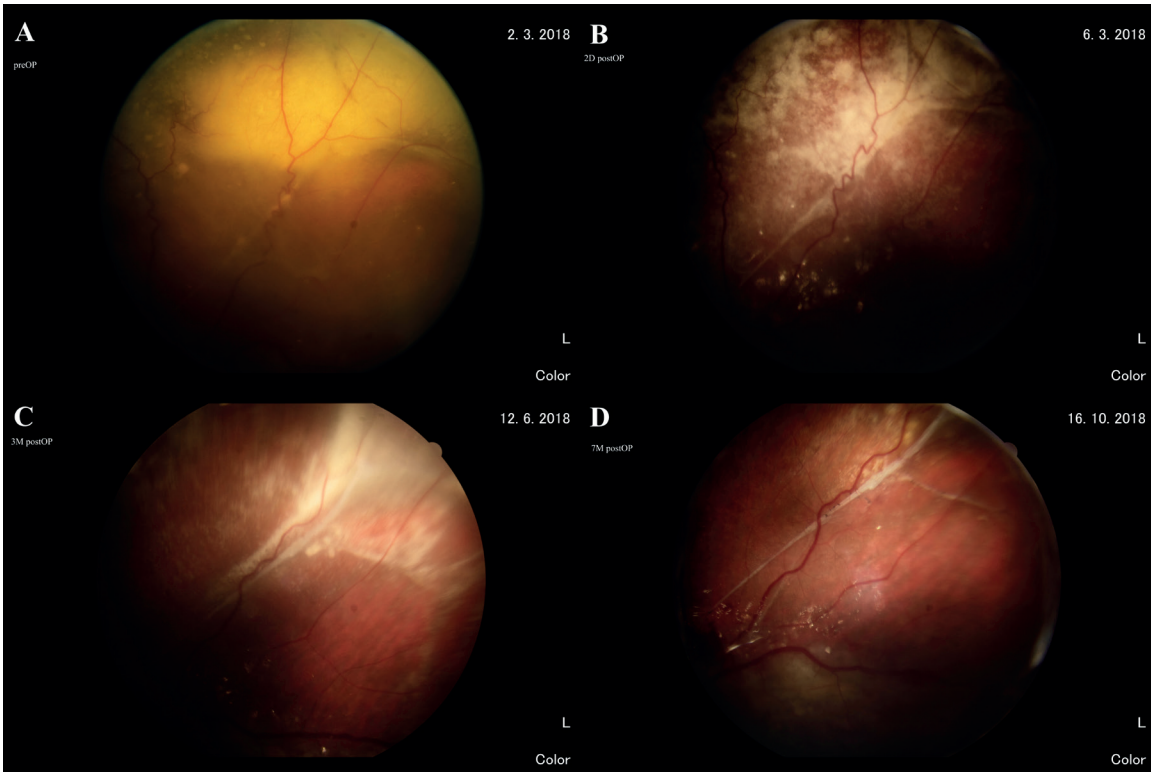


**Figure 7.** Upper retinal periphery with subretinal fibrous band. **(A)** before surgery, **(B)** 2 days after surgery, **(C)** 3 months after surgery, **(D)** 7 months after surgery

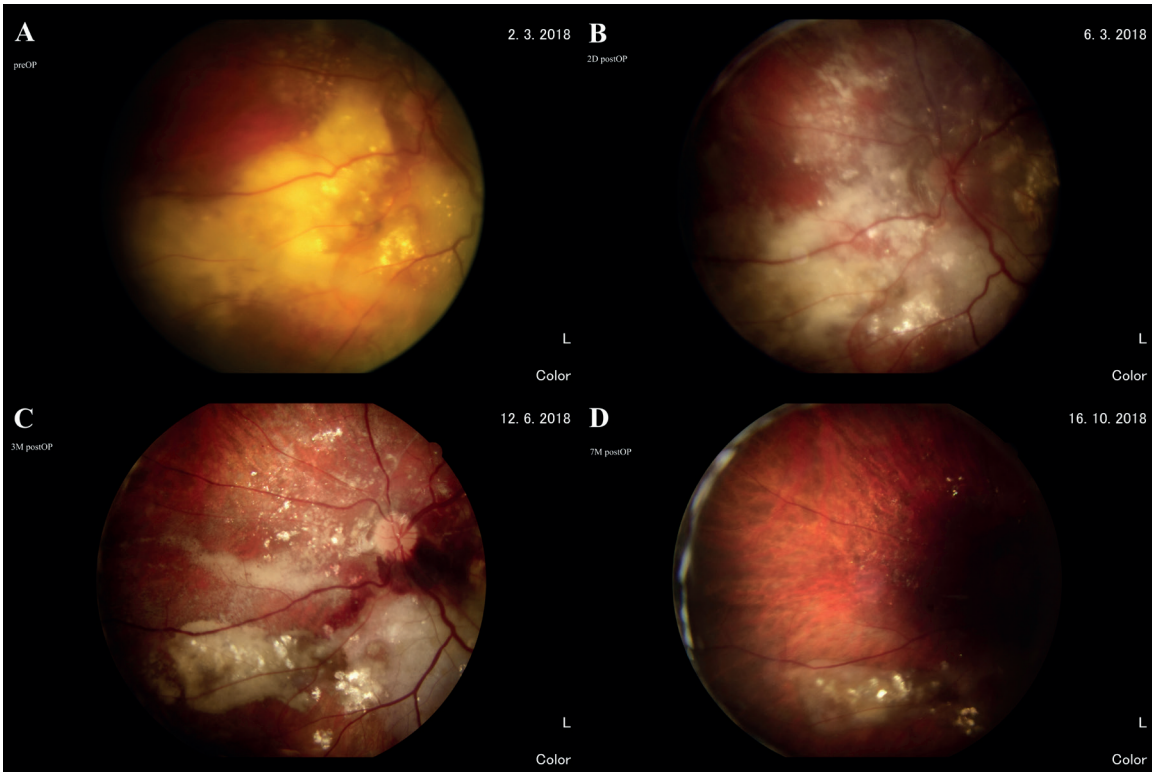


**Figure 8.** Nasal retinal periphery. **(A)** before surgery, **(B)** 2 days after surgery, **(C)** 3 months after surgery, **(D)** 7 months after surgery

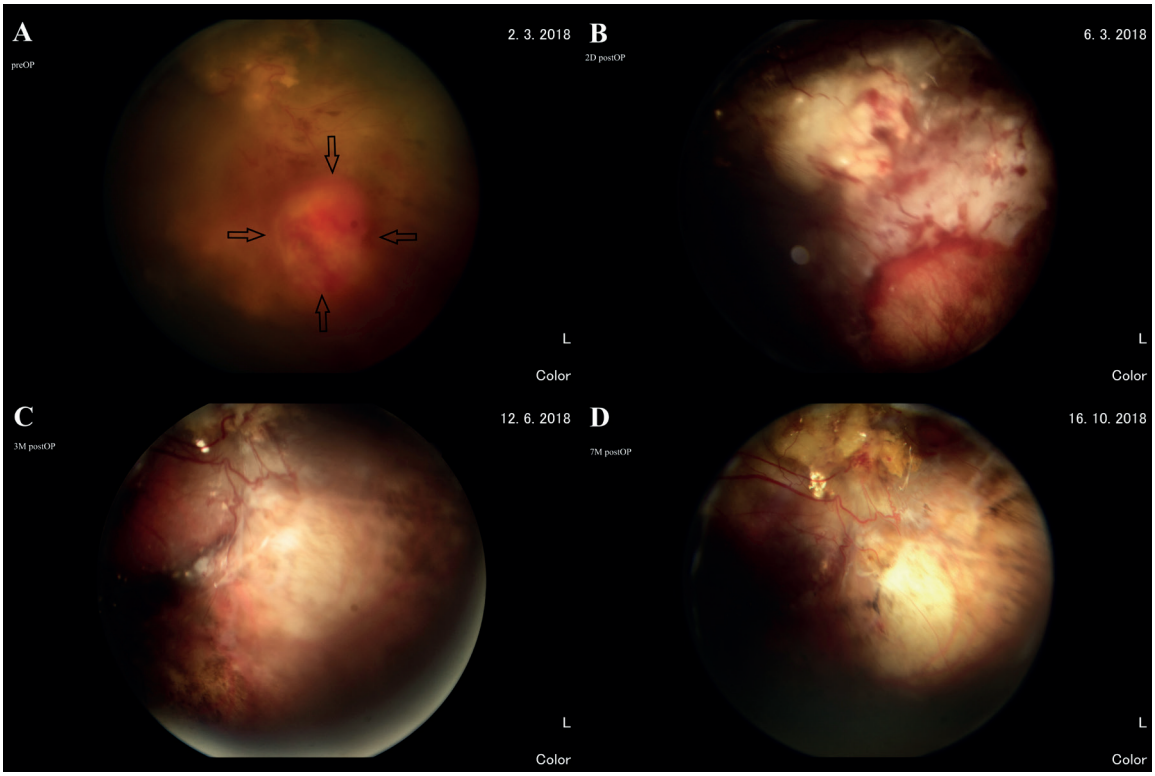


**Figure 9.** Inferior retinal periphery. **(A)** before surgery – tumor (black arrows), **(B)** 2 days after surgery (retinectomy edges treated with endolaser photocoagulation and cryoretinopexy), **(C)** 3 months after surgery**, (D)** 7 months after surgery

horizontal scissors. After creating a posterior capsulectomy, we extracted the tumor through the pupil and corneal incision. We ended the surgery with silicone oil tamponade after a previous iridectomy at the six o'clock position. We left the eye aphakic (Fig. 4).

Histological examination confirmed the presence of reactionary glial cell proliferation with melanin pigment and secondary vasoproliferation (Fig. 5). Therefore, based on histological findings, it is currently recommended to use the name "reactionary retinal glioangiosis" for VPTR [7].

Upon postoperative examination, there was a significant improvement in the local findings. The retina was completely attached with no sign of exudative detachment. The subretinal hemorrhage was gradually absorbed, hard exudates persisted in the macula, and fibrous changes occured arround the retinectomy after tumor resection. BCVA on the LE improved only slightly to the level of counting fingers at 1 meter. Gradually improved retinal findings on the LE were documented at 2 days, 3 months and 7 months after surgery compared to preoperative findings (Fig. 6–9).

## DISCUSSION

Currently there is no clear treatment protocol for VPTR. A number of different treatments have been implemented, ranging from observation to surgical intervention. Most have been based in individual cases or that of a small group of patients. Yet, the largest published study, Shields et al., used the presence of subretinal fluid, epiretinal membrane, and exudates close to the macula as criteria to treat or observe patients. Based on these criteria 51% of the cases needed treatment [1].

Cryotherapy is the most frequently used treatment. As VPTR is usually located in the periphery, cryotherapy can be performed transconjunctivaly under visual control using a binocular indirect ophthalmoscope. It is recommended to apply 2–3 freeze-thaw cycles over the entire tumor in one session. More than one cryotherapy session may be necessary to achieve complete tumor involution [8]. Adverse effects of cryotherapy include persistence of macular edema and retinal detachment due to a retinal tear adjacent to the scared area.

Laser photocoagulation has been used for small tumors. It has a limited role in larger tumors and is reserved as a supplement to other forms of treatment [4].

Photodynamic therapy is an effective treatment in retinal and choroidal vascular tumors. The successful implementation of this treatment method in VPTR was reported by Blasi et al. in tumors with an average preoperative thickness of 2.96 mm. Postoperatively, the tumors shrunk to 1.32 mm, with complete resorption of exudates in the macula without dye leakage on FA examination [10]. Its major limitation is the technical difficulties in reaching the peripheral location of the tumor.

Brachytherapy is used in larger tumors (more than 2.5 mm thick) associated with exudative retinal detachment. Aggressive cryotherapy would have to be used for such large tumors, which could lead to an intense inflammatory reaction, destruction of the surrounding tissue, vitreous hemorrhage, and increased subretinal fluid production. Therefore, Ruthenium106 or Iodine125 is used to achieve remission of such large tumors [11]. Tumor regression was achieved with brachytherapy in 97% of cases [12]. Complications associated with brachytherapy include radiation retinopathy, optic neuropathy, cataract, neovascular glaucoma, and dry eye syndrome.

Intravitreal injection of anti-vascular endothelial growth factors or corticosteroids is only used as adjunctive therapy to other destructive treatments (e.g., cryotherapy) if macular edema is the cause of decreased visual acuity [13].

Surgical resection of the tumor has been described as either local transcleral resection of medium-sized tumors with marked exudative retinal detachment [6] or as pars plana vitrectomy combined with endocryotherapy and endolaser photocoagulation [4] or transvitreal tumor resection with exudative changes in the macula which are unresponsive to cryotherapy or brachytherapy. In these cases, a standard 20-gauge pars plana vitrectomy was perfomed, with detachment of the posterior hyaloid face. After ERM peeling, diathermy was applied to all tumor vessels, and endoresection of the tumor was done using the vitrector [14]. Yeh et al. used a similar surgical technique. In these cases, 20-gauge vitrectomy with subsequent 360-degree laser around the periphery of the tumor was performed. The tumor was then removed via an enlarged superotemporal sclerostomy [15].

According to available sources, surgical resection of the tumor has so far only been documented using 20-gauge pars plana vitrectomy.

Due to the advanced stage of disease, the complete exudative retinal detachment and thickened ERM, we proceeded with surgical intervention in our patient. We decided on 23-gauge phacovitrectomy, which enabled us to perform en block resection of the tumor so we could extract the tumor through the corneal incision in the aphakic eye. In the second session, we planned to evacuate the silicone oil and proceed with possible implantation of an intraocular lens, but the patient did not show up for the next appointment.

## CONCLUSION

By performing 23-gauge par plana vitrectomy with en block resection of VPTR, we achieved in our patient improvement and stabilization of anatomical conditions on the retina without further exudative activity. BCVA on the LE improved only slightly from light perception to the level of counting fingers at 1 meter.

We assume that in the case of earlier diagnosis and intervention of the disease, we could achieve not only a good anatomical but also a functional effect in patients using less invasive procedures.

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