

# Treatment of primary vasoproliferative tumor of the retina using laser photocoagulation and intravitreal ranibizumab

Muhammad Syafiq Ahmad Musthafa<sup>1,2</sup>, Wan Norliza Wan Muda<sup>3</sup>, Norlina Ramli<sup>2,4</sup>

<sup>1</sup>Department of Ophthalmology, Hospital Sultanah Nur Zahirah, Kuala Terengganu, Terengganu, Malaysia; <sup>2</sup>Department of Ophthalmology, Faculty of Medicine, University of Malaya, Kuala Lumpur, Malaysia; <sup>3</sup>Department of Ophthalmology, Hospital Tengku Ampuan Afzan Kuantan, Pahang, Malaysia; <sup>4</sup>University Malaya Eye Research Centre, University Malaya Medical Centre, Kuala Lumpur, Malaysia

#### **Abstract**

*Background:* Vasoproliferative tumours (VPT) of the retina are benign, uncommon tumours that occur mostly in patients with no other ocular pathology. Several treatment modalities are used to treat these tumours including cryotherapy, laser photocoagulation, anti-VEGF injection, and observation.

Case presentation: We present a case of a 40-year-old female with retinal VPT. The patient received a combination of selective laser photocoagulation of the tumour and serial intravitreal injections of ranibizumab that resulted in regression of the VPT and good visual outcome.

Conclusion: Laser photocoagulation treatment should be considered for treating primary retinal VPT as it is the most available treatment modality. Combination therapy laser photocoagulation treatment with anti-VEGF provides long-term tumour regression, which is effective for the macular oedema associated with this condition.

*Keywords:* epiretinal membrane, intravitreal ranibizumab, laser photocoagulation, vasoproliferative tumour of the retina

**Correspondence:** Muhammad Syafiq Ahmad Musthafa, MD, Department of Ophthalmology, Hospital Sultanah Nur Zahirah, 20400 Kuala Terengganu, Terengganu, Malaysia. E-mail: syafiqmusthafa@yahoo.com

# Laser fotokoagulasi dan suntikan intravitreal ranibizumab sebagai rawatan primer bagi penyakit tumor retina vasoproliferatif

# **Abstrak**

Latar belakang: Tumor vasoproliferatif (VPT) pada retina merupakan tumor yang tidak merbahaya, jarang dijumpai dan sering terjadi tanpa penglibatan perubahan patologi pada tisu mata yang lain. Beberapa kaedah rawatan seperti kemoterapi, laser fotokoagulasi, suntikan intravitreal anti-faktor pertumbuhan vasoendotelial dan pemantauan klinikal didapati berkesan dalam merawat penyakit ini.

Pembentangan kes: Di sini kami melaporkan satu kes yang melibatkan seorang wanita berusia 40 tahun yang didapati mendapat VPT pada retina. Pesakit ini telah berjaya dirawat dengan laser fotokoagulasi selektif kepada tumor serta suntikan intravitreal ranibizumab yang bersiri. Hasil dari rawatan ini VPT pada retina hilang sepenuhnya dengan pengekalan penglihatan yang baik.

Kesimpulan: Rawatan laser fotokoagulasi merupakan modul rawatan yang sedia ada, perlu dipertimbangkan dalam perawatan VPT pada retina, yang mana terbukti dapat mengurangkan pertumbuhan penyakit ini. Suntikan intravitreal ranibizumab bertindak sebagi pelengkap untuk regresi tumor untuk jangka panjang terutamanya dalam mengurangkan edema di retina.

*Kata kunci:* fotokoagulasi laser, membrane epiretinal, suntikan ranibizumab, tumor vasoprolioferatif retina

# Introduction

Vasoproliferative tumours (VPT) of the retina are benign, uncommon tumours that occur mostly in patients with no other ocular pathology. VPT has a predilection for the peripheral inferior temporal retina. It may cause decreased vision due to intra- or subretinal exudates, retinal detachment, vitreous haemorrhage, preretinal fibrosis, macular oedema, and epiretinal membrane. Several treatment modalities are used to treat these tumours including cryotherapy, laser photocoagulation, anti-VEGF injection, and observation.

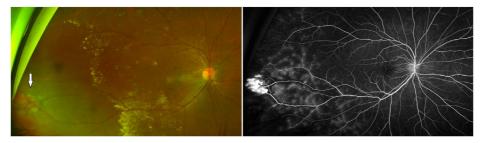


Fig. 1. (Left) White arrow showing elevated, telangiectatic aneurysmal vascular changes at temporal periphery, with hard exudates at temporal arcade. (Right) Fundus fluorescein angiography showing vascular leakage mainly at the temporal area with hyperfluorescent lesion corresponding to vascular tumour with area of significant capillary fall out area at temporal and inferotemporal quadrants.

# **Case presentation**

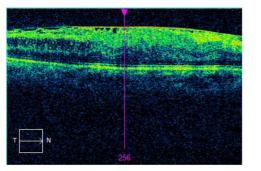
A 40-year-old Malay female with no known medical illness presented with progressive blurring of vision in the right eye for 2 months, preceded by floaters and central scotoma. The patient had no history of eye redness, pain, trauma, or eye surgery.

Upon examination, her visual acuity (VA) was 6/30 in the right eye and 6/6 in the left eye. Intraocular pressure was 16 mmHg bilaterally. Bilateral anterior segment examination was unremarkable. Fundus examination of the right eye revealed epiretinal membrane at the macula with subretinal exudates measuring 9-disc diameters near the superotemporal and inferotemporal arcades, as well as a yellowish-red tumour at the temporal periphery. The tumour was fed by a minimally dilated retinal artery and drained by a slightly dilated vein (Fig. 1, left). In the left eye, there was minimal epiretinal membrane at the macula.

Fundus fluorescein angiography revealed a hyperfluorescent lesion corresponding to the vascular lesion temporally, with late leakage along the temporal and inferotemporal arcades and an area of capillary non-perfusion peripherally (Fig. 1, right). Optical coherence tomography (OCT) of the right macula revealed a central subfield thickness of 533 microns and macular oedema with epiretinal membrane (Fig. 2).

The right eye was diagnosed with retinal VPT with epiretinal membrane and macular oedema. Treatment consisted of selective focal and barricade argon laser photocoagulation (400 shots, 150–250mW) of the peripheral retinal telangiectasia with a combination of serial intravitreal anti-VEGF (ranibizumab).

At 1-month follow-up, the patient's VA improved to 6/18, and the VPT regressed (Fig. 3, left) with a reduction in exudates and oedema. The patient received four monthly and two bimonthly intravitreal injections of ranibizumab. At the 6-month follow-up, VA improved to 6/6 and the examination revealed regressed VPT vascu-



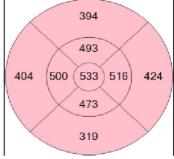


Fig. 2. Optical coherence tomography of the right eye showing epiretinal membrane and macular oedema.

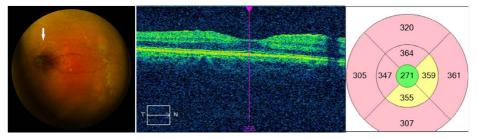


Fig. 3. (Left) Fundus photo 1 month after laser treatment showing regressed abnormal vessels surrounding laser marks (white arrow). (Right) Optical coherence tomography 6 months later showing marked reduction of macular oedema.

larization and reduced macular oedema based on OCT (Fig. 3, right). Upon 1 year of follow-up, VA remained at 6/6 and macula was dry with a central subfield thickness of 267 microns.

# **Discussion**

Retinal VPTs are rare retinal lesions that usually present in the third or fourth decade of life. Its prevalence is equal in both sexes. Around 74% exist as primary tumours, while the other 26% are secondary to pre-existing vascular, inflammatory, dystrophic ocular diseases and degenerative ocular diseases. Unlike secondary tumours, primary tumours usually present as solitary tumours.

Due to its rarity, there has been a lack of an evidence-based consensus pertaining to the optimal treatment of VPT of the retina. VPT treatment is based on the tumour's size and location as well as associated clinical features. Periodic observation of small, peripheral lesions with no evidence of macular involvement is recommended. In a report of 103 patients with VPTs, the primary treatment was observation (49%),

followed by cryotherapy (42%), laser photocoagulation (5%), or plaque radiotherapy (2%). Several case reports have been published in the literature advocating for the use of photodynamic therapy to treat primary and secondary VPT. Surgical management should also be considered for complications induced by retinal VPT, such as epiretinal membrane or tractional retinal detachment.

The presence of significant exudates and epiretinal membrane along with macular oedema in our patient indicates the need for more than observation.<sup>2</sup> The patient received laser photocoagulation treatment for the tumour and an anti-VEGF injection to alleviate macular oedema and improve vision. Krivosic *et al.* reported that direct laser photocoagulation of the retinal telangiectasia to the VPT surface induced regression of the retinal exudates. This treatment was sufficient in half the cases.<sup>8</sup> A similar finding of tumour regression was revealed in our case report following laser therapy.

Due to their natural history, retinal VPTs could lead to neovascularization, exudate formation, and tractional retinal detachment. Hence, we postulated that it would respond to anti-VEGF. Visual acuity deterioration is mostly caused by macular oedema or epiretinal membrane formation; therefore, anti-VEGF should potentially be able to decrease leakage, thus improving macular oedema, as well as inhibit neovascularization and induce regression of new vessels. While some studies indicate that intravitreal anti-VEGF may be a useful therapeutic option for these complications secondary to vasoproliferative tumours of the retina, it is still unclear whether the initially promising results will be sustained over time. 9,10

#### Conclusion

Numerous treatment modalities are available for retinal VPT depending on the lesion's manifestation and location. The treatment's objective is to preserve vision by avoiding complications such as exudative retinopathy and to induce lesion regression. In this case, we conclude that combining photocoagulation treatment with intravitreal anti-VGEF is effective at inducing long-term regression of the VPT and alleviating associated macular oedema.

# **Declarations**

#### **Consent for publication**

Informed consent was obtained from the patient for the publication of the clinical information and images used in this case report.

# **Competing interests**

None to declare.

#### **Funding**

None to declare.

#### **Acknowledgements**

None to declare.

### References

- Shields CL, Shields JA, Barrett J, et al. Vasoproliferative tumors of the ocular fundus. Classification and clinical manifestations in 103 patients. Arch Ophthalmol. 1995;113:615–623.
- 2. Henkind P, Morgan G. Peripheral retinal angioma with exudative retinopathy in adults (Coat's lesion). Br J Ophthalmol. 1966;50:2–11.
- 3. Shields JA, Shields CL, Honavar SG, et al. Clinical variations and complications of Coats' disease in 150 cases: the 2000 Sanford Gifford Memorial Lecture. Am J Ophthalmol. 2001;131:561–571.
- Saldanha MJ, Edrich C. Treatment of vasoproliferative tumors with photodynamic therapy. Ophthalmic Surg Lasers Imaging. 2008;39(2):143–145.
- Barbezetto IA, Smith RT. Vasoproliferative tumor of the retina treated with PDT. Retina. 2003;23(4):565–567.
- Heimann H, Bornfeld N, Vij O, et al. Vasoproliferative tumours of the retina. Br J Ophthalmol. 2000;84(10):1162–1169
- Irvine F, O'Donnell N, Kemp E, Lee WR. Retinal Vasoproliferative Tumors: Surgical Management and Histological Findings. Arch Ophthalmol. 2000;118(4):563–569
- 8. Krivosic V, Massin P, Desjardins L, Le Hoang P, Tadayoni R, Gaudric A. Management of idiopathic retinal vasoproliferative tumors by slit-lamp laser or endolaser photocoagulation. Am J Ophthalmol. 2014 Jul;158(1):154-161.
- 9. Saito W, Kase S, Fujiya A, Dong Z, Noda K, Ishida S. Expression of vascular endothelial growth factor and intravitreal anti-VEGF therapy with bevacizumab in vasoproliferative retinal tumors. Retina. 2013 Oct;33(9):1959-67.
- Rogers C, Damato B, Kumar I, Heimann H. Intravitreal bevacizumab in the treatment of vasoproliferative retinal tumours. Eye (London, England). 2014;28(8): 968–973.