• Letter to the Editor •

Vitrectomy combined with internal limiting membrane peeling for refractory macular telangiectasia type 1

Dan Liao[1,2], *Xiao-Li Yang*[1,2], *Rong Li*[1,2]

[1]Department of Ophthalmology, Affiliated Hospital of North Sichuan Medical College, Nanchong 637000, Sichuan Province, China
[2]Medical School of Ophthalmology and Optometry, North Sichuan Medical College, Nanchong 637000, Sichuan Province, China
Correspondence to: Rong Li. Department of Ophthalmology, Affiliated Hospital of North Sichuan Medical College, No.1, South Maoyuan Road, Nanchong 637000, Sichuan Province, China. rechelrong198222@163.com
Received: 2024-07-24 Accepted: 2025-02-19

DOI:10.18240/ijo.2025.06.25

Citation: Liao D, Yang XL, Li R. Vitrectomy combined with internal limiting membrane peeling for refractory macular telangiectasia type 1. *Int J Ophthalmol* 2025;18(6):1169-1172

Dear Editor,

Idiopathic macular telangiectasia (MacTel) type 1 is a retinal vascular disease characterized by abnormal dilation of macular capillaries, leading to metamorphopsia, progressive vision loss, and temporal scotoma enlargement. Currently, there is no standardized treatment protocol for MacTel type 1[1-2]. Treatment outcomes can vary significantly among individuals, highlighting the ongoing need for further exploration of new and more effective treatment options. This paper presents a case of refractory macular edema associated with MacTel type 1, which showed a favorable response to pars plana vitrectomy (PPV) and internal limiting membrane (ILM) peeling.

Case Presentation

Ethical Approval This study was approved by the ethics committee of the Affiliated Hospital of North Sichuan Medical College, along with the approval number 2019ER(R)107-1.

A 48-year-old Chinese female presented with reduced vision and metamorphopsia in her left eye in May 8, 2019. The patient had a 2-year history of hypertension and denied any other systemic diseases, history of radiation exposure, or significant family history. Ophthalmic examination revealed a best corrected visual acuity of 20/20 in the right eye and 20/100 in the left eye. Goldmann intraocular pressure was within normal limits, with measurements of 14.5 mm Hg in the right eye and 15.3 mm Hg in the left eye. The corneas, lenses, and vitreous bodies were clear in both eyes. The fundus of the right eye showed no abnormalities (Figure 1A). The left eye exhibited localized retinal edema in the macular area, accompanied by punctate hard exudates surrounding the edematous area. Additionally, there is a small retinal hole in the temporal peripheral region (Figure 1B). Fundus fluorescein angiography (FFA) showed microaneurysms, capillary dilatation, and leakage in the lesion area of the left eye (Figure 1C and 1D). Optical coherence tomography (OCT) showed focal thickening of the neuroepithelial layer in the macular area with interspersed punctate hyperreflectivity (Figure 1E and 1F). The patient was diagnosed with MacTel type 1 in her left eye.

Since May 8, 2019, during the initial three months of treatment, the patient received monthly intravitreal ranibizumab (Novartis, 0.5 mg/0.05 mL) for three consecutive months (Figure 2A–2C). However, the treatment was ineffective, and the macular edema did not subside; it even worsened. In August 8, and September 10, 2019 (Figure 2D and 2E), the patient received 1 mg intravitreal triamcinolone acetonide injections (Kunming Jida Pharmaceutical Co., Ltd.), but there was still no improvement in the patient's condition. By March 15, 2020 (Figure 2F), during a follow-up visit, the patient's macular edema persisted, so the patient underwent a combination of intravitreal conbercept injection (Chengdu Kanghong Biotech Co., Ltd., 0.5 mg/0.05 mL) injections and micropulse laser treatment in the macular area. In March 12, 2021 (Figure 2G), during another follow-up, the patient again received combined intravitreal conbercept injection and micropulse laser treatment in the macular area. The treatment outcomes remained unsatisfactory, causing the patient to lose confidence in the treatment, and follow-up became irregular. During a follow-up visit in January 6, 2022, it was found that the hard exudates in the macular area had increased, and the long-standing macular edema had led to cystoid degeneration in the macular region (Figure 2H). In summary, despite receiving three intravitreal anti-vascular endothelial growth factor (anti-VEGF) treatments, two intravitreal triamcinolone

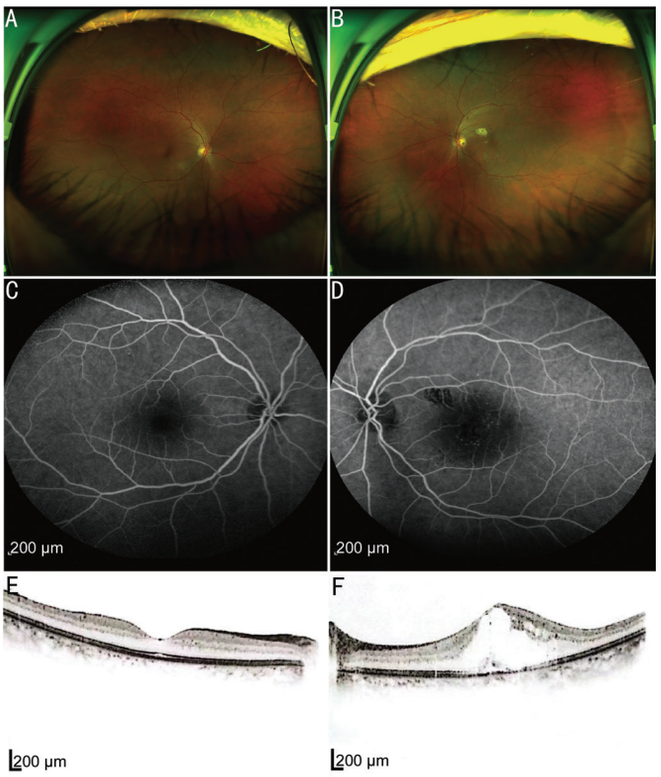

Figure 1 Images of the ocular examination at the initial diagnosis of macular telangiectasia type 1 in May 8, 2019 A: Ultrawide field laser image of the right eye; B: Ultrawide field laser image showing macular edema with surrounding punctate hard exudates in the left eye; C: FFA image of the right eye; D: FFA showing microaneurysms, capillary dilatation, and leakage in the macular of the left eye; E: OCT image of the right eye; F: OCT showing cystoid macular edema in the left eye. FFA: Fundus fluorescein angiography; OCT: Optical coherence tomography.

injections, and two combined treatments of intravitreal anti-VEGF injections with macular micropulse laser therapy over the years, the patient's condition did not improve, and her vision continued to deteriorate (Figure 2).

In January 20, 2024, her vision in the left eye decreased to counting fingers at 50 centimeters, with significant lens opacification, punctate hemorrhages in the macular area, and ring-shaped hard exudates surrounding the edematous area (Figure 3A). OCT indicated the formation of cystoid lesion in the macula (Figure 3B). She then received 25G PPV combined with ILM peeling and intravitreal air injection, along with concurrent phacoemulsification and intraocular lens implantation. During the procedure, a laser was used to seal the retinal tear in the temporal peripheral region. Two weeks postoperatively, the patient's visual acuity showed no significant improvement; however, macular edema was significantly relieved compared to preoperative levels, and cystic changes in the macular retinal tissue were observed due to prolonged macular edema lasting several years (Figure 3C and 3D). One month postoperatively, an internal lamellar

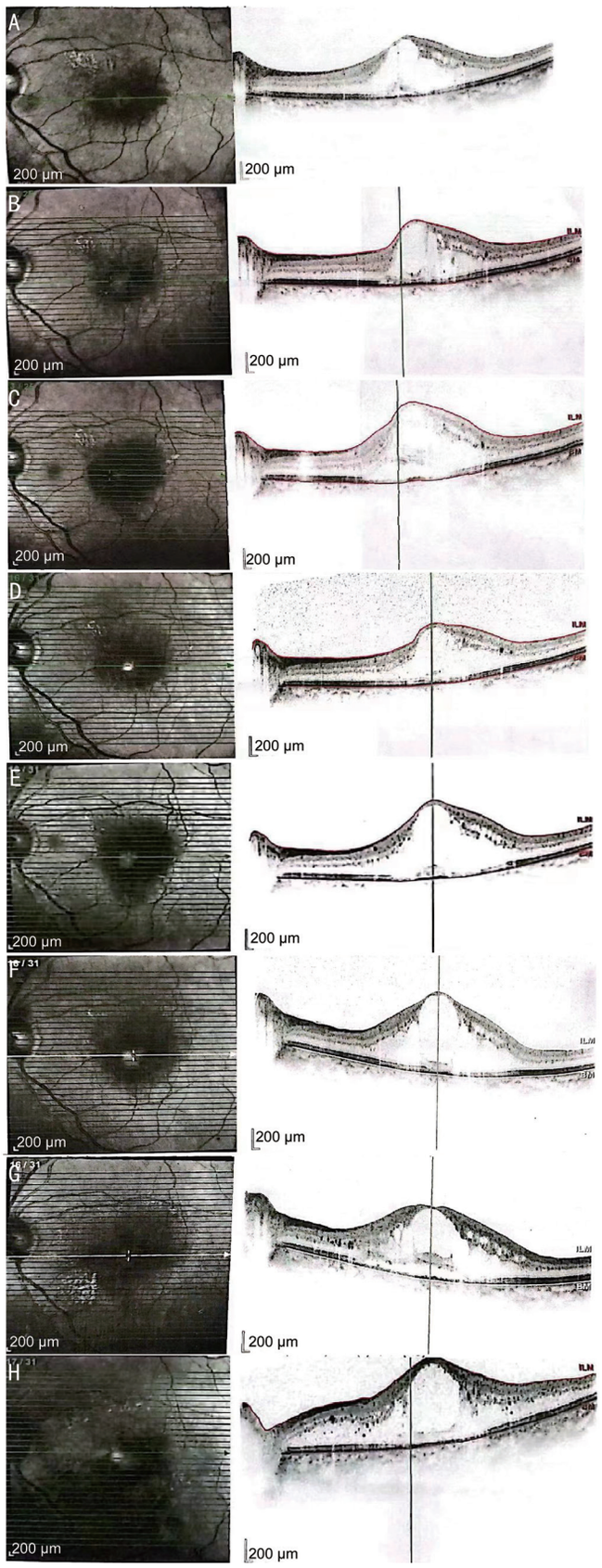

Figure 2 Follow-up OCT of the patient's left eye from May 2019 to January 2022 A: OCT image of May 8, 2019; B: OCT image of June 8, 2019; C: OCT image of July 8, 2019; D: OCT image of August 8; 2019; E: OCT image of September 10, 2019; F: OCT image of March 15, 2020; G: OCT image of March 12, 2021; H: OCT image of January 6, 2022. OCT: Optical coherence tomography.

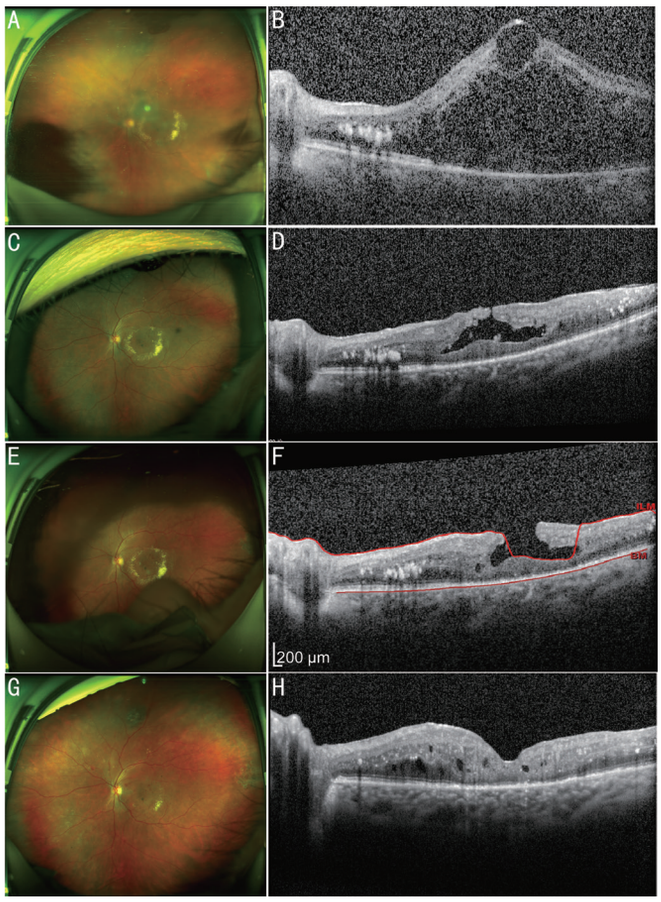

Figure 3 Preoperative and postoperative examination images of the patient's left eye A, B: Refractory macular edema and ring-shaped hard exudates in the macular area before the patient underwent PPV and ILM peeling in January 20, 2024; C, D: Ultrawide field laser image and OCT image 2wk after surgery, respectively; E, F: Ultrawide field laser image and OCT image one month after surgery, respectively; G: The fundus photograph 3mo postoperatively, with significant reduction in macular exudation; H: The OCT image 3mo postoperatively, with marked reduction in macular edema. PPV: Pars plana vitrectomy; ILM: Internal limiting membrane; OCT: Optical coherence tomography.

macular hole was detected in the affected eye (Figure 3E and 3F). At three months postoperatively, the patient's visual acuity in the left eye improved significantly to 20/400, with marked reduction in macular exudation, almost complete resolution of macular edema, and formation of the foveal contour (Figure 3G and 3H).

DISCUSSION

MacTel type 1, also known as aneurysmal telangiectasia, primarily affects middle-aged men and predominantly occurs in one eye[2]. The main symptom associated with this condition is decreased vision due to macular edema[3]. MacTel type 1 is clinically characterized by easily visible macular telangiectasis with surrounding hard exudates, which can form a ring shape and extend beyond one disc diameter. The condition is accompanied by dilated capillaries, microaneurysm formation, and occasional small hemorrhagic spots, predominantly located in the paracentral area on the temporal side of the macula[4]. These pathological changes can lead to a gradual decline in visual acuity, ultimately resulting in visual impairment. This vision loss is typically painless and predominantly affects one eye, although bilateral involvement can occur in rare cases[2].

Treating macular edema in macular telangiectasia patients is a significant clinical challenge. Effective treatments for MacTel type 1 are still under investigation due to unclear etiology and pathogenesis. Current strategies include direct retinal photocoagulation for microaneurysms, intravitreal anti-VEGF injections to reduce vascular leakage, and intravitreal triamcinolone for its anti-inflammatory properties[1,5-6]. However, some cases may be refractory to these treatments, leading to progressive vision loss. A recent study managed a MacTel type 1 case with cystoid macular edema via cystotomy and excision of the fibrinogen-rich cystoid lesion, suggesting surgery as a potential treatment for macular edema secondary to MacTel type 1[7]. Surgical interventions, including PPV and ILM peeling, have been documented to enhance visual outcomes and alleviate macular edema arising from various retinal vascular conditions, such as diabetic retinopathy and retinal vein occlusion[8-10]. Some researchers also have explored the combined use of PPV and ILM peeling for treating MacTel type 2[11].

Vitrectomy can increase oxygen levels in the inner retina[12] and improve macular vascular circulation[13]. Oxygen has been shown to inhibit vascular endothelial growth factor[14], thereby altering capillary permeability. The breakdown of the blood-retinal barrier leads to fluid leakage within the retina. Removal of the inner limiting membrane may eliminate diffusion barriers, allowing the extravasated fluid to enter the vitreous, thereby reducing the amount of extracellular fluid. Vitrectomy combined with ILM peeling has been reported to be effective in treating refractory macular edema. This procedure significantly alleviates the tangential and vertical traction forces at the foveal margins, thereby improving the pliability of the retina and reducing the severity of macular edema in diabetic macular edema patients. Furthermore, the combination of vitrectomy and ILM peeling can reduce the formation of epiretinal membranes and resolve intraretinal cystoid changes[15]. However, in patients with progressing macular cystoid degeneration, ILM peeling may remove the remaining thin layers of inner retinal tissue, leading to the development of macular holes.

This article presents a rare case of MacTel type 1 treated with PPV and ILM peeling. In this case report, PPV and ILM peeling resulted in improved macular anatomy and visual acuity, confirming the efficacy of this surgical approach for MacTel type 1. The patient experienced successful outcomes

post-surgery, including reduced subretinal hard exudation and regression of macular edema, as well as improved visual function. These results validate the effectiveness of combined vitrectomy and ILM peeling for treating MacTel type 1. For cases of MacTel type 1 with refractory macular edema, surgical intervention can be considered a viable option.

There are several limitations to the current report. First, although this study discussed how vitrectomy combined with ILM peeling can alleviate macular edema, the excised ILM tissue was too small to allow for histopathological research. Second, since this is a single case report, more clinical cases are needed to confirm the effectiveness of this treatment approach.

ACKNOWLEDGEMENTS

Authors' Contributions: Liao D: writing and revision of the manuscript, preparation of data. Yang XL: Performing surgery on the patient. Li R: revision of manuscript. All authors have read and approved the manuscript.

Foundations: Supported by Science and Technology Project of Sichuan Provincial Health Commission (No.24WXXT13); North Sichuan Medical College Doctor Start-up Fund Project (No.CBY24-QDA01).

Conflicts of Interest: Liao D, None; **Yang XL,** None; **Li R,** None.

REFERENCES

1 Cirafici P, Musolino M, Saccheggiani M, *et al*. Multimodal imaging findings and treatment with dexamethasone implant in three cases of idiopathic macular telangiectasia type 1. *Case Rep Ophthalmol* 2021;12(1):92-97.

2 Tapia Quijada HE, Mantolan Sarmiento C, Serrano García M, *et al*. Atypical bilateral presentation in idiopathic macular telangiectasia type 1. *Arch Soc Esp Oftalmol* (*Engl Ed*) 2021;96(11):602-606.

3 Pollmann AS, Mishra AV, Francisconi CLM. Ultra-widefield imaging of peripheral vascular abnormalities in idiopathic macular telangiectasia type 1. *J Vitreoretin Dis* 2021;5(6):549-552.

4 Maruko I, Iida T, Sugano Y, *et al*. Demographic features of idiopathic macular telangiectasia in Japanese patients. *Jpn J Ophthalmol* 2012;56(2):152-158.

5 Amoroso F, Miere A, Colantuono D, *et al*. Navigated focal laser of macular telangiectasia type I (adult-onset Coats' disease). *Ophthalmic Surg Lasers Imaging Retina* 2024;55(9):545-551.

6 Nakai M, Iwami H, Fukuyama H, *et al*. Visualization of microaneurysms in macular telangiectasia type 1 on optical coherence tomography angiography before and after photocoagulation. *Graefes Arch Clin Exp Ophthalmol* 2021;259(6):1513-1520.

7 Maki H, Imai H, Yamada H, *et al*. The long-term effect of cystotomy with fibrinogen clot removal for a cystoid macular edema secondary to idiopathic macular telangiectasia type 1: a case report. *Retin Cases Brief Rep* 2023;17(2):101-104.

8 Ranno S, Vujosevic S, Mambretti M, *et al*. Role of vitrectomy in nontractional refractory diabetic macular edema. *J Clin Med* 2023;12(6):2297.

9 Adelman RA, Parnes AJ, Bopp S, *et al*. Strategy for the management of macular edema in retinal vein occlusion: the European VitreoRetinal Society macular edema study. *Biomed Res Int* 2015;2015:870987.

10 Ivastinovic D, Haas A, Weger M, *et al*. Vitrectomy for diabetic macular edema and the relevance of external limiting membrane. *BMC Ophthalmol* 2021;21(1):334.

11 Bozdogan YC, Erol MK, Suren E, *et al*. Internal limiting membrane graft in full-thickness macular hole secondary to macular telangiectasia type 2. *J Fr Ophtalmol* 2021;44(9):1419-1424.

12 Stefánsson E. Ocular oxygenation and the treatment of diabetic retinopathy. *Surv Ophthalmol* 2006;51(4):364-380.

13 Li ZX, Zhang J, Lin T, *et al*. Macular vascular circulation and retinal oxygen saturation changes for idiopathic macular epiretinal membrane after vitrectomy. *Acta Ophthalmol* 2019;97(3):296-302.

14 Ahmed A, Dunk C, Ahmad S, *et al*. Regulation of placental vascular endothelial growth factor (VEGF) and placenta growth factor (PIGF) and soluble Flt-1 by oxygen—a review. *Placenta* 2000;21(Suppl A): S16-S24.

15 Jia LY, Sun YM, Ma K. Macular morphology in patients with diabetic retinopathy treated by ILM peeling: a propensity score-matched analysis. *Ophthalmic Surg Lasers Imaging Retina* 2021;52(8):420-425.