



CASE REPORT

Eales' Disease: A Successful Treatment with Oral Corticosteroid

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Abstract

Introduction: Eales' disease is an idiopathic inflammatory venous occlusion primarily affecting the peripheral retina. It commonly affects healthy young men aged 20-30 years, and 50-90% of patients develop bilateral involvement. Retinal changes include perivascular phlebitis, peripheral nonperfusion, and neovascularization. **Case Presentation:** We present a case of bilateral Eales' disease; a 38-year-old man presented to our hospital with chief complaints of decreased and blurred vision, photopsia, and floaters in both eyes. The best corrected visual acuity (BCVA) at the initial presentation was hand movement in the right eye (RE) and 0.3 in the left eye (LE). The anterior segment and intraocular pressure (IOP) were normal. Funduscopy of both eyes highlighted dense vitreous opacity. In the RE, we found perivascular sheathing, perivascular exudates, dot and flame shape intraretinal hemorrhages, and focal occlusion of retinal vessels. In the LE, we found perivascular sheathing, perivascular exudates, and dot and flame shape intraretinal hemorrhages. Initial optical coherence tomography (OCT) results show central macular thinning. The disease resolved rapidly with an oral corticosteroid before the patient underwent pars plana vitrectomy. **Conclusions:** Eales' disease can be treated with corticosteroids, anti-VEGF, laser photocoagulation, and pars plana vitrectomy. Even though the patient was planned for pars plana vitrectomy, we tried initial therapy using oral corticosteroids, which responded well to the patient's chief complaints. OCT results also showed improvement in macular thickness after the third follow-up. Oral corticosteroid therapy shows good results in Eales' disease treatment.

Keywords: Eales' disease; oral corticosteroid; vitreous opacity; retinal vascular change

Introduction

Eales disease is an idiopathic, occlusive perivasculitis primarily affecting the peripheral retina bilaterally. This condition is characterized by retinal nonperfusion, neovascularization, and recurrent vitreous hemorrhages.^[1] Eales disease was first identified in the United States, Canada, and the United Kingdom in the mid of the 19th century and early 20th century. Henry Eales^[2] first described this disease as a syndrome characterized by recurrent vitreous hemorrhage in young men, accompanied by epistaxis and constipation. Indian and Middle East have reported the most cases in the last decade, with the incidence being one in 200-250 ophthalmic patients. However, due to better health and lifestyle and lower tuberculosis (TB) rates, Eales disease appears to have declined globally.^[3] Most patients are young, healthy men between 20 and 40 years; the average onset age is typically younger among Asians than Caucasians.^[2] In Indonesia, there are no data regarding the prevalence of Eales disease, but in India, one in 135 referral ophthalmic patients and one in 200-250 general eye hospital patients have Eales' disease.^[4]

Inflammation (in the form of peripheral periphlebitis or vasculitis with perivascular exudates), retinal occlusion (superficial retinal hemorrhages, collaterals, and venovenous shunts, dot blot hemorrhages, and retinal edema), and neovascularization are three characteristic vascular responses in determining Eales disease's progression.^[5] The progression of Eales disease was divided by Charamis into four stages. In stage one, patients presented with a mild case of

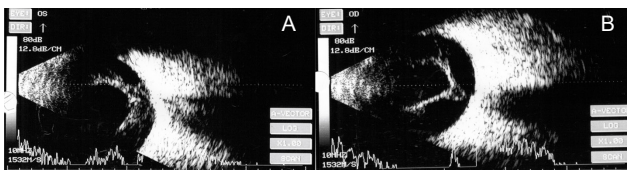


Figure 1. Ultrasonography of the (A) RE and (B) LE.

peripheral perivasculitis; stage two is characterized by severe periphlebitis that involves larger vessels; stage three was indicated by neovascularization and vitreous hemorrhage, and complications caused by traction led to stage four.^[5]

The majority of patients are asymptomatic during the inflammatory and occlusion phases. The disease starts with multiple peripheral, branch retinal vein occlusions caused by inflammation, the most common clinical manifestation is the presence of thin, solid white lines representing venous sheathing. Since the occlusions are primarily venous, they slowly develop, creating compensation manifestations such as microaneurysms, capillary telangiectasia, collaterals, corkscrew vessels, and venous beading. Long-term retinal ischemia causes peripheral neovascularization in 80% of eyes.^[6] When neovascularization causes vitreous hemorrhage, the typical symptoms of Eales disease manifest a unilateral sudden visual loss or blurring vision and floaters.^[6] Recurrence often happens and can result in traction retinal detachment due to gradual vitreous contraction, macular edema, secondary rhegmatogenous retinal detachment (RRD), and secondary neovascular glaucoma. Fundus fluorescein angiography help evaluates vasculitis,

ischemia, and neovascularization. It is essential to perform a B-scan ultrasound before surgery to evaluate posterior vitreous detachment (PVD), retina condition, and tractional membranes that may be hidden by vitreous hemorrhage.^[7] Several diseases cause retinal vascular sheathing or occlusion that may resemble Eales' disease, such as intermediate uveitis, multifocal choroiditis (ocular disease), systemic TB, syphilis (systemic inflammatory disease), and branch retina vein occlusion (noninflammatory diseases).^[7]

Early intervention of Eales disease (within 3–6 months) improve visual outcomes. Various therapeutic interventions, such as pharmacological treatment, laser therapy, and/or surgery, can be used individually or in combination, depending on the patient's condition and medical situation.^[4] Corticosteroids treat active vasculitis. Most unilateral cases respond to periocular injection corticosteroids (triamcinolone acetonide). Systemic corticosteroids (oral prednisolone, one mg/kg body weight/day) should be considered for bilateral or severe vasculitis or no response to periocular injections. Some patients need low-dose maintenance. These two routes of corticosteroids usually control Eales disease inflammation.^[6] The use of anti-VEGF (bevacizumab and ranibizumab) in Eales disease is expected to prevent retinal neovascularization and can be used as adjuvants to laser photocoagulation and vitrectomy.^[7] Peripheral laser photocoagulation in the ischemic areas bordering neovascularization is the treatment of choice for the proliferative stage. Treatment can be extended

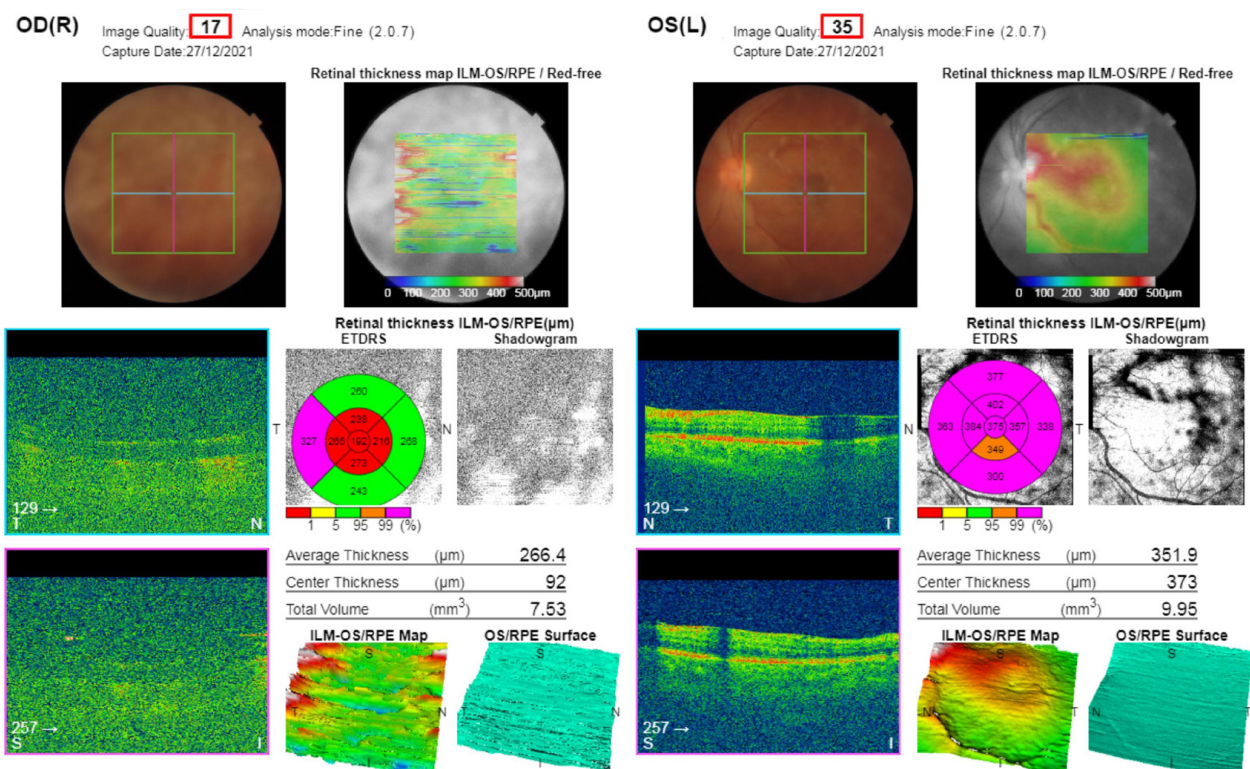


Figure 2. Initial macular OCT of the RE and LE.

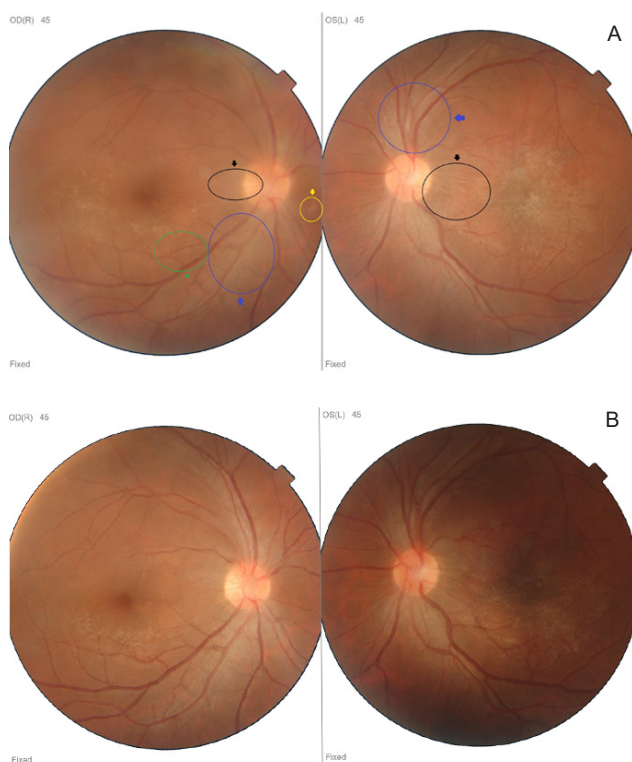


Figure 3. Fundus image of the RE and LE at (A) the first follow-up, There are perivascular sheathing (shown in blue circle), perivascular exudate (shown in yellow circle), flame-shape haemorrhages (shown in black circle), and focal occlusion of the retinal vessel (green circle) and (B) the second follow-up.

posteriorly depending on retinal ischemia spread and disc or posterior neovascularization. Photocoagulation is contraindicated in active vasculitis. In persistent vitreous hemorrhages, core vitrectomy is the choice of treatment.^[8] Eales' disease can lead to several complications, including cystoid macular edema, macular holes, persistent vitreous hemorrhage, traction retinal detachment, neovascular glaucoma, and epiretinal membrane formation.^[5]

Case presentation

In this case presentation, we present A 38-year-old man who came to Solo Eye Hospital, with a chief complaint of decreased and blurred vision in both eyes for the past two months. He also has symptoms of photopsia and floaters. There was no history of trauma, recurrent eye redness, eyeglasses, diabetes mellitus, hypertension, persistent cough, persistent vomiting, diarrhea for more than three weeks or weight loss, and free sex. General examination results within normal results. Ophthalmological examination revealed the best corrected visual acuity (BCVA) at the initial presentation was 0.3 in the left eye (LE) and hand movement in the right eye (RE). Ocular motility was normal with intraocular pressure (IOP) result 12.3mmHg on the RE and 13.0 on the LE. Anterior segment examination within normal limits in both eyes. In the posterior segment (funduscopy indirect) of RE, we found exudative retinal detachment, vitreous hemorrhage, blood dot retinal hemorrhage, and

neovascularization. We found vitreous hemorrhage, blood dot retinal hemorrhage, neovascularization, and posterior vitreous detachment (PVD) on the LE.

We consulted the patient to an internist to rule out other possibilities of systemic disease that can cause retinal vasculitis (ex. TB, syphilis, and toxoplasmosis) with the laboratory examination: leucocyte 7.800/ μ l, hemoglobin 14.6 g/dl, eritrosit 4.39 jt/ μ l, trombocvte 333.000/ μ , HbsAg (-), HIV non-reactive, fasting blood sugar 100 mg/dL, venereal disease research laboratory (VLDR) and treponema pallidum haemagglutination assay (TPAH) test negative, Anti Toxoplasma IgG & IgM negative, and X-ray thorax within normal limit. The internist stated that no systemic disease that could cause retinal vasculitis could be found in this patient.

The ultrasonography (USG) shows RE retinal detachment, while LE shows PVD (Figure 1). Initial macular optical coherence tomography (OCT) of both eyes highlighted that the macular image of the RE is less distinct due to the vitreous hemorrhage, whereas the LE shows macular edema (Figure 2). Based on history taking, ocular examination, USG, macular OCT, and labororium result, we diagnosed the patient with Eales disease and started the treatment with methylprednisolone 24 mg in the morning and afternoon.

A month after the therapy with an oral corticosteroid, the blurred vision and floaters in both eyes had already decreased. General examination results were normal. Ophthalmological examination revealed the best BCVA was 1.0 in the RE and 0.6 in the LE. Ocular motility and anterior segment examination were normal, IOP result of 13 mmHg on the RE and 14.7 on the LE. In the posterior segment examination in the RE, there is no sign of cystoid macular edema and decreased vitreous hemorrhage. There are neovascularization, cystoid macular edema, and vasculitis on the LE, with decreased vitreous hemorrhage.

A Fundus image of both eyes performed on the second visit highlighted dense vitreous opacity. In the RE, we found perivascular sheathing, perivascular exudates, dot and flame shape intraretinal hemorrhages, and focal occlusion of retinal vessels. In the LE, we found perivascular sheathing, perivascular exudates, dot and flame shape intraretinal hemorrhages (Figure 3A). The macular OCT shows thickening on the nasal and inferior segment of the RE macula and macular edema with subfoveal fluid in the LE (Figure 4A). We continue the treatment with methylprednisolone 16 mg in the morning and afternoon.

Three months after the therapy with an oral corticosteroid, blurred vision and floaters in both eyes were no longer present. General examination results were normal. Ophthalmological examination revealed that the BCVA was 1.0 in the RE and 1.0 in the LE. Ocular motility

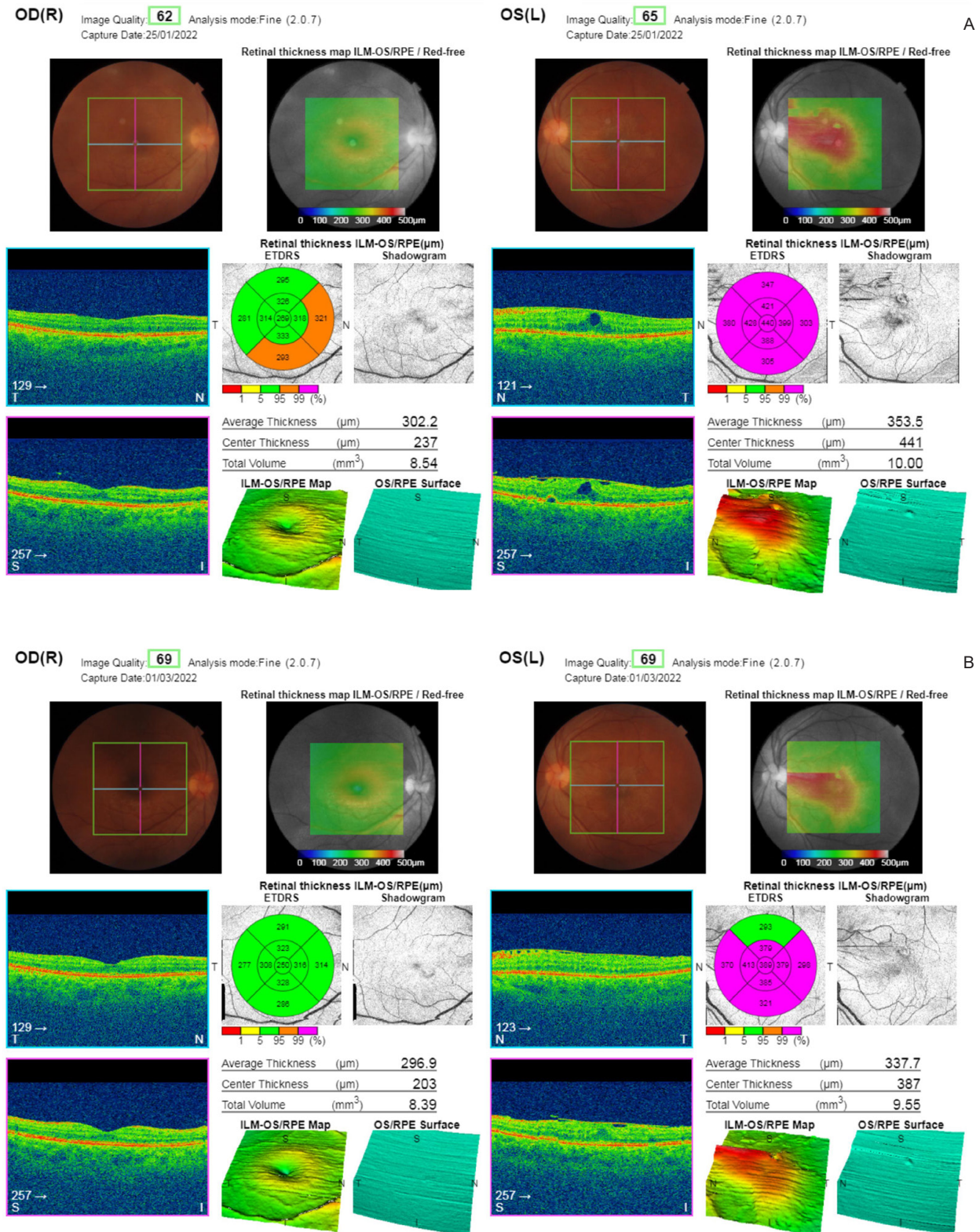


Figure 4. Macular OCT of the RE and LE at (A) the first follow-up, there is RE perimacular thickening and LE macular edema and (B) second follow-up.

and anterior segment examination were normal, with IOP result of 11.3 mmHg on the RE and 12,3 on the LE. In the posterior segment examination in the RE, there is no sign of cystoid macular edema and vitreous hemorrhage. There is decreased neovascularization, cystoid macular edema, and vasculitis on the LE. The fundus image of both eyes highlighted the decrease in perivascular sheathing and perivascular exudates with no hemorrhage in the RE, while in the LE, there are neovascularization, retinal

scarring, and perimacular exudates (Figure 3B), while the third macular OCT shows normal RE macular and macular thickening on LE (Figure 4B). OCT angiography in RE is normal, while LE shows epiretinal membrane (ERM) (Figure 5). We continue the treatment with methylprednisolone 8 mg in the morning.

Discussion and conclusions

Eales’ disease is an idiopathic inflammatory venous occlusion that primarily affects the peripheral retina

affecting predominantly healthy young men between the ages of 20 and 40. Changes to the retina include extensive periphery ischemia, perivascular sheathing, and neovascularization. Recurrent vitreous hemorrhages are the most common cause of vision loss. In Eales disease, 75% of patients have floaters, and 60% have blurred vision due to vitreous hemorrhage, although the patient often has no symptoms early in retinal perivasculitis. The patient with bilateral Eales' disease is around 50-90%.^[9] Retinal neovascularization occurs in up to 80% of patients. Neovascularization can form on the disc (NVD) or retina (NVE). This neovascularization bleeding and vitreous hemorrhage are common and often recurring, causing vision loss. Isolated vitreous hemorrhages usually resolve without vision impairment.^[6]

USG is needed to detect retinal detachment, whether traction, rhegmatogenous, or combined, when vitreous hemorrhage obscures the view of the fundus.^[10] Peripheral vasculitis is an essential sign of this disease. It looks like sheathing in peripheral blood vessels due to peripheral exudate. Signs of retinal vein blockage can appear when many of these veins are affected.

Eales' disease is treated symptomatically. It aims to reduce retinal perivasculitis and vitreous, reduce the risk of vitreous hemorrhage caused by forming new blood vessels in the retina and/or optic nerve fibers due to retinal detachment, and surgically remove unabsorbed vitreous hemorrhage and/or vitreous membrane. Corticosteroids, anti-VEGF therapy, photocoagulation with or without anterior retinal cryoablation, and vitrectomy are used in various disease stages.^[8]

In this case, a 38-year-old man came to our hospital complaining of decreased and blurred vision, photopsia, and floaters in both eyes. The BCVA at the initial presentation was hand movement in the RE and 0.3 in the LE. The anterior segment and IOP were normal. We found vitreous hemorrhage, blood dot retinal hemorrhage, and neurovascularization in the RE and LE posterior segment. USG shows RE retinal detachment in this patient, while LE shows PVD.

The patient has no history of trauma, recurrent eye redness, eyeglasses, diabetes mellitus, hypertension, persistent cough, persistent vomiting, diarrhea for more than three weeks or weight loss, and free sex. Based on lab results and internist examination, we can rule out TB, HIV, syphilis, and toxoplasma as causes of retinal vasculitis in this patient. It is important to rule out systemic infections and noninfectious diseases that can mimic Eales' disease symptoms, such as TB, syphilis, sarcoidosis, toxoplasmosis, systemic lupus erythematosus, and Behçet disease.^[11]

Initial funduscopy of both eyes highlighted dense vitreous opacity. In the RE, we found perivascular sheathing, perivascular exudates, dot and flame shape

intraretinal hemorrhages, and focal occlusion of retinal vessels. We found perivascular sheathing, perivascular exudates, dot and flame shape intraretinal hemorrhages in the LE. Initial macular OCT of both eyes highlighted that the macular image of the RE is less distinct due to the vitreous hemorrhage, whereas the LE shows macular edema. Although Eales' disease primarily affects the peripheral retina, macular involvement is possible. Macular edema and the epiretinal membrane was the most common symptom.^[12]

We choose first-line therapy for Eales disease, a corticosteroid. We choose corticosteroids as therapy because of several benefits: systemic corticosteroids are the primary treatment for active vasculitis, particularly in cases of severe bilateral involvement^[13]; corticosteroids are recommended for widespread vasculitis, vision-threatening vasculitis, and macular edema because corticosteroids reduce retinal inflammation^[4]; corticosteroid is more affordable than other therapy choices, such as anti-VEGF therapy, photocoagulation, and vitrectomy; patient does not need hospitalization and can be treated as an outpatient; no risk of infection after injection or surgery; and in most cases, Eales disease responds very well to corticosteroids. However, corticosteroids can cause side effects such as weight gain, mood changes, difficulty sleeping, increased blood sugar, increased risk of infections, cataracts, and glaucoma.^[14] Thus, we must monitor the patient monthly to adjust the dosage and stop treatment after recovery. Other therapies, such as anti-VEGF and vitrectomy, are also considered if the patient's condition does not improve (ex., severe neovascularization and persistent vitreous hemorrhage) during monthly control to the hospital.

We gave oral methylprednisolone instead of a periocular injection because this patient has bilateral Eales diseases. We start with methylprednisolone with an initial dosage of 0.6-1 mg/kg weight of the patient. In this patient, we use 24 mg given in the morning and afternoon and tapering off to 8 mg in the morning during four months of therapy. The patient responded well to oral corticosteroid therapy; blurred vision and eye floaters were absent. His BCVA was 1.0 in the RE and 1.0 in the LE, and vitreous hemorrhage already subsided in both eyes. Macular OCT results also showed improvement in macular thickness after the third follow-up. OCT angiography in RE is normal, while LE shows ERM. We also do optical coherence tomography angiography (OCT-A) in monitoring Eales' disease because it is a non-invasive imaging technique that can detect neovascularization and show deep capillary plexus changes. In patients with macular involvement, OCT-A showed enlarged superficial capillary plexus (SCP) and deep capillary plexus (DCP) areas.^{[15],[16]} Since there is an ERM in LE OCT-A, we considered performing membrane

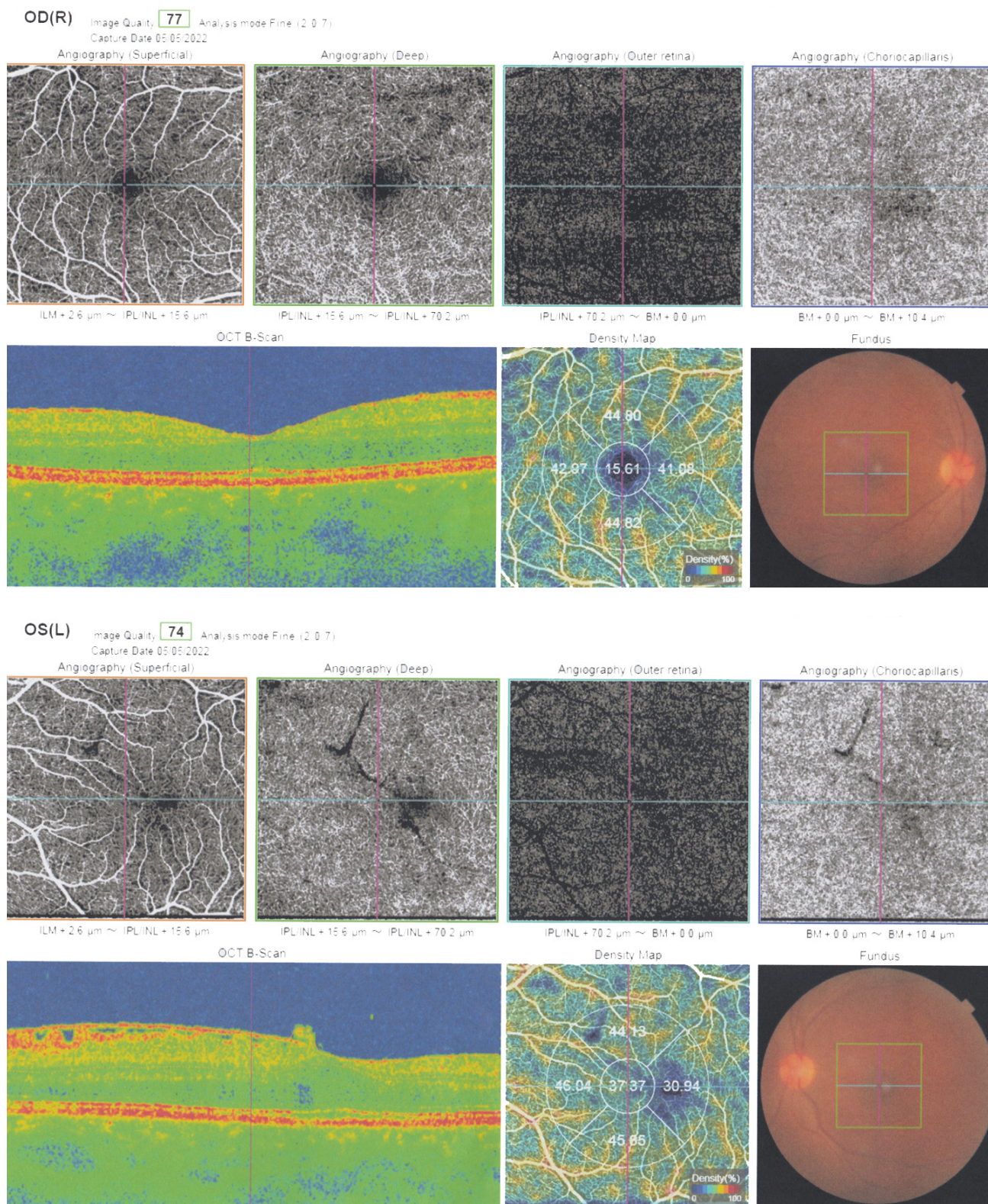


Figure 5. Angiography image of the RE and LE.

peeling surgery on the LE if the patient perceives visual impairment in the future, which is usually the patient experiences distortion (the patient sees curved or crooked lines).

Corticosteroids are the primary therapy for Eales' disease in the active perivasculitis stage to control inflammation. Oral and topical corticosteroids are used to control retinal vasculitis. Dosage must be adjusted for

each patient based on inflammation severity. In this case, oral corticosteroid therapy shows good results in Eales' disease treatment.

References

[1] Kim SJ, Fawzi A, Kovach JL, Patel S, Recchia FM, Sobrin L, et al., editors. Section 12: Retina & Vitreous. 2022-2023 Basic and Clinical Science Course, San Francisco: American Academy of Ophthalmology; 2022.

- [2] Arévalo JF, editor. Retinal and Choroidal Manifestations of Selected Systemic Diseases. New York: Springer; 2013. <https://doi.org/10.1007/978-1-4614-3646-1>.
- [3] Dalvin LA, Smith WM. Intraocular manifestations of mycobacterium tuberculosis: A review of the literature. *J Clin Tuberc Other Mycobact Dis* 2017;7:13–21. <https://doi.org/10.1016/j.jctube.2017.01.003>.
- [4] Biswas J, K.R. R, Pal B, Gondhale HP, Kharel (Sitaula) R. Long-term outcomes of a large cohort of patients with Eales' Disease. *Ocul Immunol Inflamm* 2018;26:870–876. <https://doi.org/10.1080/09273948.2017.1298817>.
- [5] Bandello F, Querques G, editors. Vascular Anomalies of the Fundus Oculi: Diagnosis and Treatment. Medical Retina (ESASO Course Series), vol. 1, Paris: Karger; 2012, p. 67–77. <https://doi.org/10.1159/isbn.978-3-8055-9991-7>.
- [6] Wilkinson CP, Hinton DR, Sadda SR, Wiedemann P, Schachat AP. *Ryan's Retina: 3 Volume Set*. 6th ed. Elsevier; 2017.
- [7] Murillo López S, Medina Medina S, Murillo López F. Eales' disease: epidemiology, diagnostic and therapeutic concepts. *Int J Retina Vitreous* 2022;8:3. <https://doi.org/10.1186/s40942-021-00354-0>.
- [8] Biswas J, Ravi RK, Naryanasamy A, Kulandai LT, Madhavan HN. Eales' disease - current concepts in diagnosis and management. *J Ophthalmic Inflamm Infect* 2013;3:11. <https://doi.org/10.1186/1869-5760-3-11>.
- [9] Murugeswari P, Shukla D, Kim R, Namperumalsamy P, Stitt AW, Muthukkaruppan V. Angiogenic potential of vitreous from proliferative diabetic retinopathy and Eales' Disease patients. *PLoS One* 2014;9:e107551. <https://doi.org/10.1371/journal.pone.0107551>.
- [10] Fu U. B-Scan ophthalmic ultrasonography of retinal detachment. *Am J Biomed Sci Res* 2019;4:327–328. <https://doi.org/10.34297/AJBSR.2019.04.000827>.
- [11] Calugaru D, Calugaru M, El Ghali C. Eales disease in a young adult man Case report. *Rom J Ophthalmol* 2017;61:306–309.
- [12] Goel N, Kumar V, Arora S, Jain P, Ghosh B. Spectral domain optical coherence tomography evaluation of macular changes in Eales disease. *Indian J Ophthalmol* 2018;66:433. https://doi.org/10.4103/ijo.IJO_845_17.
- [13] Lopes BO, Brízido MS, Reis AI, Miranda MM, Pina SM. Eales' Disease: When the rare sounds Frequent. *Case Rep Ophthalmol Med* 2021;2021:1–6. <https://doi.org/10.1155/2021/1056659>.
- [14] Grennan D, Wang S. Steroid Side Effects. *JAMA* 2019;322:282. <https://doi.org/10.1001/jama.2019.8506>.
- [15] Rajurkar K, Thakar M, Gupta P, Rastogi A. Comparison of fundus fluorescein angiography, optical coherence tomography and optical coherence tomography angiography features of macular changes in Eales disease: A case series. *J Ophthalmic Inflamm Infect* 2020;10:34. <https://doi.org/10.1186/s12348-020-00220-4>.
- [16] Alyousif NA, Alsalamah AK, Nowilaty SR, Al-Dhibi HA. Optical coherence tomography angiography features of macular epiretinal neovascularization in Eales' disease: A case report. *J Ophthalmic Inflamm Infect* 2022;12:16. <https://doi.org/10.1186/s12348-022-00289-z>.