Case Report

Taiwan J Ophthalmol 2025;15:487-490

Access this article online

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DOI:
10.4103/tjo.TJO-D-23-00049

A very unusual case report: Retinal neovascularization and tractional retinal detachment in a patient with thalassemia minor

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Abstract:

Although many ocular pathologies have been reported in patients with thalassemia major, there is a limited number of studies on the ocular findings of patients with thalassemia minor. In this report, thalassemia minor was detected in a 42-year-old male patient who presented with unilateral sudden vision loss and underwent pars plana vitrectomy due to proliferative retinopathy and tractional retinal detachment. One month after the surgery, the best-corrected visual acuity of the eye was 20/100, and the retina was fully reattached under silicone oil. Patients with thalassemia minor should undergo ophthalmologic screening with fundus examination, including the peripheral retina.

Keywords:

Pars plana vitrectomy, proliferative retinopathy, sea-fan neovascularization, thalassemia minor, tractional retinal detachment

Introduction

Thalassemias are an autosomal recessive, heterogeneous group of genetic disorders that result from a damaged synthesis of globin chains of hemoglobin and are more prevalent in the Mediterranean countries, including Turkey. Although various ocular abnormalities associated with thalassemia major (ocular surface problems, cataracts, angioid streaks, dystrophic retinal changes, retinal vascular abnormalities, and optic disc drusen) have been reported, there are only a few reports about the ocular findings of thalassemia minor.[1-4]

In this case report, the management of a middle-aged man with thalassemia minor and proliferative retinopathy is presented.

Case Report

A 42-year-old man was referred to our ophthalmology clinic with sudden visual impairment that started 1.5 months ago in his left eye. He was diagnosed with hypertension and started amlodipine 1 mg × 5 mg per day 2 months ago. At presentation, his best-corrected visual acuity (BCVA) was 20/20 in the right eye and 20/100 in the left one. Anterior segments and intraocular pressure were unremarkable in both eyes. Fundus examination showed superotemporal retinal ischemia with ghost vessels and neovascularization as sea-fan configuration associated with tractional retinal detachment with macular dragging in the left eye [Figure 1a] while the right eye was normal. Cystoid changes were detected in optical coherence tomography, and fluorescein angiography was compatible with sea-fan neovascularization, peripheral

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Submission: 29-03-2023
Accepted: 31-05-2023
Published: 22-09-2023

How to cite this article: Acan D, Karahan E. A very unusual case report: Retinal neovascularization and tractional retinal detachment in a patient with thalassemia minor. Taiwan J Ophthalmol 2025;15:487-90.

ischemia, and leakage from the newly formed vessels at the edge of this ischemic area [Figures 1a and b and 2b]. Other peripheral retinal quadrants of the affected eye and the fellow eye were examined in detail, and there was no ischemia or neovascularization elsewhere. This cynical appearance was thought to be a branch retinal vein occlusion related to hypertension. However, the patient's systemic blood pressure was 130/85 mmHg at the time of admission. The patient was consulted to the hematology department for further investigation due to his relatively young age and unusual findings.

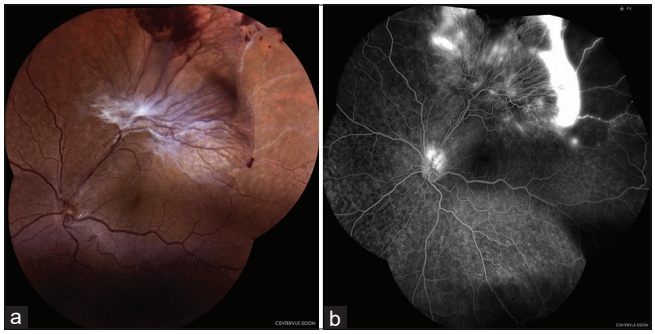

Figure 1: (a and b) Fundus photography of the left eye shows superotemporal nonperfusion with ghost vessels, retinal hemorrhages, and sea-fan neovascularization at the edge of the ischemic area. A fibrous component with traction is also observed. The superotemporal ischemic area and adjacent sea-fan neovascularization in fluorescein angiography of the left eye. Note that also optic disc shows leakage

The hematological evaluation revealed a hemoglobin level within normal limits; whereas the mean corpuscular volume, mean corpuscular hemoglobin (MCH), and MCH concentration were 74.1 fL/cell (range: 80–99.9 fL), 24.2 pg/cell (range: 27.0–31.0 pg), and 32.6 g/dL (range: 33.0–37 g/dl), respectively, and they were below normal limits. Peripheral smear showed anisocytosis without atypical cells. Hematologic consultation was concluded as the patient has thalassemia minor.

Laser photocoagulation (LP) was performed on the peripheral severely ischemic retina, avoiding the tractional detached area and neovascularization. However, 20 days later, vitreous hemorrhage was observed [Figure 2a-c]. Four days after the intravitreal bevacizumab injection, 23-gauge pars plana vitrectomy (PPV) was performed for tractional retinal detachment. Following core vitrectomy and vitreous base shaving, tractional membranes and internal limiting membrane were peeled, the retina was reattached with room air tamponade, and LP was performed to the residual far peripheral ischemic retinal areas. The vitreous cavity was filled with silicone oil due

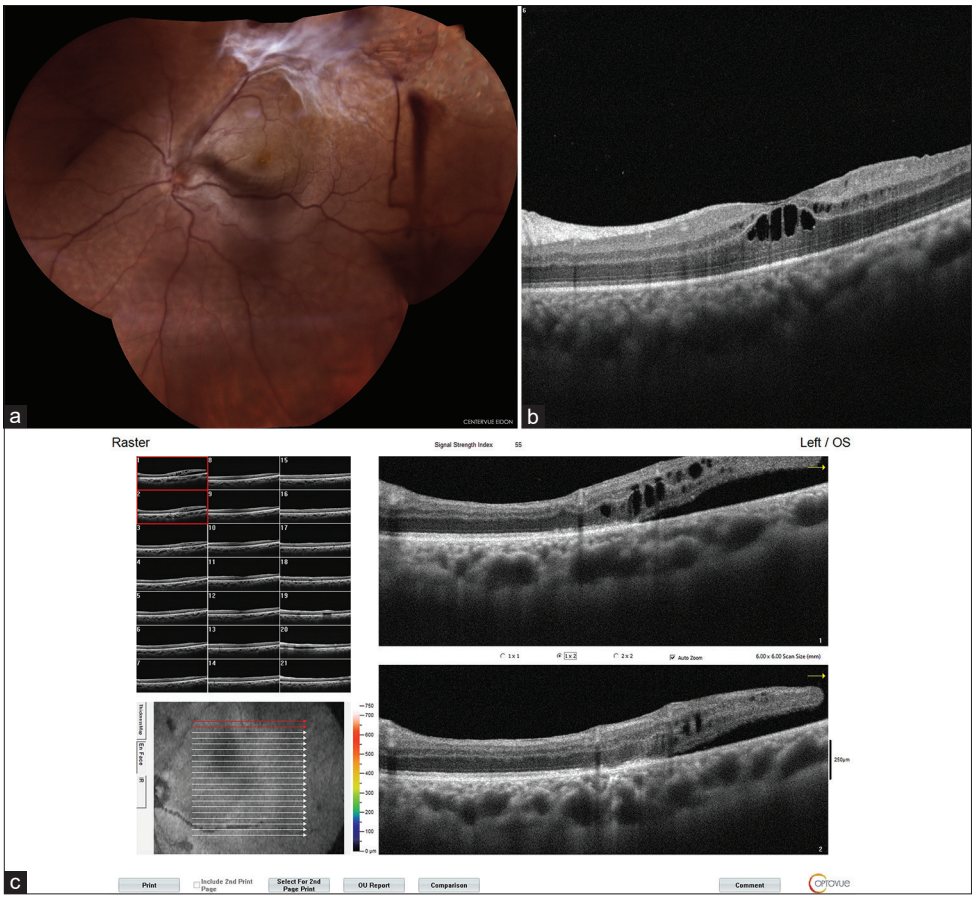

Figure 2: Photo (a) and optical coherence tomography (OCT) images(b-c) of vitreous hemorrhage and tractional retinal detachment after laser treatment. Cystoid spaces were shown with OCT, and traction excited by the fibrous tissue toward the superotemporal neovascular area was clearly distinguished

to an iatrogenic retinal tear formed during tractional membrane peeling. One month after the surgery, the BCVA of the left eye was 20/100, and the retina was fully reattached under silicone oil [Figure 3]. At the last examination, the dragging of the macula was completely gone, and the retina was completely attached.

Discussion

Unlike thalassemia major with homozygous mutation and chronic severe anemia requiring blood transfusions regularly, iron chelation, splenectomy, as well as hematopoietic transplantation, thalassemia minor with a single abnormal globin allele is usually asymptomatic and does not require any treatment. Serious ocular pathologies have been identified as a result of anemia, orbital bone marrow expansion, repeated blood transfusion, iron overload, and chelation therapy in patients with thalassemia major.[1-4] Regular ophthalmological examination is recommended for these patients, particularly those who are under chelation therapy.[2] However, ocular side effects due to thalassemia minor are much less common.

In one of the previous few reports, retinal hemorrhages and detachment in a 22-year-old African American male with a thalassemia minor were observed.[5] From a different perspective, the frequency of thalassemia among patients with intraocular hemorrhages of unknown etiology was emphasized in an article published in 1969.[6] However, detailed information about the fundus of these patients such as ischemia or neovascularization was not provided. In another study involving 96 patients with thalassemia minor, the high percentage of ocular involvement (33.3%) was interpreted by the authors as a result of the alteration of retinal function due to insufficient oxygen perfusion of the retinal receptive units.[7] Decreased central or paracentral retinal sensitivity, narrowing of isopters, peripheral scotomas, and blind spot enlargement were observed in the perimetric tests of the patients. A hypercoagulable state in thalassemia was emphasized, and it was reported that in patients who do not need regular transfusion with hemoglobin levels <9–10 g/dL, ineffective erythropoiesis and chronic hemolysis cause the production of thrombogenic red blood cells and microparticles and increase the risk of thrombosis, particularly in older ages.[8] The hemoglobin of the patient in this report was 14.4 g/dL and within normal limits. However, the patient still developed ischemic retinal vein thrombosis. Although hypertension may also have been triggered, the fundus findings of the patient were different from classical branch retinal vein occlusion. From this point of view, we thought that although anemia is much less severe in thalassemia minor patients, thromboembolism and ischemia-related pathologies can be encountered if a more detailed routine ophthalmological examination is performed.

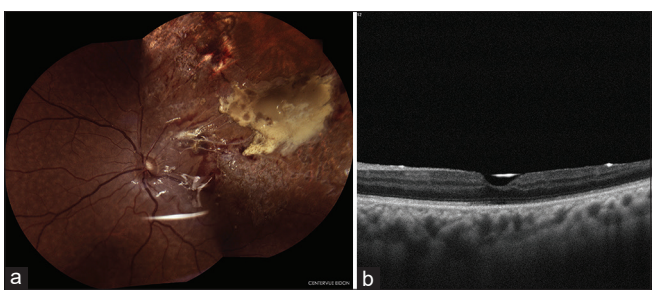

Figure 3: One month after pars plana vitrectomy, the retina is fully reattached under silicone oil tamponade(a), and no traction or dragging on the macular region(b)

Recently, two patients with beta-thalassemia minor as the cause of unilateral proliferative retinopathy were reported.[9] Significant peripheral ischemia and neovascularization were prominent in these cases. In our case, the tractional retinal detachment was present in addition to these findings; the patient was treated with PPV and silicone oil tamponade as it could not be stabilized by laser therapy. To the best of our knowledge, our case is the first case in the literature to have PPV applied for tractional retinal detachment due to thalassemia minor.

Conclusion

Thalassemia minor should be considered in the differential diagnosis of patients presenting with proliferative retinopathy, and patients with thalassemia minor should also be screened for ocular complications intermittently; furthermore, the peripheral retina should be evaluated in detail by dilated fundoscopy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published, and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Data availability statement

The datasets generated during and/or analyzed during the current study are available from the corresponding author on reasonable request.

Financial support and sponsorship

Nil.

Conflicts of interest

The authors declare that there are no conflicts of interest in this article.

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