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**CASE REPORT**

# Central retinal vein occlusion in a young adult Case report

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## Abstract

Purpose. To report the case of a 48-year-old man with unilateral central retinal vein occlusion.

Methods. The clinical, hematologic and hypercoagulability evaluations of the patient were thoroughly and specifically carried out.

Results. The central retinal vein occlusion was of nonischemic type and was associated with polyglobulia, hyperleukocytosis, hypercholesterolemia, multiple dental foci, and pulmonary sequelae of tuberculosis.

Conclusions. When a patient less than 50 years of age experiences central retinal vein occlusion, other mechanisms, such as the hyperviscosity syndrome or the inflammatory condition (inflammation of the central retinal vein) should be specifically considered and accounted for.

**Keywords:** central retinal vein occlusion, young adult, intravitreal bevacizumab, hyperviscosity syndrome, focal phlebitis

## Introduction

Central retinal vein occlusion (CRVO) is a significant cause of vision impairment and can occur at any age [1]. However, 90% of the CRVO patients are older than 50 years at the disease occurrence and only 10% of them are younger than 40 years [2]. The etiology can be quite varied, but age can be helpful in determining the differential diagnosis. Patients older than 50 years usually have common systemic vascular conditions such as hypertension and diabetes. However, when a CRVO occurs in a patient of less than 50 years old, other mechanisms should be specifically considered and accounted for.

Herein, we present a case of central retinal vein occlusion in a young adult, given the issues that may arise from the establishment of the positive, differential and etiopathogenic diagnose of the disease.

## Case presentation

A 48-year-old man presented to the clinic with complaints of decreased and blurred vision as well as photopsias in his right eye over the previous 3 months. The patient's medical history revealed primary pulmonary tuberculosis characterized by the primary complex in the

chest, 13 years before, which had been treated for 8 months with full recovery. No other systemic disease has been reported. Additionally, the patient presented multiple dental foci. His best-corrected visual acuity on presentation was 20/100. Anterior segment and intraocular tension were normal. Dilated ocular fundus examination found dotted and flame-shaped intraretinal hemorrhages throughout the fundus, often along the nerve fiber layer in all 4 quadrants, engorgement and tortuosity of the major retinal veins, papilloretinal edema, telangiectatic capillary bed, and small cotton wool spots located in the area of the optic nerve head and alongside the temporal vessels (Fig. 1). The visual field using the Goldmann perimeter was normal and the Humphrey static achromatic automatic perimetry (central 30-2 threshold test) exhibited a significant enlargement of the blind spot (the big blind spot syndrome [BBSS]) (Fig. 2). The macular optical coherence tomography (Stratus OCT, Carl Zeiss Meditec, Dublin, CA) revealed subretinal fluid with serous detachment of the macula, thickening of retina up to 400 microns, and small cystic changes within the neurosensory retina (Fig. 3). The ocular ecography showed that the optic nerve head area was elevated with 1.5 mm (Fig. 4).

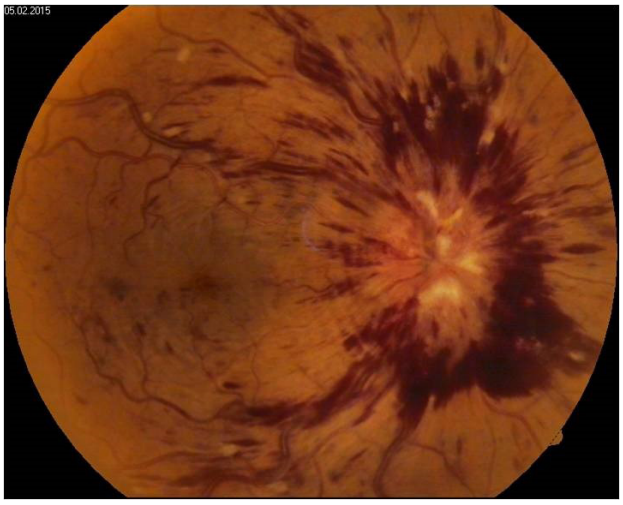



**Fig. 1** A 48-year-old man with nonischemic retinal vein occlusion in his right eye. Dilated fundus examination revealed flame-shaped hemorrhages in all 4 quadrants, papilloretinal edema, and small cotton wool spots

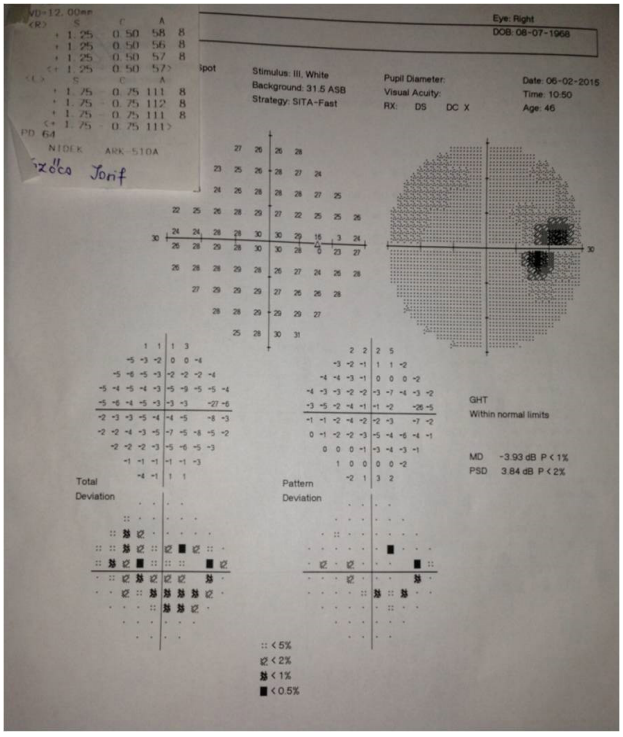



**Fig. 2** The Humphrey static automatic achromatic perimetry (central 30-2 threshold test) showing a significant enlargement of the blind spot (the BBSS)

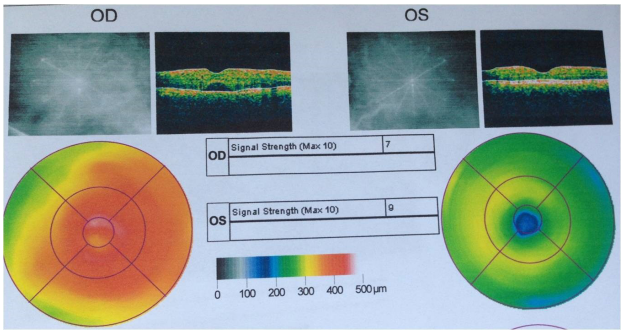
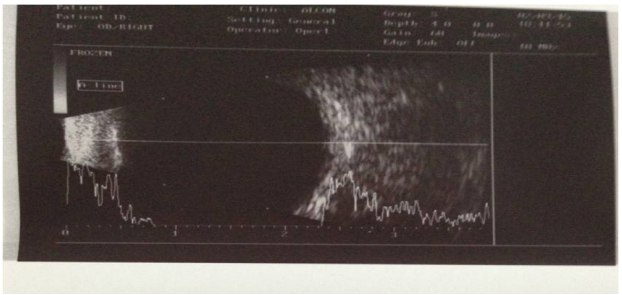



**Fig. 3** The Stratus OCT of the macula exhibiting subretinal fluid, serous detachment of the macula, thickening of retina up to 400 microns, and small cystic changes within neurosensory retina



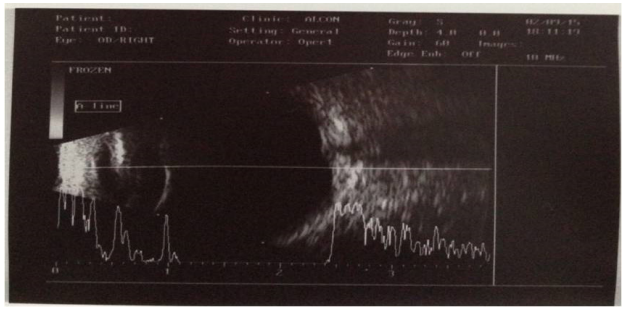


Fig. 4 The ocular ultrasonography revealing a 1.5mm elevation of the optic nerve head area

The pneumo-phtisiological examination included tuberculin intradermal reaction that identified pulmonary sequelae of tuberculosis (calcified primary complex) with no signs of disease reactivation. Results of the serologic testing revealed polyglobulia (6,2 million of red blood cells; 52% hematocrit), hyperleukocytosis (12,360 white blood cells) with neutrophilia (94%) and lymphomonocitopenia (4.9% and 0.7%, respectively), and hypercholesterolemia (183.8 mg/ dL low-density lipoprotein cholesterol). The rest of the hematologic evaluations were normal (antithrombin III, macroglobulins, paraproteins, total proteins and serum protein electrophoresis, cryoglobulins, plasma homocysteine, factor V [proaccelerin], factor V Leiden, antiphospholipid antibodies [anticardiolipin antibodies and lupus anticoagulant], cytomegalovirus, Lyme titer, C and S anticoagulant proteins, activated protein C resistance, antinuclear antibody test, syphilis serology [VDRL], and test for the human immunodeficiency virus).

Taking into account all the clinical, hematologic and hypercoagulability assessments performed, the diagnoses of unilateral nonischemic central retinal vein occlusion in a young adult; polyglobulia; hyperleukocytosis; hypercholesterolemia; multiple dental foci; and pulmonary sequels of tuberculosis, were established.

The treatment consisted of 4 consecutive intravitreal injections of bevacizumab (Avastin; Genentech Inc., South San Francisco, CA, USA) administered off-label at a dose of 2.5 mg (0.1 ml) per injection [3], with each injection spaced approximately 45 days apart. Steroids as well as hyperosmotic agents and carboanhydrase inhibitors were systemically applied. After 4 months of treatment, visual acuity improved to 20/ 40 and then to 20/ 20 after 1 year of observation. No recurrence of the disease occurred.

## Discussion

CRVO (central retinal vein occlusion) was labeled as being of nonischemic type [4], due to the following findings: visual acuity with mild impairment of 20/ 100 at presentation, which improved to 20/ 20 at the end of the follow-up; mild tortuosity and engorgement of the retinal vessels; absence of the relative afferent pupillary defect, which was a marker of extended retinal ischemia; milder degrees of intraretinal hemorrhages in all 4 quadrants; moderate papilloretinal edema; normal peripheral visual field without central scotoma; presence of the BBSS; small number of cotton wool spots.

The differential diagnosis of our case had to be done with other diseases that cause intraretinal hemorrhages and disc swelling (Table 1). The differentiation was quite easy taking into account the age as well the clinical, hematologic and hypercoagulability evaluations of our reported case.

**Table 1.** Conditions that were differentiated from our reported case

| | |
|---|---|
| Diabetic retinopathy | Inflammatory optic neuropathy |
| Hypertensive retinopathy | Behcet's disease |
| CRVO in patients older than 50 years | Sarcoidosis |
| Eales disease | Systemic lupus erythematosus |
| Sickle cell disease | Tuberculosis |
| Ischemic optic neuropathy | Syphilis |
| Optic disc swelling | Lyme disease |
| Papillary drusen | Purtscher retinopathy |
| Antiphospholipid antibody syndrome | Dysproteinemias |
| Hyperhomocysteinemia | Acquired immunodeficiency syndrome |

We believe that the CRVO pathogenesis in our reported case was a multifactorial process. There was an aggregate of elements that contributed to the destabilization of a preexisting hemodynamic balance (Table 2).

**Table 2.** Factors that could be involved in the pathogenesis of central retinal vein in our reported case

| |
|---|
| **Predisposing factors:** |
| - Natural constriction of central retinal vein at the site of the lamina cribrosa; |
| - Lamina cribrosa rigidity; |
| - Focal phlebitis/ periphlebitis. |
| **Destabilizing and precipitating factors:** |
| - Blood hyperviscosity (polyglobulia, hyperleukocytosis, and hypercholesterolemia); |
| - Slowing the blood flow down with subsequent arterial perfusion insufficiency; |
| - Turbulence of the hemodynamics; |
| - Changes in the local fibrinolysis and coagulation; |
| **Mechanical factors:** |
| - Shear stresses due to hyperviscosity causing damage and proliferation of the vascular endothelial cells; |
| - Leukostasis; |
| - Throttle mechanism by thrombus |

For the venous occlusion picture to become manifest in humans there must exist a narrowing of venous lumen while it passes through the optic nerve head [5]. This narrowing is always localized beyond the visible portion of central retinal vein or behind the lamina cribrosa. Predisposing factors responsible for this localized narrowing of the vein are manifold, e.g. natural constriction of central retinal vein at the site of the lamina cribrosa [6]; lamina cribrosa rigidity; focal phlebitis and periphlebitis. This venous inflammation in our case could be due to a nonspecific hyperergic reaction triggered by the pulmonary sequelae of tuberculosis and/ or by the multiple dental foci. A series of destabilizing and precipitating factors came to add to those mentioned above. They were represented by hemorheological anomalies i.e., slowing the blood flow down with subsequent arterial perfusion insufficiency, blood hyperviscosity (due to polyglobulia, hyperleukocytosis, and hypercholesterolemia), and changes in the local fibrinolysis and coagulation. Additionally, mechanical factors involved were shear stresses due to hyperviscosity causing damage and proliferation of the vascular endothelial cells, leukostasis, and eventually a throttle mechanism [7] in the central retinal vein in the region of the lamina cribrosa perhaps by thrombus formation.

## Conclusions

When a patient less than 50 years old experiences a central retinal vein occlusion, other mechanisms, such as a hyperviscosity syndrome or an inflammatory condition should be specifically considered and accounted for.

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**Disclosures**

The authors have no proprietary of commercial interest in any of the materials discussed in this article; they declare no conflict of interest. The authors have nothing to disclose: NONE.

## References


1. Călugăru D. Etiologia ocluziei venei centrale a retinei. Oftalmologia. 2011; 55(2):27-37.
2. Prisco D, Marcucci R. Retinal vein thrombosis: risk factors, pathogenesis, and therapeutic approach. Pathophysiol Haemost Thromb. 2002; 32(5/6):308-311.
3. Călugăru D, Călugăru M. Injection scheme for intravitreal bevacizumab therapy for macular oedema due to central retinal vein occlusion: results of a muticenter study. Acta Ophthalmol. 2016; 94(1):e80-e81.
4. Călugăru D, Călugăru M. Intravitreal bevacizumab in acute central/ hemicentral retinal vein occlusions: three-year results of a prospective clinical study. J Ocul Pharmacol Ther. 2015; 31(2):78-86.
5. Kaur C, Fulds WS, Ling EA. Blood-retinal barrier in hypoxic ischaemic conditions: basic concepts, clinical features, and management. Progress in Retinal and Eye Research. 2008; 27(6):622-647.
6. McAllister, Gilles ME, Smithies LA, Rochtchina E, Harper CA, Daniell MD, Constable IJ, Mitchell P. The central retinal vein bypass study: a trial of laser-induced chorioretinal venous anastomosis for central retinal vein occlusions. Ophthalmology. 2010; 117(5):954-965.
7. Williamson TH. A "throttle" mechanism in the central retinal vein in the region of the lamina cribrosa. Br J Ophthalmol. 2007; 91(9):1190-1193.