

Case Report

# Presentation of acute central retinal vein occlusion in scleroderma



Faisal Malik<sup>a</sup>, Ahmed Al Habash<sup>b,\*</sup>

## Abstract

Central retinal vein occlusion (CRVO) is a rare complication of scleroderma. Here we report a case of a 30-year-old man who was diagnosed to have scleroderma in the rheumatology and dermatology clinic. During treatment with systemic steroids and immunosuppressive therapy the patient developed a sudden decrease of vision in the right eye and was diagnosed to have right CRVO with macular edema on fundus examination. After three consecutive Intravitreal bevacizumab (IVB) injections for macular edema, best-corrected visual acuity (BCVA) improved from 20/80 to 20/25. All ocular and systemic causes of CRVO other than scleroderma were excluded in our patient by thorough clinical examination and investigations, suggesting that scleroderma was the most possible etiology in his condition.

**Keywords:** Central Retinal Vein Occlusion, Scleroderma, Systemic sclerosis, Eye manifestations, Intravitreal bevacizumab (IVB), Macular edema

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## Introduction

Scleroderma or systemic sclerosis (SSc) is a generalized chronic connective tissue disorder of unknown etiology.<sup>1</sup> One of the critical abnormalities in SSc is a diffuse fibroproliferative vasculopathy, most commonly involving the microvasculature system,<sup>2</sup> which includes perivasculitis and fibrinoid degeneration of blood vessels.<sup>1</sup> Scleroderma usually appears in patients between 35 and 55 years, women being 2–3 times more frequently affected than men.<sup>1</sup>

Scleroderma patients commonly report eye problems, with dry eyes being a typical complaint. Conjunctiva and eyelid abnormalities are also often seen, including injection, telangiectatic vessels and stiff or tight eyelids. Diffuse iris atrophies and lens opacities are also experienced, although the use of corticosteroids as part of treatment is likely to

be the cause of the latter of these.<sup>1</sup> However, involvement of the posterior segment is often subclinical and visual loss as a direct result of the disease is rare.

Here we report a case of Central retinal vein occlusion (CRVO) with decreased vision, which is a rare complication of scleroderma. Patient was treated with Intravitreal bevacizumab (IVB) injections for macular edema and responded well with vision improvement. To our knowledge, CRVO with scleroderma was reported only twice in the literature.<sup>3,4</sup> We herein report a third case with that very rare complication.

## Case report

A 30-year-old man was diagnosed as scleroderma in the rheumatology and dermatology clinic three years ago. He

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<sup>a</sup> Department of Ophthalmology, King Fahd Hospital of the University, Khobar, Saudi Arabia

<sup>b</sup> Department of Ophthalmology, College of Medicine, University of Dammam, Dammam, Saudi Arabia

\* Corresponding author at: Department of Ophthalmology, College of Medicine, University of Dammam, P.O. Box 31010, Khobar 31952, Saudi Arabia. Mobile: +966 507017777.

e-mail addresses: [ahmedalhabash@gmail.com](mailto:ahmedalhabash@gmail.com), [ahabash@ud.edu.sa](mailto:ahabash@ud.edu.sa) (A. Al Habash).



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presented with features of thickening, tightening and induration of the facial skin. There were associated sclerodactyly, periungual telangiectasia, digital pitting, calcinosis in the right fourth finger and positive Raynaud's phenomena. History was negative for any medical problems other than scleroderma.

All investigations at presentation which include thyroid functions, complete blood count (CBC), prothrombin time (PT), partial thromboplastin time (PTT), random blood sugar (RBS), renal function tests (RFTs), liver function tests (LFTs), rheumatoid factor, HLA-B51, HLA-27, anti-double-stranded DNA, anti-scleroderma antibody, anti-JO-1, anti-ribonucleoprotein, anti-Sjogren's antibody SSA, Anti-Sjogren's antibody SSB, C3, C4 complement levels, centromere protein antibody, Smith antibody, anti liver/kidney microsomal antibody (ANA) speckled ((1:320)), erythrocyte sedimentation rate (ESR) 67 mm/h normal (0–20) mm/h and C-reactive protein (CRP) 0.4 mg/dl normal (0–0.3) mg/dl.

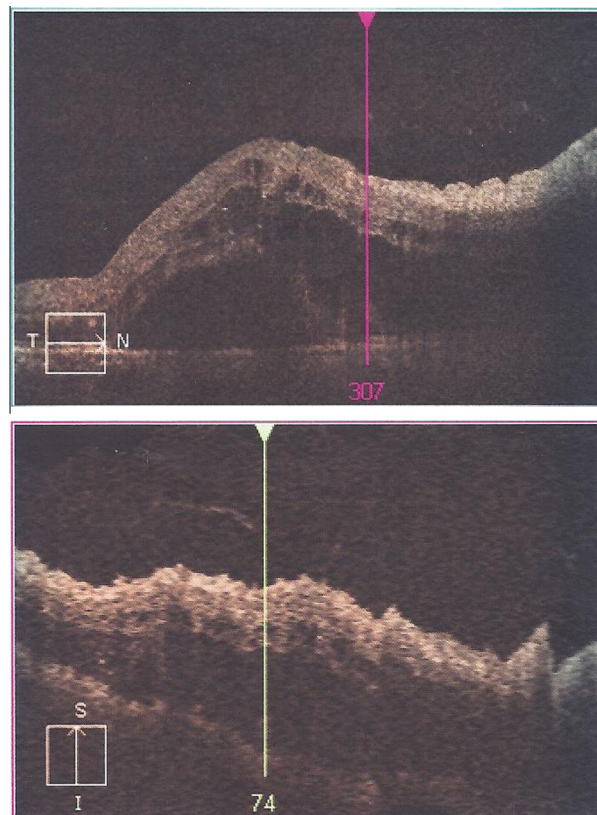
A treatment in the form of oral prednisolone and mycophenolate mofetil (Cellcept) was started since that time.

As the patient was on routine follow up in the ophthalmology clinic for dry eye, an incidental finding of tortuous retinal vessels in both eyes on fundus examination was documented.

Two years after the patient was diagnosed to have scleroderma, he presented to our clinic with a sudden decrease of vision in the right eye of one-day duration. On examination his Blood pressure was 125/75 mmHg, and best-corrected visual acuity (BCVAs) were 20/80 and 20/20 in the right and left eyes, respectively. Both pupils found to be normal with no relative afferent pupillary defect (RAPD). Anterior segment was within normal for both eyes. Intraocular pressure was normal in both eyes. The fundus of the right eye showed tortuous vessels with multiple retinal hemorrhages, cotton wool spots and optic nerve swelling (Fig. 1). Fundus fluorescein angiography (FFA) showed capillary nonperfusion of less than 10 disk diameters. Macular edema and neurosensory detachment were also detected in the right eye by optical coherence tomography (OCT) (Fig. 2). Fundus examination of the left eye showed mild tortuous vessels (Fig. 3).



**Figure 1.** Tortuous vessels with multiple retinal hemorrhages, cotton wool spots and optic nerve swelling in the fundus of the right eye.



**Figure 2.** OCT of the right macula on presentation showing macular edema with neurosensory detachment.



**Figure 3.** Mild tortuous retinal vessels of the left eye.

Based on the previous findings, a diagnosis of non-ischemic CRVO in the right eye was made, in which the patient received three consecutive IVB injections for the macular edema on monthly basis.

After six-months of follow up, his right BCVA was improved to 20/25 and the macular edema was resolved (Fig. 4). Retinal hemorrhages, cotton wool spots and optic nerve swelling were resolved leaving only a track of some sclerosed blood vessels with no neovascularization in the retina, iris or the iridocorneal angle. Intraocular pressure was

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