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Case report

Vasculitic central retinal vein occlusion: The presenting sign of seronegative rheumatoid arthritis

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ABSTRACT

Purpose: To report the case of a patient who presented with a vasculitic central retinal vein occlusion (CRVO), which was the result of an undiagnosed systemic inflammatory condition, seronegative rheumatoid arthritis (RA).

Observations: The patient presented with reduced vision in the left eye and polyarthralgia. Fundoscopic examination revealed a central retinal vein occlusion (CRVO) with concurrent evidence of vasculitis. Work-up for polyarthralgia included comprehensive serologic testing for connective tissue disease, including Vectra® disease activity (DA) testing. Results of these studies confirmed the diagnosis of seronegative rheumatoid arthritis (RA). Systemic steroid therapy was initiated with subsequent anatomic and visual improvement.

Conclusions and importance: We hypothesize that the systemic inflammation—a hallmark of RA—led to the development of a vasculitic CRVO and, thus, the retinal manifestations served as the disease marker that prompted thorough work-up of the patient's disease, even in the face of initial seronegativity. This case serves as a reminder that, in the setting of CRVO and polyarthralgia, systemic inflammatory conditions must be considered as the underlying etiology. Further, this case report highlights our evolving understanding of the role that serologic markers play in the diagnosis and monitoring of RA.

1. Introduction

Rheumatoid arthritis (RA) is estimated to affect 0.8% of the world's population, making it the most common inflammatory arthritis [1]. It is characterized by a painful, persistent, and often symmetrical polyarthritis that primarily involves the synovial tissues. Additionally, RA can be categorized and broadly divided into two categories based on serology: seropositive RA and seronegative RA. Approximately 40% of seropositive RA patients experience extra-articular involvement of multiple organ systems, including the eyes [2]. Less is known about seronegative RA and its extra-articular features, but generally speaking seronegative RA is felt to be less aggressive, with fewer joint erosions and better response to treatment than seropositive RA [3]. The ophthalmic sequelae of RA vary widely and range from relatively benign findings, such as keratoconjunctivitis sicca and episcleritis, to serious vision-threatening conditions, including anterior scleritis, necrotizing scleritis, scleromalacia perforans, peripheral ulcerative keratitis and retinal vasculitis secondary to posterior scleritis [4].

1.1. Case report

Personal identifying information was removed from this report because informed consent to publish such information was not obtained. Our patient presented, in the sixth decade of life, with acute painless vision loss in the left eye that had progressed over two weeks. In addition, the patient had noticed new onset floaters. The patient had no past ocular, medical, surgical, or pertinent family history, was not taking any medications and at presentation (as well as on subsequent visits) had a normal blood pressure. Further questioning did reveal that the patient had been experiencing progressive joint stiffness of the wrists and hands for several months; these symptoms were most prominent in the morning and seemed to improve as the day progressed. The involved joints were mildly edematous and tender to the touch at the metacarpophalangeal joints of both hands, suggesting the presence of active synovitis. Despite the progressive nature of this pain, the patient had not sought medical attention and had not taken any

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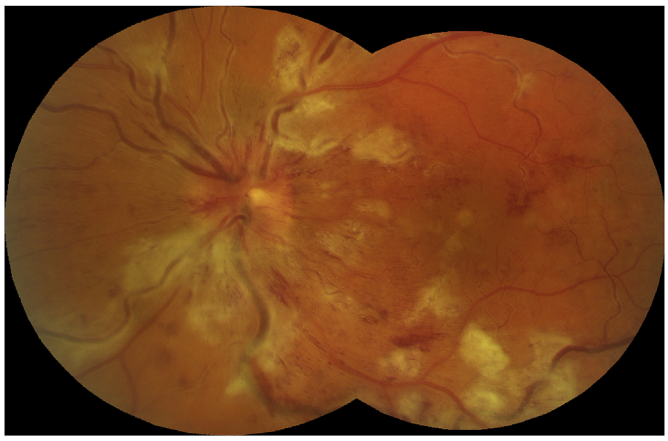

Fig. 1. Widefield color fundus photograph shows dilated and tortuous veins with scattered flame-shaped hemorrhages, cotton-wool spots and perivascular exudation.

medication to manage the pain. In an attempt to uncover a possible unifying etiology for the combination of polyarthralgia and decreased vision in the left eye, the patient was specifically asked about a history of hypertension, hyperlipidemia, hypercoagulable states, immunocompromised states and autoimmune disease; all of which the patient denied.

On ophthalmic examination, the patient's Snellen visual acuity (BCVA) was 20/20 in the right eye (OD) and count fingers (CF) at 2 feet in the left eye (OS). Intraocular pressures were 15 mm Hg OD and 16 mm Hg OS. Anterior segment examination was unremarkable. Importantly, no cell or flare was detected in the anterior chamber. Dilated fundus exam of the left eye showed vitreous cell, dilated and tortuous vessels with perivascular exudation, scattered flame-shaped hemorrhages and significant macular edema—all consistent with the diagnosis of a central retinal vein occlusion (CRVO) (Fig. 1). In the early phase, widefield fluorescein angiography (FA) demonstrated multiple venous filling defects with associated stretches of capillary non-perfusion (Fig. 2A). In the late phase (Fig. 2B), widefield fluorescein angiography was positive for venous staining and showed mild perivenous leakage. Spectral-domain optical coherence tomography (SD-OCT) detected extensive intraretinal fluid (between the outer nuclear layer and the ellipsoid zone), scattered intraretinal fluid in the other layers and mild vitritis (Fig. 3A). Examination of the OD was unremarkable. The patient was started on frequent difluprednate 0.05% drops in the OS, and a broad workup for inflammation and vaso-occlusive disease was initiated in conjunction with the Rheumatology service.

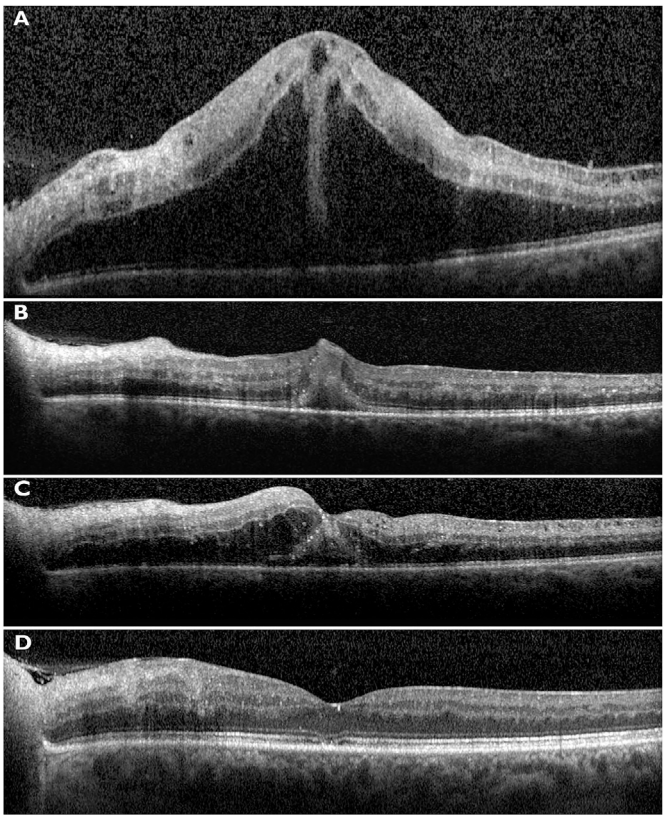

Fig. 3. **(A)** Spectral-domain optical coherence tomography (SD-OCT) at the time of presentation showed significant macular edema and vitreous cell; **(B)** On day three of the initial topical steroid therapy, SD-OCT showed marked improvement of the intraretinal fluid; **(C)** On day ten, the macular edema recurred despite continued topical therapy. Intravitreal ranibizumab was administered at this time. **(D)** At 3.5 months of monthly ranibizumab treatment, the patient's visual acuity improved to 20/30 and there was a resolution of the macular edema.

At three days' follow-up, the patient's VA OS had improved to 20/80 in the left eye, and there was marked improvement of the intraretinal fluid on SD-OCT without any steroid mediated intraocular pressure increase (Fig. 3B). Based on this response, a sub-Tenon injection of triamcinolone acetonide was used to supplement the topical therapy. Surprisingly, however, the initial work-up for occlusive vascular disease was largely uninformative: aside from a homogenously-positive anti-nuclear antibody (ANA; Titer 1:80) and a mildly elevated Erythrocyte Sedimentation Rate (ESR; 29 mm/h). Other antibodies such as rheumatoid factor (RF), anti-

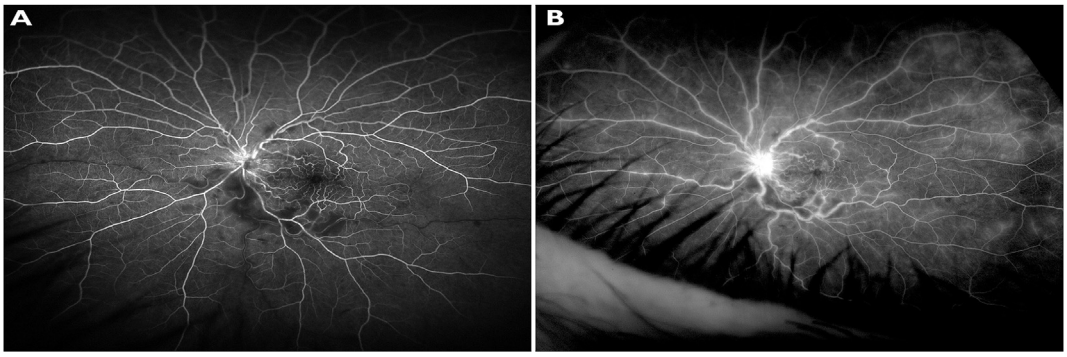

Fig. 2. **(A)** Early phase widefield fluorescein angiography showed capillary non-perfusion. **(B)** Late phase widefield fluorescein angiography showed staining of the vessel wall and perivascular leakage.