



Idiopathic retinal arterial occlusive vasculitis in the setting of multiple arterial occlusions

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ABSTRACT

Purpose: We present a patient with vaso-occlusive retinal vasculitis to summarize this uncommon entity and review the clinical features and management challenges applicable to patients with retinal vasculitis.

Observations: A 76-year-old male presented with sudden-onset severe central vision loss. On examination, vitreous hemorrhage, neovascularization of the optic nerve, peripheral segmental periphlebitis, vessel sclerosis, vascular sheathing, and retinal hemorrhages were observed, and a diagnosis of active vaso-occlusive retinal vasculitis was made. The patient then underwent a complete infectious, inflammatory, and neoplastic workup which returned negative. The patient was treated with locally with a sub-Tenon's injection of 40 mg triamcinolone on presentation and later with oral prednisone. At three-month follow-up, vision improved to 20/300 with regressing neovascularization and clearing of vitreous hemorrhage in the right eye (OD).

Conclusions: Considering novel associations of occlusive retinal vasculitis, it is important to recognize that idiopathic occlusive retinal vasculitis, although uncommon, can occur and represents a prototypical disease form. It is imperative that these patients have a complete infectious, inflammatory, and neoplastic workup owing to the possible overlap of masquerade clinical signs and symptoms.

1. Introduction

Retinal vasculitis is a sight-threatening inflammatory condition characterized by perivascular inflammation that may result in vascular occlusion.¹ Causes of retinal vasculitis are classified as either secondary to infection, neoplasia, or other systemic inflammatory diseases.² Depending on etiology, retinal vasculitis can be associated with predominant arterial or venous involvement or both.² Rarely, retinal vasculitis may occur as an isolated idiopathic condition.³ Vision loss due to retinal vasculitis can occur secondary to cystoid macular edema and macular ischemia; moreover, retinal neovascularization can produce vitreous hemorrhage, fibrovascular proliferation, or tractional retinal detachment.

Here we present a patient with idiopathic vaso-occlusive retinal vasculitis to summarize this uncommon entity and review the clinical features and management challenges applicable to patients with retinal vasculitis.

2. Case presentation

A 76-year-old male with a medical history of asthma, arthritis and hypertension presented with sudden-onset severe central vision loss in the right eye (OD). Ophthalmic examination demonstrated a best corrected visual acuity (BCVA) of 2/200 OD and 20/30 in the left eye (OS). Pupillary response, intraocular pressures and anterior segment exam were unremarkable. Dilated fundus examination OD revealed vitreous hemorrhage, neovascularization of the optic disc (NVD), peripheral segmental periphlebitis, vessel sclerosis, vascular sheathing, retinal hemorrhages, and areas of retinal whitening surrounding the macula. No vitreous cells or acute vitritis was noted (Fig. 1). Dilated fundus examination OS revealed mildly sclerotic vessels along with mild vitreous opacities either from vitreous hemorrhage or old inflammation. No signs of acute vitritis were present (Fig. 1). No hearing loss, tinnitus, or peripheral neuropathy was noted on history and physical examination. Optical coherence tomography (OCT) demonstrated inner retinal thinning in both eyes (Fig. 2). Fluorescein angiography revealed

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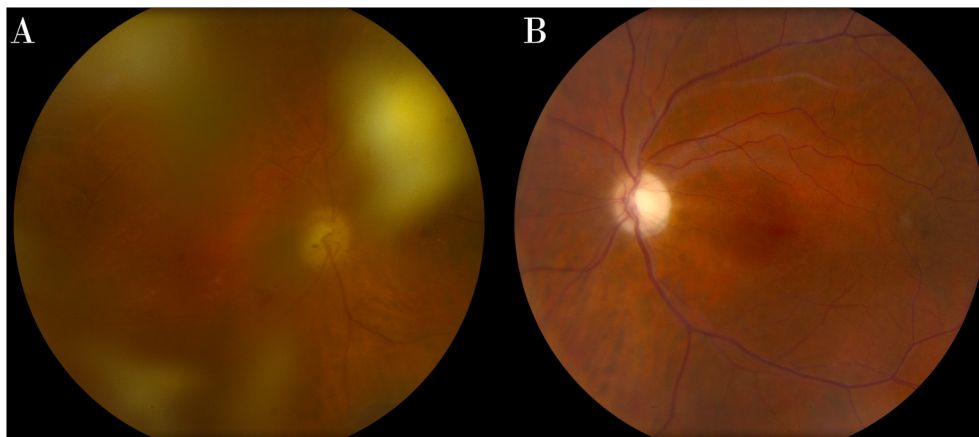


Fig. 1. Fundus photograph of right eye (A) shows vitreous hemorrhage, neovascularization of the optic disc, peripheral segmental periphlebitis, vessel sclerosis, vascular sheathing, and trace retinal hemorrhage. Left eye (B) reveals sclerotic vessels, optic disc pallor, and mild vitreous opacities.

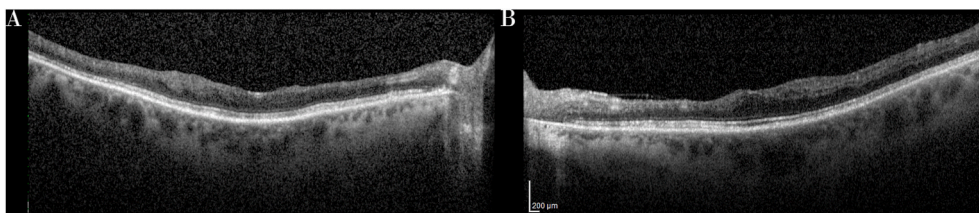


Fig. 2. Optical coherence tomogram illustrates inner retinal thinning secondary to macular ischemia more prevalent in right eye (A) compared to left eye (B).

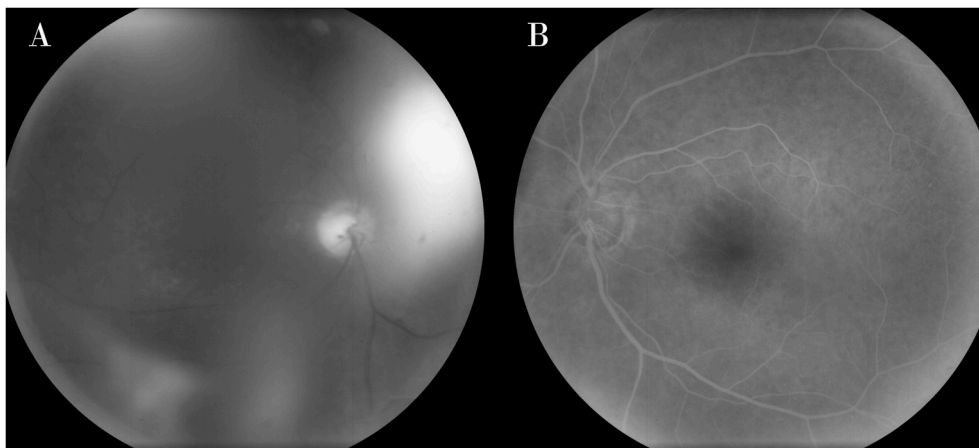


Fig. 3. Fluorescein angiogram demonstrating late hyperfluorescence in the mid-periphery secondary to vascular leakage in the right eye (A). There are no signs of abnormal late leakage seen in the left eye (B).

significant late hyperfluorescence secondary to vascular leakage OD, and no abnormal late leakage OS (Fig. 3).

The patient underwent a complete infectious, inflammatory and neoplastic workup including syphilis serology, Lyme serology antinuclear antibody, rheumatoid factor, homocysteine, antineutrophil cytoplasmic antibody, angiotensin converting enzyme, complete blood count with differential, erythrocyte sedimentation rate, C-reactive protein and lysozyme. Additional testing consisted of HLA-DR1, HLA-DR4, HLA-B27, pathergy for Behcet’s disease, anterior chamber polymerase chain reaction testing for toxoplasmosis and herpes viruses (herpes simplex virus, herpes zoster virus, cytomegalovirus, Epstein-Barr virus), tuberculosis serum quantiferon, magnetic resonance imaging of the head, chest plain film radiography, computerized tomography scans of the chest, abdomen and pelvis and positron emission tomography. All

workup testing and imaging was negative.

Given the constellation of clinical findings - including perivascular inflammation, loss of vision from macular ischemia, negative infectious, neoplastic, or systemic inflammatory causes - a diagnosis of active idiopathic vaso-occlusive retinal vasculitis was made. The patient was treated locally with a sub-Tenon’s injection of 40 mg triamcinolone on the day of presentation. After negative work-up, the patient was treated by their primary care doctor with oral prednisone. At three-month follow-up, vision improved to 20/300 with regressing neovascularization and clearing of vitreous hemorrhage OD.

3. Discussion

This case highlights the presentation of idiopathic vaso-occlusive

retinal vasculitis associated with multiple branch retinal artery occlusions in 2nd order vessels in the superotemporal quadrant. Idiopathic vaso-occlusive retinal vasculitis was first described by Jampol et al.⁵ and is characterized by retinal microinfarcts with subsequent retinal neovascularization resulting in retinal hemorrhages and vitreous hemorrhage.

Arterial occlusions in active retinal vasculitis are likely a result of thrombotic vascular changes induced by local inflammatory-mediated endothelial injury.⁶ Additionally, active vascular sheathing or cuffing with perivascular inflammatory infiltrate is typical of this idiopathic form of retinal vasculitis.⁶ In this case, we believe the patient's NVD was most likely secondary to ischemia from multiple branch retinal artery occlusions. PRP was initially considered for the patient given the level of neovascularization. However, the patient was treated with a sub-Tenon's triamcinolone injection to reduce active local ocular inflammation. Full regression of NVD was observed on follow-up and, thus, PRP was not pursued. Complete visual recovery is not common in this clinical setting due to irreversible macular ischemia. Given inner retinal atrophy was observed on initial OCT and the patient's complicated history of idiopathic retinal vasculitis OS, this may have been an initial chronic presentation of her retinal vasculitis OD.

Of note, there are reports of intraocular lymphoma as a cause of retinal vasculitis findings such as periphlebitis.⁷ Given this patient's older age, lymphoma was considered as a cause of their retinal findings however no significant findings were seen clinically, on blood work, as well as computerized tomography imaging.

Recently, there has been significant discussion regarding occlusive retinal vasculitis reported after intravitreal injection of brolocizumab.⁸⁻¹³ While this pathogenic mechanism is unclear, patients demonstrate similar findings such as intraocular inflammation, evidence of vascular occlusion and retinal ischemia presenting after intravitreal injection.⁹ There are also several reports hemorrhagic occlusive retinal vasculitis (HORV) after uncomplicated cataract surgery with prophylactic intracameral vancomycin.¹⁴⁻¹⁷ HORV often presents with macular ischemia, sectoral retinal hemorrhage along venules, as well as retinal vasculitis and arterial occlusion.¹⁴ The postulated mechanism of injury in HORV is thought to be a hypersensitivity or drug-mediated reaction.

4. Conclusion

We present a patient with idiopathic vaso-occlusive retinal vasculitis to characterize this uncommon entity and review the clinical features and management challenges in patients with retinal vasculitis. Given the novel associations of occlusive retinal vasculitis after intravitreal injection of brolocizumab and uncomplicated cataract surgery with prophylactic vancomycin, it is important to recognize that idiopathic occlusive retinal vasculitis, although uncommon, can occur and represents a prototypical disease form. Since idiopathic vaso-occlusive retinal vasculitis is a diagnosis of exclusion, it is imperative that these patients have a complete infectious, inflammatory, and neoplastic workup owing to the possible overlap of masquerade clinical signs and symptoms.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

- PW: No disclosures
- EC: Consultant (Alcon, Allergan, Chendgu Kanghong Biotechnology, Genentech, Novartis, Ophthotech, Opthea); Cofounder/Equity Holder (Citrus Therapeutics)
- DA: Advisory Board, Consultant, Speaker (Alcon, Allergan, Bayer, Genentech, Novartis, Regeneron); Cofounder, Equity Holder (Citrus Therapeutics)

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