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Case report

Simultaneous central retinal artery occlusion and optic nerve vasculitis in Crohn disease

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ABSTRACT

Purpose: To describe a case of Crohn disease presenting as occlusive vasculitis resulting in a central retinal artery occlusion (CRAO) in one eye and transient ischemic optic neuropathy in the fellow eye.
Observations: An 18-year-old patient recently diagnosed with biopsy-proven Crohn disease presented with CRAO OD after a previous episode of transient visual loss OS. Extensive workup was negative for other autoimmune or infectious etiologies. The patient was started on intravenous methylprednisolone for 72 h followed by maintenance dose of azathioprine and oral prednisone. Signs of inflammation resolved gradually with some improvement of visual acuity despite developing optic atrophy.
Conclusions and importance: To our knowledge, this is the first case of unilateral CRAO and bilateral optic nerve occlusive vasculitis in Crohn disease, which should be considered as an etiology of retinal vascular occlusive disorders especially in young patients. It is important for ophthalmologists to be aware of the ophthalmic risks associated with Crohn disease as aggressive treatment with systemic steroids and immunosuppressive agents is often needed.

1. Introduction

Crohn disease is a granulomatous inflammatory bowel disease first reported by Crohn in 1925.[1] It is characterized by skipped transmural inflammation mainly affecting the distal ileum. Crohn disease is the most common inflammatory bowel disease that is associated with extraintestinal manifestations. In fact, about 5–10% of affected patients present with ocular complications of both the anterior and posterior segments of the eye.[2] Such complications include kertatitis, uveitis as well as retinal vasculitis. In this article, we report the first case of simultaneous central retinal artery occlusion (CRAO) and optic nerve vasculitis in the fellow eye of an 18-year-old man with biopsy-proven Crohn disease (Fig. 1).

2. Case report

An 18-year-old man was hospitalized in our institution for the management of an acute gastro-intestinal exacerbation of a recently diagnosed Crohn disease. He also complained of a sudden painless loss of vision in the right eye two days prior to hospital admission. He also reported a 5-min transient superior altitudinal field loss in the left eye one week prior to his admission. There was no other previous ocular complaint. The patient was diagnosed with biopsy-proven Crohn disease four months prior to presentation (Fig. 1). Over two weeks prior to presentation, the patient reported fatigue, a 15-lbs weight loss, watery diarrhea, fever, myalgia, and arthalgia. At the time of ophthalmic consultation, medical therapy had not yet been initiated.

On examination, visual acuity was hand motion in the right eye and 20/200 improving to 20/70 with pinhole in the left eye. There was a relative afferent pupillary defect (RAPD) in the right eye. The left eye pupil was found to be very slow to react to light. A ciliary flush was noted in both eyes. Anterior chamber exam of both eyes showed presence of 1 + cells. The dilated fundus exam of the right eye showed diffuse retinal whitening compatible with CRAO with juxtapapillary cilioretinal sparing (Fig. 2A). The exam was normal in the left eye (Fig. 2B). Fluorescein angiography (FA) of the right eye showed extreme delay in retinal circulation (Fig. 2C and E). The FA of the left eye was significant for mid and late phases increasing hyperfluorescence of the optic nerve (Fig. 2D and F). The patient blood work-up was remarkable for an elevated C-reactive protein of 184 mg/dl and a leukocytosis of 22.9 10^9 per L. His immunological and hypercoaguable workup for lupus anticoagulant, anti-neutrophil cytoplasmic antibodies, protein C, S and anti-thrombin

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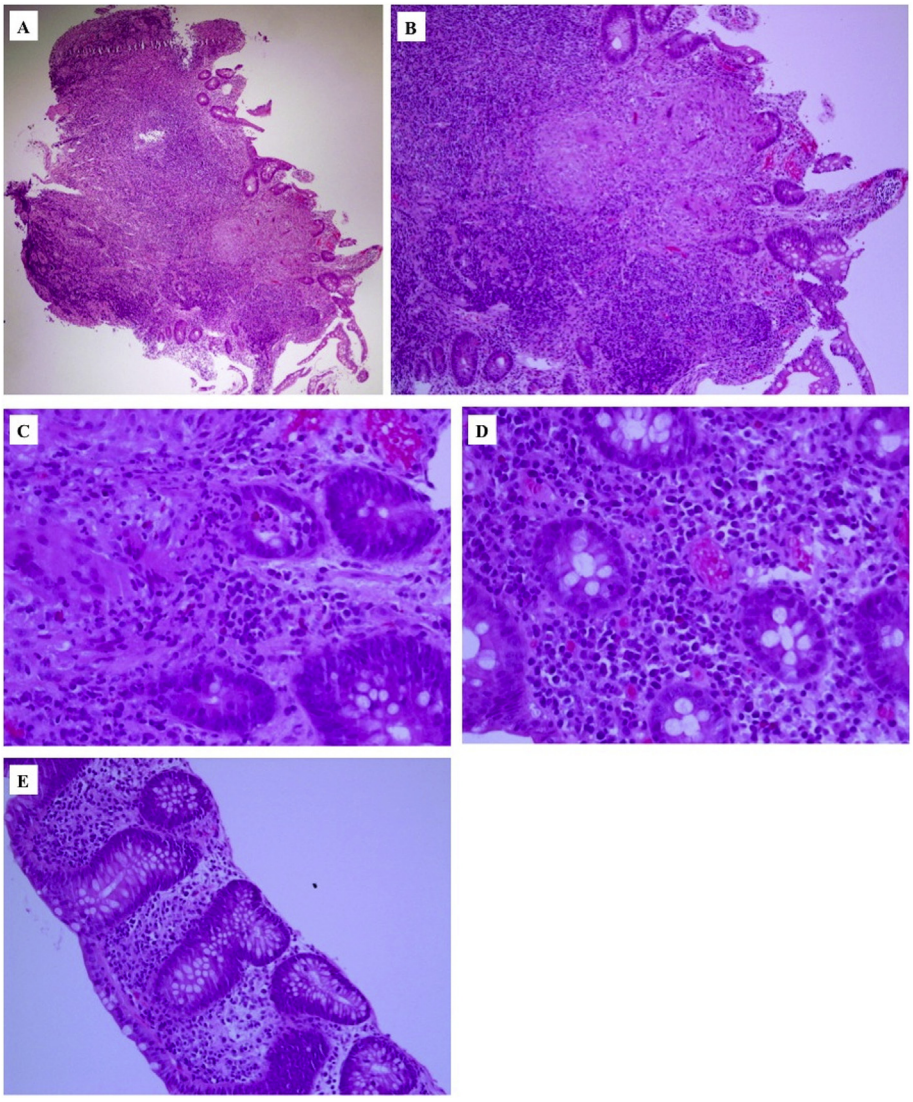

Fig. 1. H&E histology slides of our 18-year-old patient's terminal ileum. A. Low power of terminal ileum mucosa. **B**. Granuloma and dense chronic active inflammation. **C**. Cryptitis and crypt abscess. **D**. Dense lymphoplasmacytic infiltrate in colonic mucosa**. E**. Mild gland budding and/or branching. *The histology slides are courtesy of Dr. Sangeeta Sandhu, assistant professor, McGill University, pathologist, Lakeshore General Hospital, Montreal, Quebec, Canada*.

III levels were negative or normal. Transesophageal echography, carotid Doppler and neck MRA were performed to rule out an embolic source were all unremarkable. An abdominal CT showed abdominal wall thickening at the ileocecal junction, consistent with an inflammatory etiology. In view of these findings, the patient was started on intravenous methylprednisolone (20 mg) q8hr as well as prednisolone acetate drops q1hr and dexamethasone 0.1% ointment qhs OD. Azathioprine was also started. Intravenous methylprednisolone was stopped after 72 h and replaced with daily oral prednisone (50 mg) and ASA (325 mg).

At one-month follow-up, the patient's right eye visual acuity improved to counting fingers. His visual acuity in the left eye was 20/50 improving to 20/25 with pinhole. The patient missed 3 color plates (out of 17) when testing his left eye with the Ishihara color vision test. There was a right RAPD with the left pupil also found to be slow to react to light. The dilated fundus exam of the right eye showed significant and diffuse optic nerve pallor. The previously reported retinal whitening was resolved (Fig. 3A). The dilated fundus exam of the left eye was significant for moderate temporal pallor of the optic nerve (Fig. 3 B). FA was unremarkable OU (Fig. 3C–F).

3. Discussion

The average age for patients with CRAO is 58.5 years.[3] The incidence of CRAO in patients under 30-year-old is about 1 in 50,000 patients.[3] These CRAO cases are generally associated with anticardiolipin antibodies, rheumatic heart disease, aortic or mitral regurgitation, systemic lupus erythematosus and homocystinuriabib3.[3,4] CRAO has also been reported in neoplasms such as T-cell lymphoma.[5] Although etiologically different, the visual prognosis of CRAO in very young adults are similar.[3]

In our case, the etiology of the arterial occlusion is likely inflammatory as evidenced by improvement in vision and disc leakage following anti-inflammatory treatment. In contrast with embolic arterial occlusion, which is characterized by transient acute occlusion until the emboli dislodges downstream, inflammatory arterial occlusions are characterized by persistent blockage especially when the underlying etiology is poorly treated. The

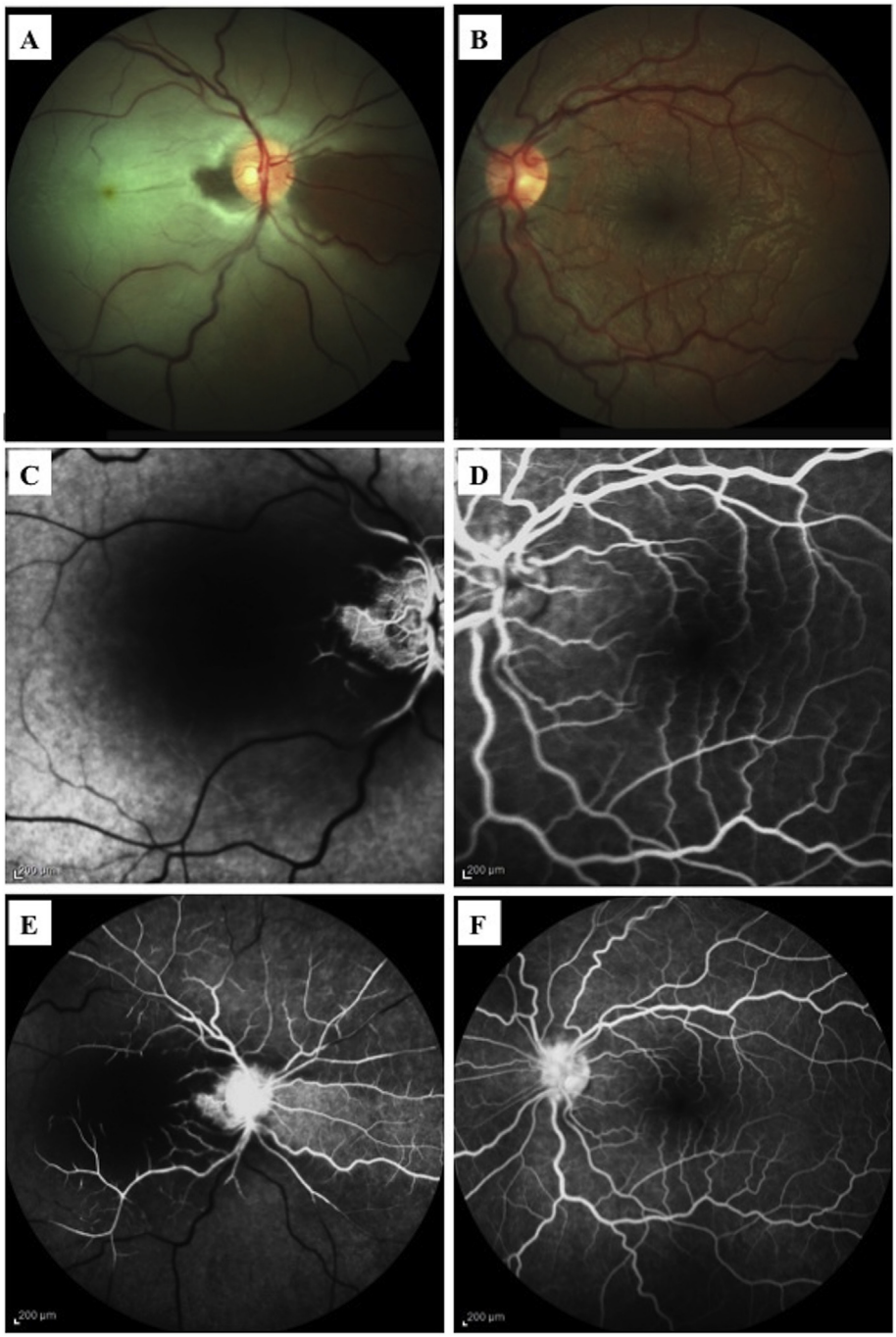

Fig. 2. Initial presentation of our 18-year-old patient with central retinal artery occlusion (CRAO) and optic nerve vasculitis in association with Crohn disease. A. OD fundus photo showing diffuse retinal pallor compatible with CRAO with juxtapapillary cilioretinal sparing exam. **B**. OS fundus photo showing a normal exam. **C**. OD early phase fluorescein angiography (FA) showing delayed arterial filling of major arcades with cilioretinal sparing. **D**. Normal OS early phase FA**. E**. OD mid phase FA showing delayed arterial filling of major arcades with cilioretinal sparing and optic nerve diffuse leakage. **F**. OS mid phase FA showing optic nerve patchy leakage.

occlusive vasculopathy in Crohn disease is believed to result from blood vessel wall disruption due to a combination of focal arteritic submucosal glaucomatous changes and chronic adherence of inflammatory cells.[6]

Ocular findings in Crohn disease, although uncommon, include conjunctivitis, uveitis, and scleritis. More visually threatening retinal vascular occlusion were previously reported in Crohn disease.[7,8] Particularly, Falavarjani et al. reported a case of unilateral CRAO with cilioretinal artery sparing and optic nerve atrophy in a 9-year-old boy[9,10]. Unilateral branch retinal artery occlusion (BRAO) and non-ischemic central retinal vein occlusion were also reported.[7–10] Interestingly, Saatci et al. reported a case of unilateral retinal vasculitis with a BRAO that required laser panretinal photocoagulation for neovascularization.[11] Optic nerve involvement including papillary inflammation, neuroretinitis and optic neuropathy, has been previously reported in association with Crohn disease.[12] Heuer et al. reported a case of bilateral ischemic optic neuropathy and retinal vasculitis in a 24-year-old patient.[13]

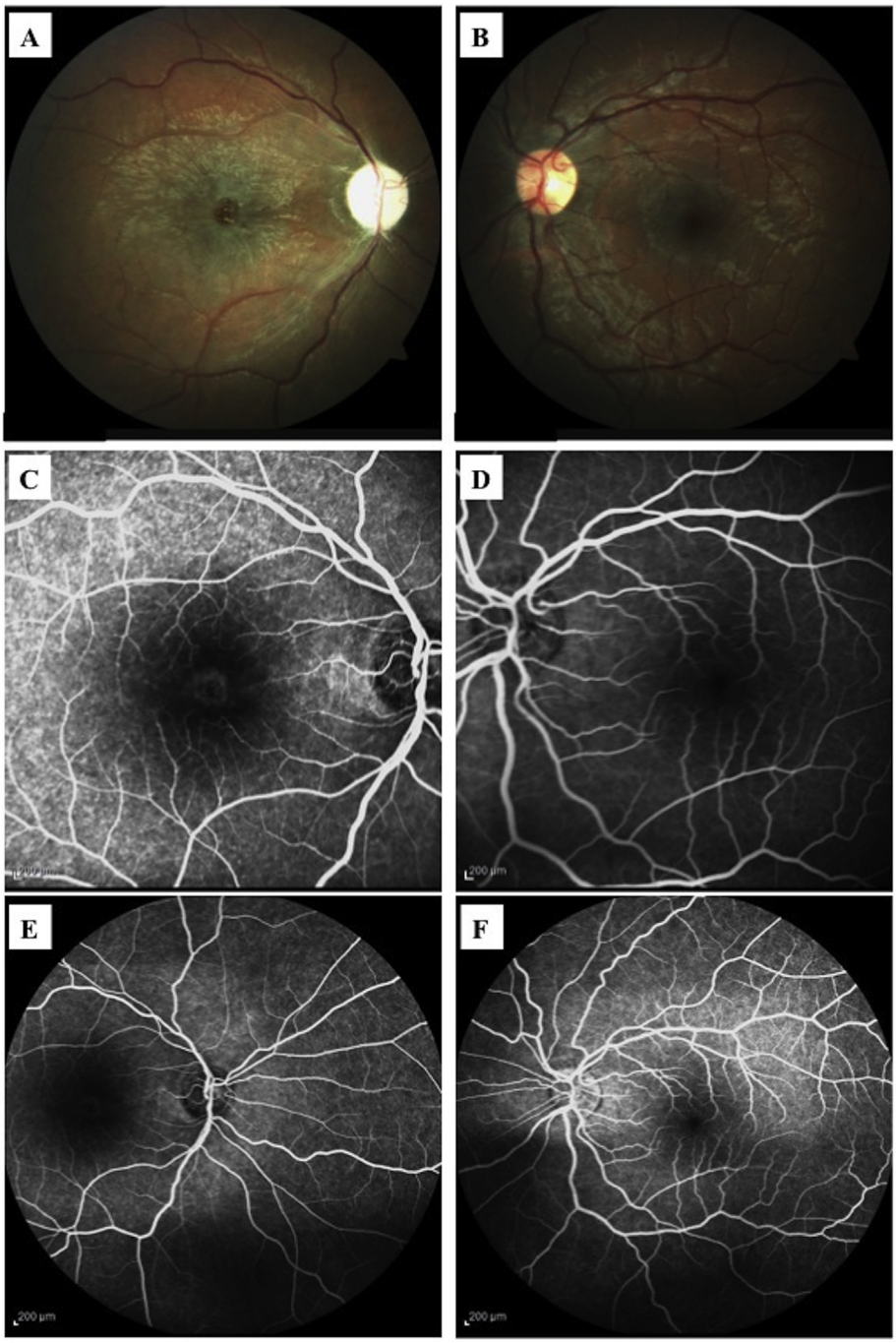

Fig. 3. Follow-up exam of our 18-year-old patient with central retinal artery occlusion (CRAO) and optic nerve vasculitis in association with Crohn disease. A. OD fundus photo showing marked diffuse optic nerve pallor. **B**. OS fundus photo showing moderate temporal optic nerve pallor. **C and E**. OD early and mid phases fluorescein angiography (FA) showing grossly normal vascular flow. **D and F**. OS early and mid phases FA showing grossly normal vascular flow.

To our knowledge, this is the first case of Crohn disease related bilateral optic nerve occlusive vasculitis presenting with CRAO in one eye and amaurosis fugax in the fellow eye.

In conclusion, CRAO in the pediatric and young adult patients is unusual. A systemic hypercoaguable, inflammatory and infectious etiologies must always be ruled out. Crohn disease should be considered as an etiology of retinal vascular occlusive disorders especially in young patients. Aggressive treatment with systemic steroids and immunosuppressive agents is needed during the acute presentation as well as for chronic maintenance to prevent and/or minimize flare-ups.

Patient consent

Consent to publish the case report was not obtained. This report does not contain any personal information that could lead to the identification of the patient."

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Conflict of interest

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Authorship

Each of the authors has contributed to, read and approved this manuscript. All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of interest

None of the authors has any conflict of interest, financial or otherwise.

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