Case Report

Central Retinal Vein Occlusion-like Appearance: A Precursor Stage in Evolution of Frosted Branch Angiitis

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Abstract

Purpose: To report a young man with a central retinal vein occlusion (CRVO)-like appearance which later evolved to frosted branch angiitis (FBA).

Case Report: A 28-year-old Indian man presented with optic disc swelling, hyperemia, peripapillary hemorrhages, and dilated tortuous veins in the left eye, 6 months after being diagnosed with idiopathic FBA in the right eye. Within 3 days of presentation, the left eye developed FBA, which was promptly and successfully treated with oral steroids.

Conclusion: A CRVO-like picture may be the first stage of FBA. Young patients with CRVO and intraocular inflammation should be followed closely for early detection of FBA. Early initiation of oral steroids may preserve visual acuity in such cases.

Keywords: Central Retinal Vein Occlusion; Optic Disc Hyperemia; Uveitis; Vasculitis

J Ophthalmic Vis Res 2017; 12 (4): 440-2

INTRODUCTION

Frosted branch angiitis (FBA) is a rare disorder which was first described by Ito et al in a 6-year-old Japanese boy with panuveitis and widespread retinal vasculitis.[1] Since the initial report in 1976, only a small number of cases have been reported in the literature and the majority of them are from Japan.[2] To the best of our knowledge, there are only about 9 reported cases of FBA associated with central retinal vein occlusion (CRVO). [3-6]

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Received: 26-08-2015 Accepted: 28-01-2016

Access this article online

Quick Response Code:

Website: www.jovr.org

DOI: 10.4103/jovr.jovr_84_15

We found no previous reports elaborating any preclinical stage prior to development of clinically detectable FBA. In view of the rarity of the disease, very little is known about the natural history of FBA with CRVO. Here, we report a case of idiopathic FBA with CRVO, which might be helpful in understanding the natural course of the disease.

CASE REPORT

A 28-year-old otherwise healthy Indian man presented with mild nonspecific discomfort in the left eye. He had been diagnosed with idiopathic FBA in the right eye 6 months earlier [Figure 1a], at that time a complete blood count, differential count, erythrocyte sedimentation rate,

How to cite this article: Kumawat B, Tripathy K, Venkatesh P, Chawla R, Sharma YR. Central retinal vein occlusion-like appearance: A precursor stage in evolution of frosted branch angiitis. J Ophthalmic Vis Res 2017;12:440-2.

chest X-ray, Mantoux test, and angiotensin-converting enzyme level were normal. Serology for human immunodeficiency virus and syphilis was non-reactive. On presentation, the right eye had achieved visual acuity of 6/12 (20/40, logMAR 0.30) after resolution of the previous episode of FBA. The left eye had 6/6 (20/20, logMAR 0) vision. Intraocular pressure on applanation tonometry was 14 and 12 mmHg in the right and left eyes, respectively. There were occasional anterior chamber cells in both eyes with no keratic precipitates. The left eye showed marked optic disc hyperemia, mild blurring of the disc margins, and dilated tortuous veins with flame-shaped hemorrhages around the disc [Figure 1b]. Fundus fluorescein angiography showed normal arterial, arteriovenous, and venous phases in both eyes. There was mild early disc leak in the left eye, which increased with time [Figure 1c]. There was no abnormal perivascular leakage to suggest vasculitis in either eye. A diagnosis of impending CRVO in the left eye was made and the patient was further evaluated to rule out disorders associated with CRVO in younger individuals. The cardiovascular evaluation was unremarkable. A complete hemogram, with bleeding time, clotting time, prothrombin time, activated partial thromboplastin time, and serum homocysteine level, were within normal limits. Serum antinuclear antibody and lupus anticoagulant were negative. Three days later, the patient presented with decreased visual acuity of 6/18 (20/60, logMAR 0.48) in the left eye. The left eye showed increased disc edema with hyperemia, macular edema, and thick perivascular exudation along the major vessels (predominantly venous) in the lower half of the fundus, typical of FBA [Figure 2a]. Differential leucocyte count, erythrocyte sedimentation rate, C-reactive protein, chest X-ray, Mantoux test, peripheral blood smear, and angiotensin-converting enzyme level were normal. Serology for human immunodeficiency virus was negative. The anterior segment of the left eye showed cells with no keratic precipitates. The patient was started on oral prednisolone 1 mg/kg/day. Four weeks later, macular edema had resolved completely, with a few precipitated hard exudates nasal to the fovea. Small segments of patchy vascular sheathing were evident along the vessel wall of inferior arcade arterioles [Figure 2b]. There was complete visual recovery to 6/6 (20/20, logMAR 0) in the left eye. Oral steroids were tapered off in the following 4 weeks. No recurrence of the disease was noted in either eye during 6 months of follow-up.

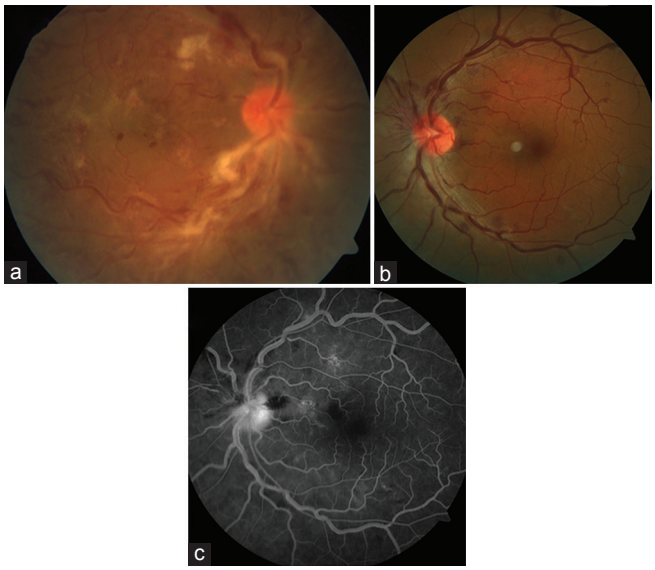

Figure 1. (a) The right eye in this patient has dilated tortuous veins, paravascular infiltrates surrounding both veins, and arteries suggestive of frosted branch angiitis 6 months before involvement of the left eye. (b) At presentation, the left eye shows marked disc hyperemia, mild blurring of the disc margins, and dilated and tortuous veins with flame-shaped hemorrhages around the disc. (c) A fluorescein angiogram showing mild disc leak and normal arteriovenous transit time, with no abnormal vascular leakage in the left eye.

DISCUSSION

Typical idiopathic FBA has been described as a primary immune response to a number of provoking antigens and is not associated with pre-existing intraocular inflammation.[2] The exact nature of these antigens is unknown but may include herpes simplex virus, varicella zoster virus, Epstein-Barr virus, cytomegalovirus (CMV), measles, rubella, adenovirus, Coxsackie virus, tuberculoprotein, and antistreptolysin.[2] Idiopathic FBA is treated by systemic steroids. Subcutaneous adalimumab has been successfully used in a patient with recurrent idiopathic FBA and a history of an acute steroid-induced episode of psychosis during a previous attack of FBA.[7] Atypical FBA secondary to concurrent inflammation is most

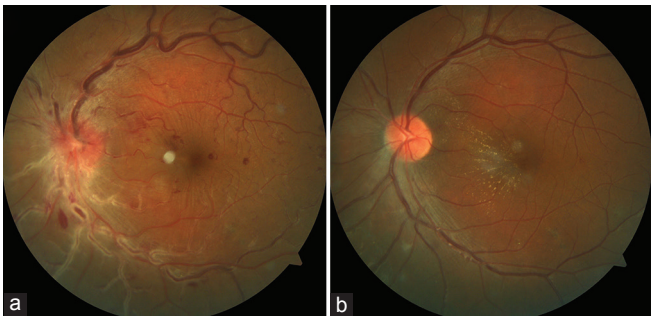

Figure 2. (a) Three days after the initial presentation, the same eye shows increased disc edema with hyperemia, macular edema, and thick perivascular exudation along the major vessels (predominantly venous), typical of frosted branch angiitis. (b) Four weeks after starting oral steroids, the macular edema resolved completely, with precipitated hard exudates on the nasal side of the fovea with patchy vascular sheathing in the left eye.

commonly seen in association with CMV retinitis.[2] Another form of FBA that may be infiltrative rather than an inflammatory process is associated with ocular lymphoma or leukemia.[2] FBA has been reported in association with CRVO in about nine patients in the past.[3-6] Six of these nine patients were in the third or fourth decade of life. The exact causal relationship between FBA and CRVO has not been established. In most of the reported cases, involvement was unilateral and FBA was documented together with CRVO. Abu El-Asrar et al[6] reported a patient who developed non-perfused CRVO after development of FBA in the right eye. Later, the patient was diagnosed with CRVO in other (left) eye which, 10 days later, developed FBA. Unfortunately both eyes were lost due to subsequent neovascular glaucoma. Clinical presentation of this case has some similarities with our patient.

This patient had a high IgM titer of antiphospholipid antibody and atheromatous plaques in both common carotid arteries, in the origin of the right internal carotid artery, and in the left internal carotid artery. These concurrent findings suggest that, in this case, the development of CRVO like findings cannot be attributed to the FBA alone. Our patient was similarly diagnosed as having FBA in the right eye and subsequently presented with a CRVO-like picture in the left eye before development of FBA. Within 3 days, the patient developed typical FBA in the left eye as well. Fortunately, our patient responded well to steroids. We hypothesize that in cases presenting with CRVO before the FBA appearance, the main trunk of central retinal vein first becomes involved where it runs through the optic nerve resulting in a CRVO-like appearance. Perivascular inflammation then spreads centrifugally over the next few days, and FBA is established. However, histopathological data and larger case series of similar cases are required to validate this hypothesis. In our patient, prompt treatment with oral steroids led to resolution of this CRVO-like appearance with no sequelae of venous occlusion, which suggests that immediate initiation of anti-inflammatory therapy might be useful to prevent blinding sequelae of venous occlusion like neovascular glaucoma.

Our experience with this patient suggests that a CRVO-like appearance of the fundus may be part of the natural history of FBA. Since FBA itself is painless and visual acuity may not be affected initially, the patient may not present to a clinician at this very first stage of the condition. Young patients with a CRVO-like picture may progress to FBA. Therefore, a high degree of clinical suspicion and early follow-up is required for young patients who present with a CRVO-like picture and signs of anterior or posterior segment inflammation for early detection and timely management of FBA.

Financial Support and Sponsorship

Nil.

Conflicts of Interest

There are no conflicts of interest.

REFERENCES

1. Ito Y, Nakano M, Kyu N, Takeuchi M. Frosted-branch angiitis in a child. *Rinsho Ganka Jpn J Clin Ophthalmol* 1976;30:797–803.
2. Walker S, Iguchi A, Jones NP. Frosted branch angiitis: A review. *Eye Lond Engl* 2004;18:527–533.
3. Foss AJ, Headon MP, Hamilton AM, Lightman S. Transient vessel wall sheathing in acute retinal vein occlusions. *Eye Lond Engl* 1992;6(Pt 3):313–316.
4. Seo MS, Woo JM, Jeong SK, Park YG. Recurrent unilateral frosted branch angiitis. *Jpn J Ophthalmol* 1998;42:56–59.
5. Kaburaki T, Nakamura M, Nagasawa K, Nagahara M, Joko S, Fujino Y. Two cases of frosted branch angiitis with central retinal vein occlusion. *Jpn J Ophthalmol* 2001;45:628–633.
6. Abu El-Asrar AM, Al-Obeidan SA, Abdel Gader AGM. Retinal periphlebitis resembling frosted branch angiitis with nonperfused central retinal vein occlusion. *Eur J Ophthalmol* 2003;13:807–812.
7. Hedayatfar A, Soheilian M. Adalimumab for treatment of idiopathic frosted branch angiitis: A case report. *J Ophthalmic Vis Res* 2013;8:372–375.