

Neuroretinitis, frosted branch angiitis, and paracentral acute middle maculopathy in a young female

Simar Rajan Singh, Mohit Dogra, Faisal Thattaruthody, Ramandeep Singh, Mangat R Dogra

Key words: Frosted branch angiitis, neuroretinitis, PAMM, syphilis

A 23-year-old woman presented to the ophthalmologist with sudden, painless decrease of vision in her right eye (RE) since 4 days. Ophthalmic examination revealed a visual acuity of counting fingers at 1 m in the RE and 20/20 in the left eye. There was a presence of relative afferent pupillary defect with 2+ cellular reaction in the anterior chamber of the RE. Posterior segment examination revealed vitritis, optic disc edema, hard exudates, and subretinal fluid in the posterior pole. There was a wedge-shaped patch of retinal opacification temporal to the fovea along with exuberant perivascular exudation of the supero-nasal and supero-temporal retinal arterioles, suggestive of frosted branch angiitis (FBA) [Fig. 1a]. Ultrawide field fluorescein angiography showed leakage of the optic

disc, capillary non-perfusion areas in the superior half of the retina [Fig. 1b]. The left eye was normal. Optical coherence tomography passing through the fovea showed the presence of subretinal fluid and cystoid macular edema along with hyper-reflectivity of the temporal inner nuclear layer [Fig. 1c]. A diagnosis of right eye panuveitis with

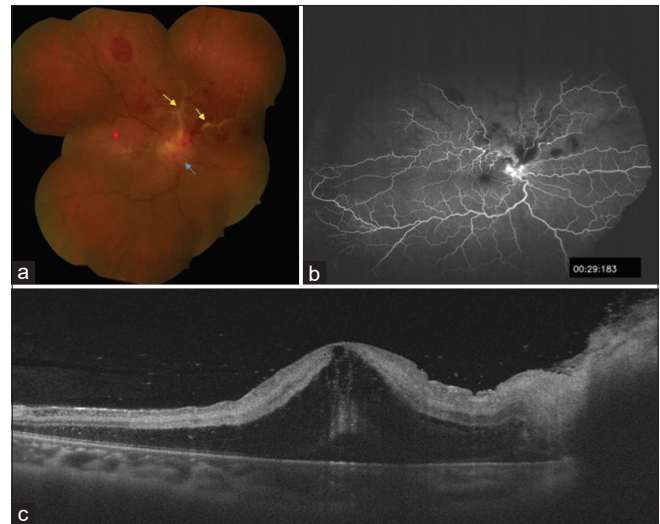


Figure 1: Neuroretinitis with Frosted branch angiitis in a young female. (a) Fundus picture of the right eye (RE) showing optic disc edema (blue arrow), wedge-shaped opacification of the retina temporal to the fovea (red arrow) along with sheathing of the superior and nasal retinal arterioles (yellow arrows) suggestive of frosted branch angiitis. (b) Late phase ultrawide field fluorescein angiogram of the RE showing non-perfusion in the area of the vasculitis with leakage over the disc. (c) Optical coherence tomography demonstrating paracentral acute middle maculopathy

Access this article online	
Quick Response Code:	Website: www.ijo.in
	DOI: 10.4103/ijo.IJO_10_20

Advanced Eye Centre, Department of Ophthalmology, Post Graduate Institute of Medical Education and Research, Chandigarh, India

Correspondence to: Dr. Mohit Dogra, Advanced Eye Centre, Post Graduate Institute of Medical Education and Research, Chandigarh - 160 012, India. E-mail: mohit_dogra_29@hotmail.com

Received: 07-Jan-2020

Revision: 18-Mar-2020

Accepted: 21-Mar-2020

Published: 20-Aug-2020

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Cite this article as: Singh SR, Dogra M, Thattaruthody F, Singh R, Dogra MR. Neuroretinitis, frosted branch angiitis, and paracentral acute middle maculopathy in a young female. Indian J Ophthalmol 2020;68:1962-3.

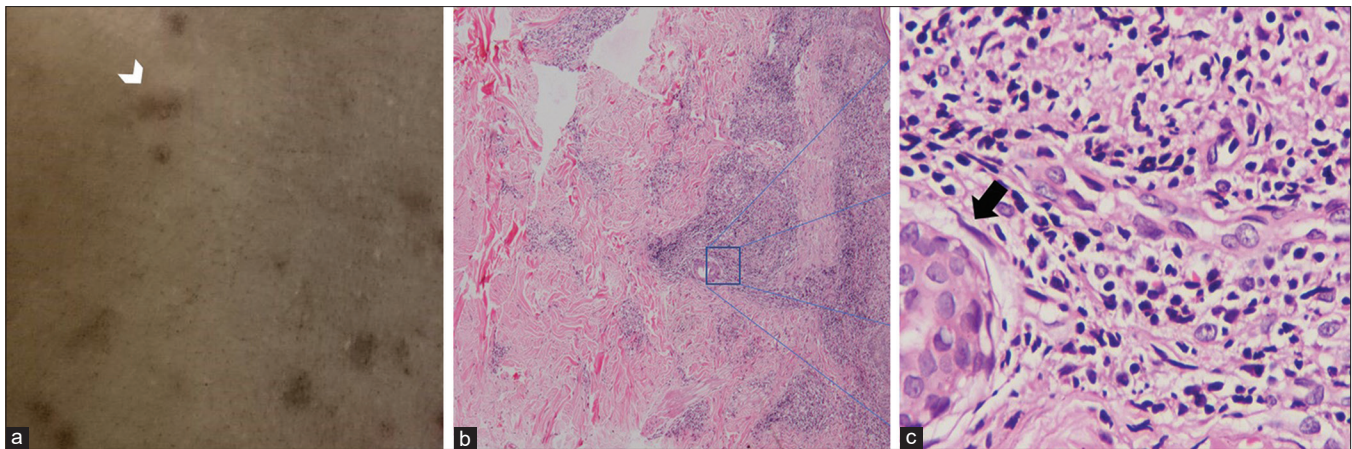


Figure 2: Skin lesions with biopsy findings characteristic of syphilis. (a) Multiple reddish-pink, non-itchy lesions over the back (white arrowhead). (b) Biopsy from the skin lesions showing perivascular distribution of inflammatory cells, with predominance of plasma cells and histiocytes [hematoxylin and eosin, magnification 100×]. (c) High magnification image from the marked area showing infiltration and thickening of the vessel wall (black arrowhead) [hematoxylin and eosin, magnification 400×]

neuroretinitis, paracentral acute middle maculopathy, and frosted branch angiitis was made. A review of systems revealed reddish-pink maculopapular rash over the trunk and extremities, including the palms and soles [Fig. 2a]. Biopsy from the skin lesions revealed the presence of inflammatory cells consisting mainly of plasma cells and histiocytes in a perivascular distribution [Fig. 2b] along with infiltration and thickening of the vessel wall [Fig. 2c]. The dermatological findings were highly suggestive of syphilis.^[1]

Discussion

Syphilis, also known as “*the great mimicker*,” can affect almost every structure of the eye.^[2] Although less common, FBA form of vasculitis can be seen as a manifestation of syphilis.^[3] The diagnostic feature of this case was the skin rash. A high index of suspicion is required to diagnose ocular syphilis, which is managed like neurosyphilis.^[4,5] Our patient was treated with 2 g of intravenous ceftriaxone twice a day for 14 days, due to non-availability of aqueous Penicillin G. Prophylactic scatter laser photocoagulation of the non-perfused retina was done to prevent retinal and anterior segment neovascularization. Oral steroids (0.5 mg/kg body weight) were added after 72 h of starting systemic antibiotics in order to reduce the inflammatory insult to the optic nerve and retina. At 9 months follow-up, the visual acuity improved to counting fingers at 3 m with complete resolution of optic disc edema and vasculitis. Recovery of visual acuity was modest as the patient developed optic disc pallor and foveal atrophy.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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