Central retinal artery occlusion in a child with angiolymphoid hyperplasia with eosinophilia orbit

V S Vijitha, Anasua Ganguly Kapoor, Ruchi Mittal[1], Aditya Kapoor[2]

Key words: Angiolymphoid hyperplasia of orbit with eosinophilia, ocular oncology, pediatric proptosis

An 8-year-old-boy presented with rapid onset proptosis with vision loss of right eye following blunt trauma. Examination showed right abaxial, nontender proptosis [Fig. 1a and b] with inferior dystopia with limitation of motility in all directions of gaze, conjunctival corkscrew vessels, and minimal congestion. Visual acuity in the right eye was hand movements with afferent pupillary defect. Fundus showed features of central retinal artery occlusion [Fig. 1c]. B Scan orbit showed hypoechoic lesion posterior to globe causing conical deformation of posterior globe (guitar-pick sign) indicative of high orbital pressure[1] [Fig. 1d]. Left eye was normal. Computed tomography (CT) orbit showed homogenous isodense mass with irregular margins in the right superior orbit extending to intraconal space till the apex, indenting the globe [Fig. 1e and f]. Prompt ocular massage and intravenous mannitol were administered. Immediate paracentesis and orbitotomy with debulking of the ill-defined, soft, nonencapsulated mass was performed. Histopathology revealed fibromuscular and fibrofatty tissue diffusely replaced by proliferating vessels of varying sizes lined by plump epithelioid-like endothelial cells with pale cytoplasm amidst numerous eosinophils with plasma cells and lymphocytes in a fibrous stroma [Fig. 1g; H and E 40X]. CD31 decorated the endothelial cells [Fig. 1h, 20X] confirming the diagnosis of angiolymphoid hyperplasia with eosinophilia of orbit. Peripheral blood eosinophilia was present. The child was started on oral steroids and proptosis improved significantly. However, there was no improvement in vision.

Access this article online

Quick Response Code:

Website: www.ijo.in

DOI: 10.4103/ijo.IJO_1422_19

Ophthalmic Plastic Surgery and Ocular Oncology Services, LV Prasad Eye Institute, [2]Vitreo Retina Services, LV Prasad Eye Institute, Vijayawada, Andhra Pradesh, [1]Kanupriya Dalmia Ophthalmic Pathology Laboratory, LV Prasad Eye Institute, Bhubaneswar, Odisha, India

Correspondence to: Dr. Anasua Ganguly Kapoor, The Operation Eyesight Universal Institute for Eye Cancer, Consultant Ophthalmic Plastic Surgery and Ocular Oncology Services, LV Prasad Eye Institute, Vijayawada, Andhra Pradesh, India. E-mail: anasua21@yahoo.com

Received: 03-Aug-2019 Revision: 03-Oct-2019
Accepted: 24-Oct-2019 Published: 22-Nov-2019

Cite this article as: Vijitha VS, Ganguly Kapoor A, Mittal R, Kapoor A. Central retinal artery occlusion in a child with angiolymphoid hyperplasia with eosinophilia orbit. Indian J Ophthalmol 2019;67:2059-60.

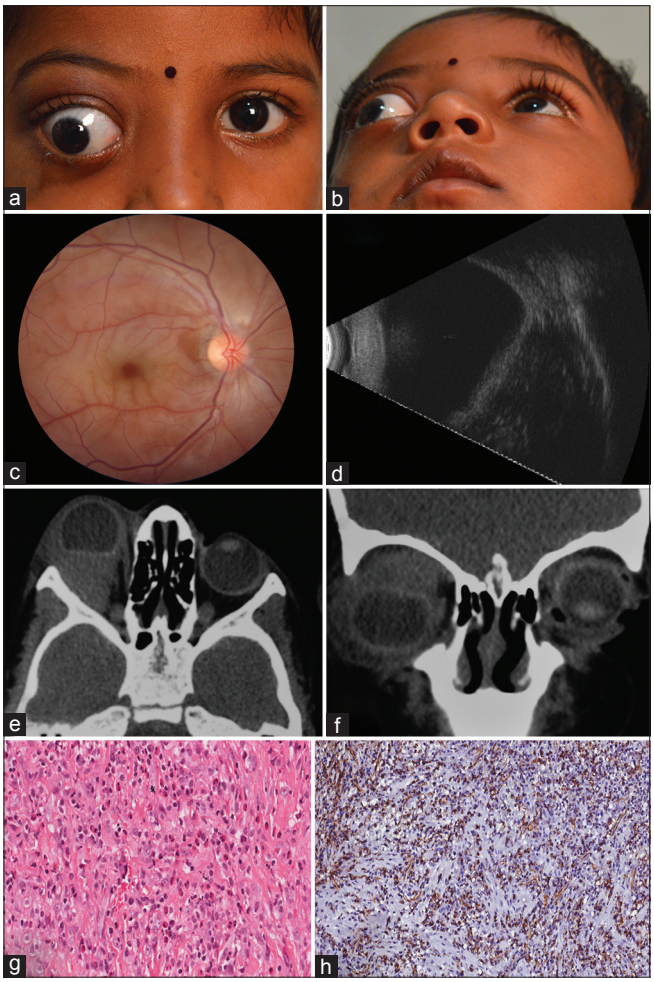

Figure 1: (a) Clinical image showing inferolateral dystopia; (b) Worms view showing abaxial proptosis; (c) Fundus photograph of the right eye showing diffuse whitening of the posterior pole and a cherry red spot; (d) Ultrasonography of the orbit showing hypoechoic lesion posterior to the globe with few low to moderate reflective dot echoes; (e and f) Axial and coronal sections of CT scan showing a well-defined homogenously isodense mass causing significant proptosis and indentation on the globe; (g) Photomicrograph showing vessels of varying caliber lined by plump endothelial cells with pale cytoplasm amidst numerous eosinophils and few lymphocytes in a fibrous stroma. (Hematoxylin and Eosin 40X); (h) The endothelial cells are highlighted by CD31 (20X)

Angiolymphoid hyperplasia with eosinophilia is a benign disorder usually presenting as subcutaneous nodules in the head and neck region.[2] Few cases of orbital involvement have been reported in literature.[2,3] Middle-aged women are commonly affected and occasional reports in pediatric age group exist. Response to steroids is usually good but recurrences are known to occur. Surgery is limited to debulking due to the diffuse nature of the tumor. Immunosuppressants are indicated for recurrent or refractory cases unresponsive to steroids.[3] Rare instance of spontaneous resolution in an immunosuppressed patient has been described.[4] Clinicians need to be aware of this entity, which infrequently affects children, in order to aid in early diagnosis and appropriate treatment.

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.

References

1. Theoret J, Sanz GE, Matero D, Guth T, Erickson C, Liao MM, *et al*. The "guitar pick" sign: A novel sign of retrobulbar hemorrhage. CJEM 2011;13:162-4.
2. Seregard S. Angiolymphoid hyperplasia should not confused with Kimura's disease. Acta Ophthalmol Scand 2001;79:91-3.
3. Mukherjee B, Kadaskar J, Priyadarshini O, Krishnakumar S, Biswas J. Angiolymphoid hyperplasia of the orbit and adnexa. Ocul Oncol Pathol 2016;2:40-7.
4. Kumari P, Kasturi N, Nagarajan G, Senthamizh T, Ramesh BK, Bheemanathi HS. Spontaneous regression of angiolymphoid hyperplasia with eosinophilia of lacrimal gland in an HIV-positive patient. Indian J Ophthalmol 2019;67:1334-5.