Recurrent posterior scleritis with secondary choroidal osteoma in a child

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Key words: Choroidal osteoma, immunoglobulin G4 (IgG4) disease, posterior scleritis

Posterior scleritis common in adults is characterized by disc swelling, retinal striae and T-sign on ultrasound. Pediatric posterior scleritis is a rare entity. Choroidal osteoma (CO) may be secondary to inflammatory etiologies like idiopathic orbital inflammation and sclero-uveitis.

A 12-year old boy, presented with intermittent dull aching pain and decreased vision in the right eye (RE) for 3 years and was on systemic steroid therapy. Fundus photographs taken 6 months back revealed an elevated choroidal lesion superior, nasal and temporal to the optic disc [Fig. 1a] with hyperfluorescence and delayed staining on the corresponding fluorescein angiogram (FA) [Fig. 1b and c] and T-sign on B-scan ultrasound (USG) [Fig. 2].

We noted best-corrected visual acuity of 20/30 in RE and a nontender periorbital fullness superolaterally. The eye was quiet. The juxtapapillary choroidal lesion had decreased in size, with well-defined borders [Fig. 3a]. There was normal autofluorescence [Fig. 3b].

On FA, hyperfluorescence had resolved [Fig. 3c]. Indocyanine green angiogram showed hypocyanescence [Fig. 3d]. There was no T-sign on USG, but a high-reflective spike adjacent to the optic disc suggestive of calcification was seen [Fig. 4]. Optical coherence tomography showed a thick hyper-reflective layered matrix [Fig. 5]. Serum IgG4 was raised (3.12 g/L reference 0.012–1.699). Mantoux, QuantiFERON TB-gold, RA, ANA, c and p ANCA were negative. The left eye was normal. Pediatric evaluation was normal. Oral steroids were tapered and regular ophthalmic and pediatric follow-up was advised.

Discussion

A diagnosis of recurrent posterior scleritis and "possible" IgG4-related ophthalmic disease was considered based on recommended criteria.[1] The choroidal osteoma in our case appears to be secondary to inflammation-induced dystrophic ossification. Systemic surveillance was recommended as IgG4-related multifocal fibrosclerosis, pseudotumor, retroperitoneal fibrosis, autoimmune pancreatitis, and thyroiditis have been observed.[2,3]

IgG4-related ocular inflammation is characterized by IgG4 positive plasmacytic infiltration, fibrosis of the involved organs, and elevated serum IgG4 levels. Histopathology from tissue confirms the diagnosis.[1] While oral steroids are the first-line treatment in IgG4 disease, immunomodulators and Rituximab could be considered for recurrences.[4]

IgG4-related disease is an emerging diagnosis to consider in recurrent ocular inflammation. Early diagnosis prevents extensive investigations and multiple biopsies. The unique association of two lesser known entities like secondary choroidal osteoma and IgG4 inflammatory disease with recurrent posterior scleritis is highlighted in this report.

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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

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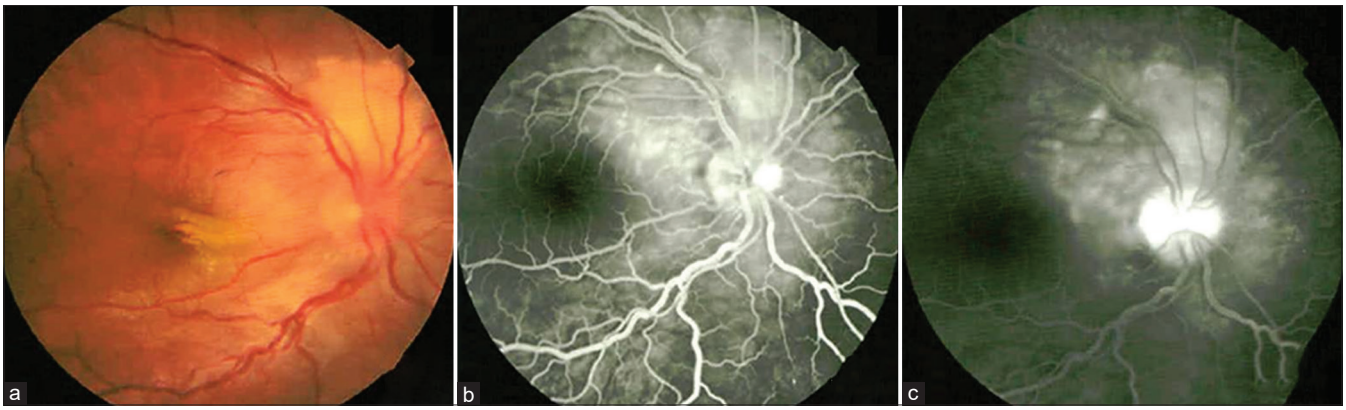

Figure 1: (a) Color fundus photo of right eye taken 6 months prior to presentation to us showing exudative retinal detachment and choroidal folds suggestive of posterior scleritis; (b) Corresponding fluorescein angiogram (FA) showing hyperfluorescence in early phase; (c) FA showing staining in the late phase

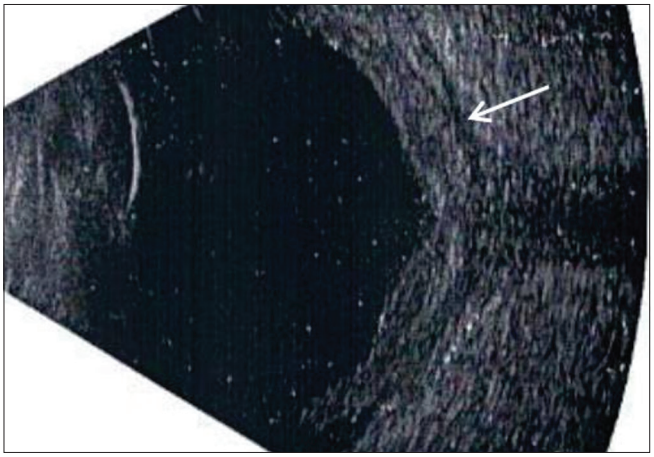

Figure 2: Ultrasound B scan taken 6 months prior to presentation to us showing subtenons fluid (T-sign) (white arrow) suggestive of posterior scleritis

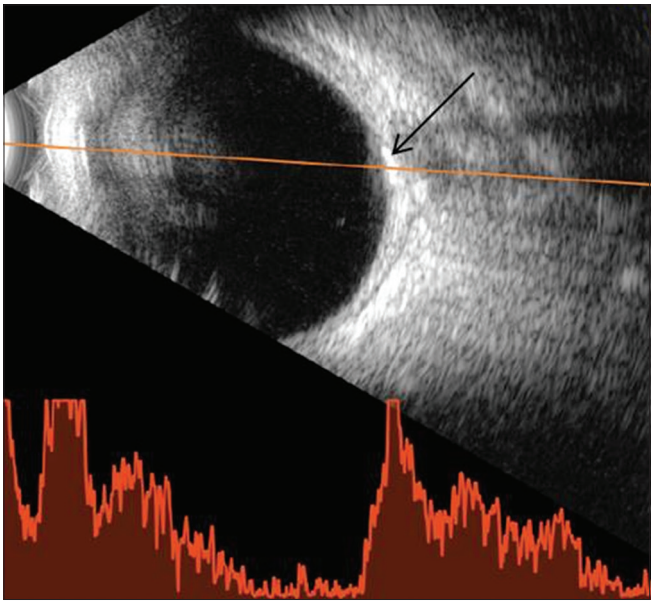

Figure 4: Ultrasound B scan taken on presentation to us showing a linear high reflective lesion noted over the ocular coats adjacent to ONH (black arrow) causing back shadowing persisting in low gain suggestive of calcification and/or choroidal osteoma with the absence of T sign

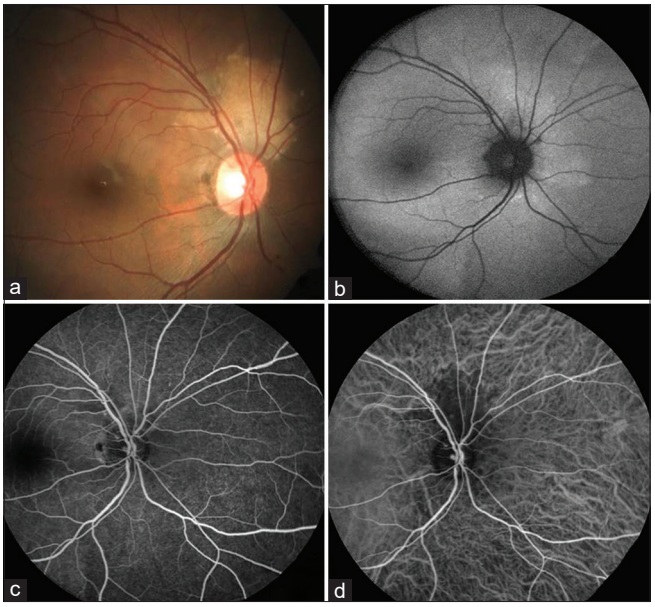

Figure 3: (a) Colour fundus photograph of right eye taken on presentation to us showing a juxtapapillary orange yellow choroidal lesion (decreased size) superior to disc; (b) Fundus Autoflourescence (FAF) showing normal autofluorescence; (c) FA on presentation to us showing resolution of hyper fluorescence and no disc leak; (d) Indocyanine Green Angiography (ICG) showing corresponding hypocyanascence

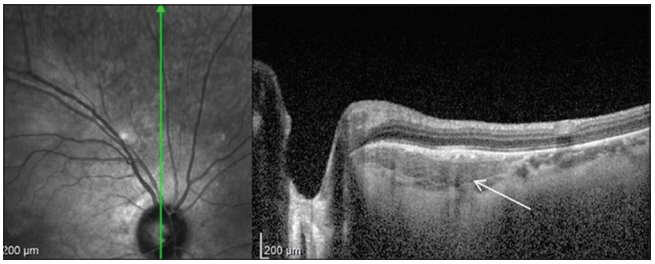

Figure 5: Spectralis OCT through the choroidal lesion showing a thick hyper-reflective layered matrix within hypo-reflective echoes (white arrow) and increased choroidal thickness superior to disc

and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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