Papillophlebitis as an initial presentation of Eales’ disease

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Abstract:

Eales’ disease (ED), which is an idiopathic obliterator vasculitis, is a diagnosis of exclusion. The optic nerve involvement in ED is not very common. We report a case of ED in a 36-year-old male who initially presented as papillephlebitis. He presented with complaints of decreased vision in his right eye for 2 months. Fundus examination revealed that optic disc edema with hemorrhages, and he was started on oral corticosteroid after the exclusion of infectious etiology. Fundus examination after 2 months revealed that resolution of optic disc edema, but active periphlebitis with multiple superficial retinal hemorrhages involving inferior and inferotemporal quadrant. Based on negative laboratory results and clinical findings a diagnosis of ED was considered. Regular monitoring of patients with papillephlebitis is recommended.

Keywords:

Corticosteroid, Eales’ disease, optic disc edema, papillephlebitis, retinal vasculitis

Introduction

Eales’ disease (ED) is an idiopathic obliterator vasculitis that primarily affects the peripheral retina.[1,2] ED primarily affects young adults and is characterized by three overlapping stages of periphlebitis, venous occlusion, and retinal neovascularization. The diagnosis of ED is mostly clinical and requires exclusion of other systemic or ocular conditions that could present with similar clinical pictures. The most common presentation of ED is a sudden visual loss secondary to vitreous hemorrhage and vasculitis. We report a case of a 36-year-old male who initially presented with papillephlebitis and subsequently developed peripheral retinal vasculitis.

Case Report

A 36-year-old nondiabetic, nonhypertensive healthy Indian male presented to us with complaints of decreased vision in the right eye for the past 2 months. He initially presented to an ophthalmologist with a swollen optic disc, tortuous, and engorged retinal vessels along with scattered, splinter hemorrhages extending from the disc [Figure 1]. Fundus examination of the left eye was within the normal limits. Subsequent investigations revealed an erythrocyte sedimentation rate (ESR) of 17 mm/h (Westergren), a negative Mantoux test, and a normal high-resolution computerized tomography of the chest. His serum angiotensin-converting enzyme and lysozyme were within the normal limits. Based on interferon-gamma release assay, which was positive, the treating ophthalmologist decided to start him on oral corticosteroid and antitubercular drugs (ATT).

At the presentation to our clinic after 2 months, his best-corrected visual acuity was 6/9 in the right eye and 6/6 in the left eye. The slit-lamp examination of the anterior segment was normal in both eyes. Pupils were normally reacting and his color vision was within the normal limits. Fundus examination of the right eye revealed optic disc edema with hemorrhages involving the inferotemporal quadrant. Based on negative laboratory results and clinical findings a diagnosis of ED was considered. Regular monitoring of patients with papillephlebitis is recommended.

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eye revealed remarkable resolution of disc edema and peripapillary lesion, but showed active periphlebitis with multiple superficial retinal hemorrhages involving the inferior and inferotemporal quadrant [Figure 2a]. Fundus examination of the left eye was unremarkable. Fundus fluorescence angiography confirmed the presence of retinal periphlebitis and capillary nonperfusion areas in right eye [Figure 3]. Clinically, there was no evidence of macular edema and the patient refused to undergo swept-source optical coherence tomography due to financial constraint. He was thoroughly investigated, which revealed negative antinuclear antibody and negative antineutrophilic cytoplasmic antibodies and negative serologies for syphilis, human immunodeficiency virus, and toxoplasma. Based on clinical, angiographic, and laboratory investigations, the diagnosis of ED was performed. The patient was started on tablet mycophenolate mofetil 1000 mg two times daily, tablet prednisolone 60 mg/day, and was advised to continue ongoing ATT for 6 months and stop.

He was seen again after 1 month. Slit-lamp examination of the right eye revealed a quiet anterior chamber, occasional cells in the anterior vitreous. Fundus examination of the right eye revealed resolving periphlebitis in the inferior and inferotemporal quadrant [Figure 2b] with no evidence of any neovascularization clinically and angiographically. He was advised to continue the same treatment.

**Discussion**

ED can cause a diagnostic and therapeutic challenge to the ophthalmologist. ED has a vast spectrum of clinical presentations and though it has been primarily labeled as idiopathic obliterative vasculopathy and considered as a diagnosis of exclusion, various etiologies have been implicated with ED.\(^3\) The disease is classically described as bilateral, but asymmetric involvement is not uncommon. Posterior pole involvement is relatively uncommon in ED; even rarer is the optic disc involvement as the initial presentation. The largest case series on ED was reported from our center and included 898 eyes of 500 patients with ED; however, we did not find papilloephlebitis as an initial manifestation of ED.\(^4\) Macular involvement is rare but has been reported in extensive variety of the disease and in the late stages of proliferation.\(^5\) An aggressive clinical phenotype of ED, similar to central retinal vein occlusion was labeled as central ED by various authors.\(^6\) Saxena and Kumar\(^6\) described eleven consecutive cases of central ED. All of them were male and had raised ESR of \(>20\) mm/h. Majority of them were unilateral (81.8\%) and had good visual outcomes following treatment with systemic steroid. In a retrospective analysis of 565 eyes of 466 patients with ED, Atmaca e\(t\) al.\(^7\) reported central ED in 1.8\% of the patients. However, the presentation of our case was unique – the peripheral phlebitis was preceded by papillephlebitis. To the best of our knowledge, such a presentation of ED was not reported.

\[\text{Figure 1: Fundus photograph of the right eye showing a swollen disc with blurred margin, tortuous and engorged retinal vessels along with scattered splinter hemorrhages extending from the disc}\]

\[\text{Figure 2: (a) Fundus photograph of the right eye showing active periphlebitis with multiple superficial retinal hemorrhages involving inferior and inferotemporal quadrant (b) Fundus examination of the right eye showing resolving periphlebitis in inferior and inferotemporal quadrant after 1 month}\]

\[\text{Figure 3: (A to D) Fundus fluorescein angiography revealing retinal periphlebitis and capillary nonperfusion areas in the right eye}\]
in literature till date. To conclude papillephlebitis can be the initial presentation of ED and should be monitored closely to observe the further course of the new vascular inflammation which can involve the periphery of the retina.

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.

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Conflicts of interest
There are no conflicts of interest.

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