Saudi Journal of Ophthalmology (2015) 29, 160–164

Case Report

Unusual presentation of a multiple sclerosis case involving central retinal artery occlusion

Alberto Galvez-Ruiz [a,c,*], Sawsan R. Nowilaty [b]

Abstract

The term intermediate uveitis (IU) refers to a subgroup of uveitis in which the vitreous is the site of greatest inflammation. Patients with multiple sclerosis (MS) have a greater frequency of IU compared with the general population.
The IU associated with MS is characterized by the presence of pars planitis (occasionally accompanied by anterior uveitis) and the presence of peripheral retinal vasculitis in the form of periphlebitis (venous sheathing) in 6–26% of patients.
We present a patient with an unusual initial presentation of MS involving central retinal artery occlusion (CRAO) in the right eye (RE).
Although retinal vascular changes are asymptomatic in the majority of MS patients, the spectrum of impairment ranges from simple peripheral retina periphlebitis to the presence of peripheral occlusive retinal vasculitis in 6.5% of patients.
This atypical case may represent an extreme of the spectrum of retinal vasculitis associated with demyelinating disease.

Keywords: Central retinal artery occlusion, Retinal vasculitis, Multiple sclerosis, Intermediate uveitis, Periphlebitis

http://dx.doi.org/10.1016/j.sjopt.2014.12.001

Introduction

The term intermediate uveitis (IU) refers to a subgroup of uveitis in which the vitreous is the site of greatest inflammation. When IU is accompanied by snowbanks or snowballs, the condition is referred to as pars planitis.[1]

Patients with IU may present with inflammation in the anterior chamber (which is generally minimal) and peripheral vascular sheathing.[1,2]

Patients with multiple sclerosis (MS) have a greater frequency of IU compared with the general population. According to the publications we consulted, between 0.4% and 26.9% of patients with MS develop uveitis, and between 7.8% and 14.8% of patients with IU develop MS.[1,3] However, other diseases are associated with IU, including Lyme disease, syphilis, cat scratch disease, and sarcoidosis.[1]

We present a patient with an unusual initial presentation of MS involving central retinal artery occlusion (CRAO) in the right eye (RE). This highly atypical case may represent one extreme of the retinal vasculitis spectrum associated with demyelinating diseases.

Case report

We present a 46-year-old woman who was admitted to the emergency room in 2010 with a sudden decrease in the RE visual acuity (VA), which had an onset several hours prior. She had mild pain with eye movement. The patient did not have a relevant medical history and was medicated only with oral contraceptives.

In the emergency room, the blood pressure was 101/67, and the examination revealed a VA of 6/200 in the RE and

Received 9 September 2014; received in revised form 19 October 2014; accepted 3 December 2014; available online 11 December 2014.

[a] Neuro-ophthalmology Division, King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia
[b] Vitreoretinal Division, King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia
[c] Unidad de Neuro-oftalmología, Hospital Ruber Internacional, Spain

* Corresponding author at: Neuro-ophthalmology Division, King Khaled Eye Specialist Hospital, Riyadh, Saudi Arabia.
e-mail address: algarui@yahoo.com (A. Galvez-Ruiz).

Peer review under responsibility of Saudi Ophthalmological Society, King Saud University

Production and hosting by Elsevier

Access this article online:
www.saudiophthaljournal.com
www.sciencedirect.com

20/20 in the left eye (LE). The intraocular pressure was 15 mmHg in the RE and 14 mmHg in the LE.

The pupils were isochoric and normoreactive to light and accommodation, but with the presence of a relative afferent pupillary defect in the RE. The slit lamp revealed RE conjunctival ciliary injection and the presence of mild inflammation, 1+ cells, and flare in the anterior chamber. There were no posterior synechiae.

Conspicuous in the fundus of the eye was a hyperemic, slightly elevated optic disk, with ischemic pallor of the retina at the posterior pole that was compatible with acute CRAO and a preserved cilioretinal artery (Fig. 1A). Likewise, there was vascular sheathing and vitritis with snowballs.

The fluorescein angiogram showed mild signs of retinal vasculitis, retinal arterial filling delay, diffuse retinal capillary non-perfusion in the posterior pole compatible with CRAO. Preservation of the retinal circulation around the cilioretinal artery was visible, as was contrast leakage from the optic disk in the subsequent frames (Fig. 1B–C). The macular optical coherence tomography (OCT) showed increased retinal reflectivity consistent with CRAO (Fig. 2).

Given the presence of typical periventricular demyelinating lesions (some showing contrast uptake) in the cranial and orbital MRI, as well as slight enhancement of the optic nerve after contrast injection, it was believed that the patient might have had optic neuritis in addition to uveitis (Fig. 3). Consequently, treatment was initiated with intravenous methylprednisolone, 1 g every 24 h for three days (single dose), followed by a tapered oral dose (1 mg/kg tapered gradually in 2 weeks). Likewise, topical treatment was initiated using a corticosteroid in the RE (prednisolone acetate drops three times a day, gradually reducing the dose in 4 weeks).

Upon ophthalmologic examination two months later, the VA was 20/50 in the RE and 20/20 in the LE. There was no inflammation in the anterior chamber, and the fundus of the RE displayed global pallor of the optic nerve with retinal arteriolar attenuation at the posterior pole. The neurological examination was normal. The VA improvement was believed to be caused by the decreased in macular thickening or optic neuritis improvement after corticosteroid treatment.

The requested blood analyses, which included C-reactive protein (CRP), Anti-nuclear Ab (ANA), Anti-DNA, Treponema pallidum hemagglutination assay (TPHA), Rapid Plasma Reagin (RPR), Angiotensin converting enzyme (ACE), serum Lysozyme, and coagulation risk profile, were all within normal limits. Likewise, a chest CT scan, purified protein derivative (PPD), EKG, echocardiogram, and Doppler of the carotids were obtained, with results within normal limits.

Goldmann's visual field revealed a residual central and temporal islet in the RE and normal findings in the LE (Fig. 4).

During two years of follow-up, the patient remained clinically stable. However, the appearance of new periventricular lesions on subsequent MRIs, combined with the suspicion of initial optic neuritis, has led to the diagnosis of MS and the initiation of treatment with interferon beta-1 by the Neurology Service (Fig. 5).

Discussion

We present a patient with an unusual initial presentation of MS combining a CRAO with a possible optic neuritis in the RE. Such an atypical case may represent one extreme of the retinal vasculitis spectrum associated with demyelinating disease.

The IU associated with MS is characterized by the presence of pars planitis (occasionally accompanied by anterior uveitis) and the presence of peripheral retinal vasculitis in the form of periphlebitis (venous sheathing) in 6–26% of patients.[3]

In the majority of patients, these retinal vessel changes are asymptomatic. Regardless, certain studies have shown that the presence of peripheral vascular disease in a patient with isolated sheathing in typical optic neuritis may predict the development of MS in the future, with a relative risk of 11.4%.[4,5] Other studies have linked the presence of vascular sheathing with the severity of the neurological dysfunction in MS patients.[3,6]

Nonetheless, despite the fact that in the majority of patients, these retinal vascular changes are asymptomatic, there are numerous publications demonstrating a spectrum that ranges from simple peripheral retinal periphlebitis to the presence of peripheral occlusive retinal vasculitis in 6.5% of patients.[1,4,7] This condition leads to other complications, such as retinal ischemia, neovascularization, retinal detachment, vitreous hemorrhage,[2] or neovascular glaucoma.[8]

This patient belongs within the group of patients with occlusive retinal vasculitis associated with demyelinating disease. However, this case is unusual in that the intense retinal vasculitis was associated with CRAO.

The pathogenic mechanism that occurs in retinal vasculitis is that of infiltration of the retinal venules by T-cell lymphocytes, of which 35–90% are CD4+.[1]

Recent immunogenetic studies have revealed that the HLA-DR15 allele may encode a predisposition for the development of MS and pars planitis, which demonstrates that the processes share a similar immunopathogenic substrate.[9,10]

In patients for whom MS and IU are associated, MS tends to be the first disease to present clinically. Thus, in the study by Biousse,[3] neurological symptoms occurred nine years prior to the start of uveitis in 57.1% of cases. In addition, only one patient in the study had an initial presentation with simultaneous ocular and neurological symptoms.

Our patient's visual symptoms led to the diagnosis of MS through a cranial MRI, which showed typical demyelinating lesions. However, she had not previously presented with neurological symptoms.

With respect to the differential diagnosis of occlusive retinal vasculitis, in addition to the demyelinating etiology, we must consider sarcoidosis, Behçet's disease, and tuberculosis. Additionally, as in MS, all of these diseases may alter the central nervous system (for example, neuro-Behçet and neuro-sarcoidosis), thus further complicating the differential diagnosis in certain cases.[8]

A study conducted in Tunisia described 18 patients with CRAO secondary to posterior uveitis. The most frequent etiology was toxoplasmosis, followed by rickettsiosis and Behçet's disease. There were no patients with demyelinating disease, and only one patient presented with an idiopathic etiology.[11] Similar studies have shown that the presence of CRAO associated with MS is infrequent. Nonetheless, there are publications in which the presence of CRAO and branch retinal artery occlusion (BRAO) is described in optic neuritis patients, with similar frequencies from demyelination versus post-vaccination.[12–15]

In the study by Kahloun et al., various mechanisms are proposed to explain the occurrence of retinal arterial occlusions