Case Report

Regression of peripapillary choroidal neovascular membrane in a patient with sarcoidosis after oral steroid therapy

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Abstract

Choroidal neovascular membrane (CNV) may occur in patients with posterior uveitis. Treatment of patients with corticosteroids induces regression of the inflammation in the posterior pole with downregulation of many cytokines including vascular endothelial growth factors. We report herewith, a case of biopsy proven sarcoidosis that developed posterior uveitis and peripapillary CNV membrane and subretinal hemorrhage with fluid. The patient was treated with systemic steroids. She demonstrated progressive regression of the CNV membrane and complete resolution of the subretinal hemorrhage and fluids. In conclusion, control of the posterior segment inflammation is crucial in the resolution of the CNV membrane in uveitis and the intravitreal anti-vascular endothelial growth factor may not be always indicated.

Keywords: Sarcoidosis, Peripapillary CNV, Oral steroids

Introduction

Sarcoidosis is a chronic multisystem inflammatory disorder of unknown etiology. The disease is characterized by non-caseating granulomata that affect many organs, including the lungs, lymph nodes, skin, heart, liver, muscles and eye. Ocular lesions are common among patients with sarcoidosis.1 Choroidal neovascular (CNV) membrane occurs rarely in patients with sarcoidosis, but can be vision-threatening when it involves peripapillary locations.2–4 Peripapillary CNV membrane is characterized clinically by the presence of a CNV membrane adjacent to the disc which may lead to subretinal hemorrhage, fluid, or exudates.5 We report herewith, a case of peripapillary CNV membrane in a patient with sarcoidosis which showed regression after oral steroid therapy.

Case report

A 40-year-old woman presented to The Eye Center, Riyadh, KSA with itching and dryness of both eyes for long duration. Her best corrected visual acuity (BCVA) was 20/60 in the right eye and 20/30 in the left eye. The decrease of vision in the right eye was due to anisometropic amblyopia. Schirmer test was 0 mm in both eyes. Slit-lamp biomicroscopy and funduscopy were normal bilaterally. She was diagnosed with dry eye syndrome and was treated with topical lubricants and the application of punctal plugs. On her follow up visit, she came complaining of increased irritation in both eyes and xerostomia. A labial biopsy of the accessory salivary glands was performed by one of us (KFT) and tissue specimens were subjected to histopathologic evaluation.

Received 27 January 2014; received in revised form 18 February 2014; accepted 25 February 2014; available online 6 March 2014.

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This Study was supported in part by a Special Fund from The Eye Center and The Eye Foundation for Research in Ophthalmology, Riyadh, Saudi Arabia.

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Histopathology revealed non-caseating granuloma that was consistent with sarcoidosis. The granuloma was composed of epithelioid histiocytes, multinucleated giant cells and mononuclear cells. There were no lymphoepithelial lesions to suggest Sjögren’s syndrome or malignancy. Modified Ziehl–Neelsen stain was negative for mycobacteria.

Later, the patient developed anterior granulomatous uveitis in the right eye with mutton-fat keratic precipitates and three large Koepppe nodules (Fig. 1). She was referred to a pulmonologist for further evaluation and was found to have mild restriction of the lung function tests. Chest X-ray revealed bilateral hilar lymphadenopathy. Serum Alkaline phosphatase was 300 U/L (normal range 30–125). Percutaneous liver biopsy was done and histopathological evaluation of biopsy specimens was consistent with sarcoidosis. The patient was diagnosed with sarcoidosis affecting the lungs and liver.

Subsequently, the patient presented with the history of blurring of vision in the left eye. Her best corrected visual acuity (BCVA) was 20/60 in the right eye and 20/30 in the left eye. Flaremetry with KOWA FM-600 Laser Flaremeter was 10 photons/ms in the right eye and 8 photons/ms in the left eye. Biomicroscopy revealed anterior granulomatous uveitis in both eyes. Funduscopy of the right eye was normal and the left eye revealed the peripapillary CNV membrane with subjacent hemorrhage (Fig. 2). Optical coherence tomography was done and revealed subretinal fluid adjacent to the optic nerve head and dry macula with smooth vitreoretinal interface (Fig. 3). Fundus fluorescein angiography showed staining of the CNV membrane and adjacent hypofluorescence corresponding to the area of hemorrhage.

The patient was given prednisone 20 mg orally daily and topical prednisolone acetate to both eyes. She was maintained on 10 mg oral prednisone. After two months the CNV membrane started to regress in size and areas of hemorrhages were noted to clear. Funduscopy of the left eye revealed marked regression of the CNV membrane and complete resolution of the peripapillary hemorrhages. Oral prednisone was tapered and discontinued. The patient was followed-up for a period of one year with no recurrence of the CNV membrane (Fig. 4).

Discussion

Peripapillary CNV membrane may occur in association with several conditions including age-related macular degeneration which is the most common cause (45.2% of the cases), multifocal choroiditis (4.3%), angioid streaks (2.6%), histoplasmosis (1.7%), choroidal osteoma (1.7%), optic disc drusen (0.9%), and congenital disc anomaly (0.9%). In cases of uveitis, chronic disc edema may present prior to the onset of the disease. Chronic disc swelling may induce peripapillary CNV membrane by increasing the potential space between the
Bruch’s membrane and the swollen disc tissue facilitating ingrowth of choroidal vessels around the termination of the Bruch’s membrane. Alternatively, chronic disc edema may result in the deformation of the peripapillary Bruch’s membrane leading to breaks in the Bruch’s membrane which allows ingrowth of the choroidal neovessels under the retinal pigment epithelium (RPE). Clinically, peripapillary CNV membrane is characterized by the presence of a CNV membrane adjacent to the optic nerve producing subretinal hemorrhage, fluid, and/or exudates. Several treatment modalities for sarcoid-related peripapillary CNV membrane have been reported. These include systemic, oral and periocular steroids, argon laser photocoagulation and intravitreal anti-vascular endothelial growth factor agents.

In our case, the initial presentation of ocular sarcoidosis was with signs and symptoms of dry eye syndrome that were treated with topical lubricants and punctal plugs. Later, patient developed xerostomia and labial biopsy of the accessory salivary glands was performed. The biopsy revealed sarcoidosis that was subsequently confirmed by a biopsy of the liver. The diagnosis of systemic sarcoidosis was established in our patient prior to the development of ocular manifestations of the disease. Ocular involvement started with anterior granulomatous uveitis without posterior segment affection. Peripapillary CNV membrane developed later in the left eye. The CNV membrane responded to oral prednisone therapy without the need for intravitreal or periocular injections.

CNV membrane in cases of sarcoidosis was found to respond well to oral steroids. Corticosteroids induce regression of the inflammation in the posterior pole in cases of posterior uveitis with downregulation of many cytokines including VEGF. The good response to oral prednisone seen in our patient emphasizes the importance of trial of oral immunosuppression before considering periocular injections of steroids or intravitreal injections of vascular endothelial growth factor inhibitors.

**Conflict of interest**

The authors declared that there is no conflict of interest.

**References**