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Case Report

Idiopathic Orbital Inflammatory Disease Attack Associated with Large Serous Detachment Without Anterior and Posterior Segment Inflammation

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Abstract

Idiopathic orbital inflammatory disease is a benign, non-infectious, and non-neoplastic space-occupying orbital and peri-orbital inflammation with no identifiable local or systemic causes. Patients usually present with eyelid and periorbital erythema and edema, proptosis, and decreased eye movements. In this report, a case of large serous retinal detachment accompanying a unilateral idiopathic orbital inflammatory disease attack is discussed. It demonstrates that very extensive and highly serous retinal detachment may accompany idiopathic orbital inflammatory disease attacks. Dramatic improvement in clinical findings and imaging can be observed with high-dose steroid treatment in a short time.

Keywords: Idiopathic orbital inflammatory disease, orbital inflammation, retina, serous detachment, steroids

Introduction

Idiopathic orbital inflammatory disease is a benign, non-infectious, and non-neoplastic space-occupying orbital and peri-orbital inflammation with no identifiable local or systemic causes. After Graves' disease and lymphoproliferative disorders, idiopathic orbital inflammatory disease is the third most common ophthalmologic disease of the orbit, accounting for approximately 8–11% of all orbital diseases. For example, in a study of patients evaluated for a suspected orbital mass, 11% of histopathologically proven cases consisted of inflammatory lesions (1). In these patients, eyelid and periorbital erythema and edema, conjunctival congestion, proptosis, ptosis, diplopia, light sensitivity, decreased eye movements, and pain with eye movements may be present. Dacryoadenitis, myositis, scleritis, or diffuse soft tissue inflammation may occur (2-4). Symptoms are usually unilateral.

In this report, a unique case with large serous retinal detachment accompanying a unilateral idiopathic orbital inflammatory disease attack without any findings of anterior or posterior segment inflammation is discussed. This case report was performed in compliance with the Declaration of Helsinki.

Case Report

A 52-year-old female patient, who was followed up in the oculoplastics departments of Kocaeli University Faculty of Medicine, Department of Ophthalmology, with the diagnosis of idiopathic orbital inflammatory disease, applied to our clinic with complaints of redness in her right eye and swelling of the eyelid for 3 days. The best corrected visual acuity was 20/200 on the right eye and 20/20 on the left eye. The right eye was 3–4 mm proptotic with restricted extraocular movement in all directions of gaze. There was

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no relative afferent pupillary defect. Intraocular pressure was 18 mmHg bilaterally. In the biomicroscopic examination, there was no anterior chamber reaction bilaterally. There was intense chemosis, injection, and periorbital edema on the right eye; the left eye was normal. Fundus examination revealed serous retinal detachment without vitreous inflammation on the right eye, and the left eye was normal. Optical coherence tomography (OCT) showed a large and high serous detachment on the right eye (Fig. 1a). Choroidal thickness measurements with enhanced depth imaging (EDI) OCT were 261 µ in the right eye and 257 µ in the right eye. We performed FFA for the patient. We did not see any dark spots or leakage on the vascular bed. There was no capillary leakage or any inflammation finding. B-scan ultrasonography (U/S) revealed serous retinal detachment without posterior scleritis findings, including T-sign. On the contrast-enhanced orbital magnetic resonance imaging examination, diffuse inflammatory signal changes and enhancement were observed in the intraconal fatty tissue with proptosis on the right eye (Fig. 2). A systemic workup, including vasculitis panels and infectious markers, was performed, and all evaluations were normal. The patient was treated with 2 × 500 mg of methylprednisolone for 3 days. We continued the steroid therapy with 16 mg of methylprednisolone (Prednol®) twice a day for a total of 15 days. We discontinued the therapy without tapering. On examination in the 1st week, visual acuity increased to 20/25 on the right eye, serous detachment regressed, and retinal pigment epithelial changes were observed in fundus examination and OCT (Fig. 1b).

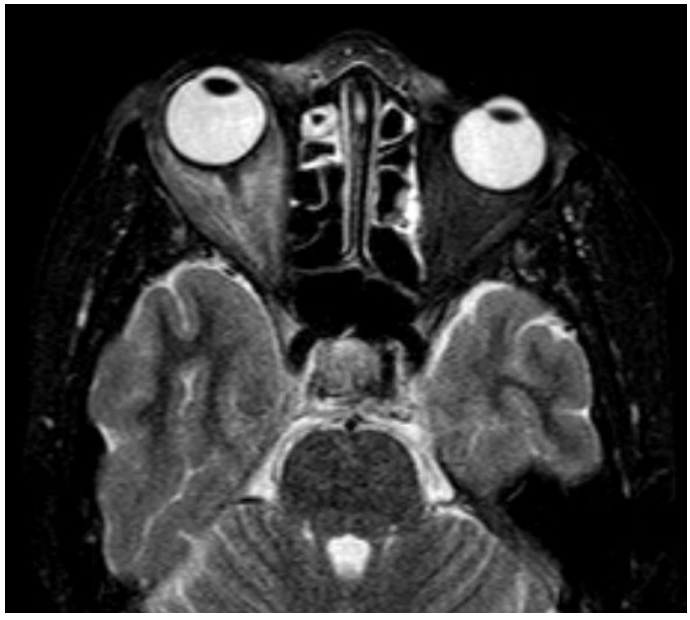

Figure 2. Contrast-enhanced magnetic resonance imaging section of the orbit at admission.

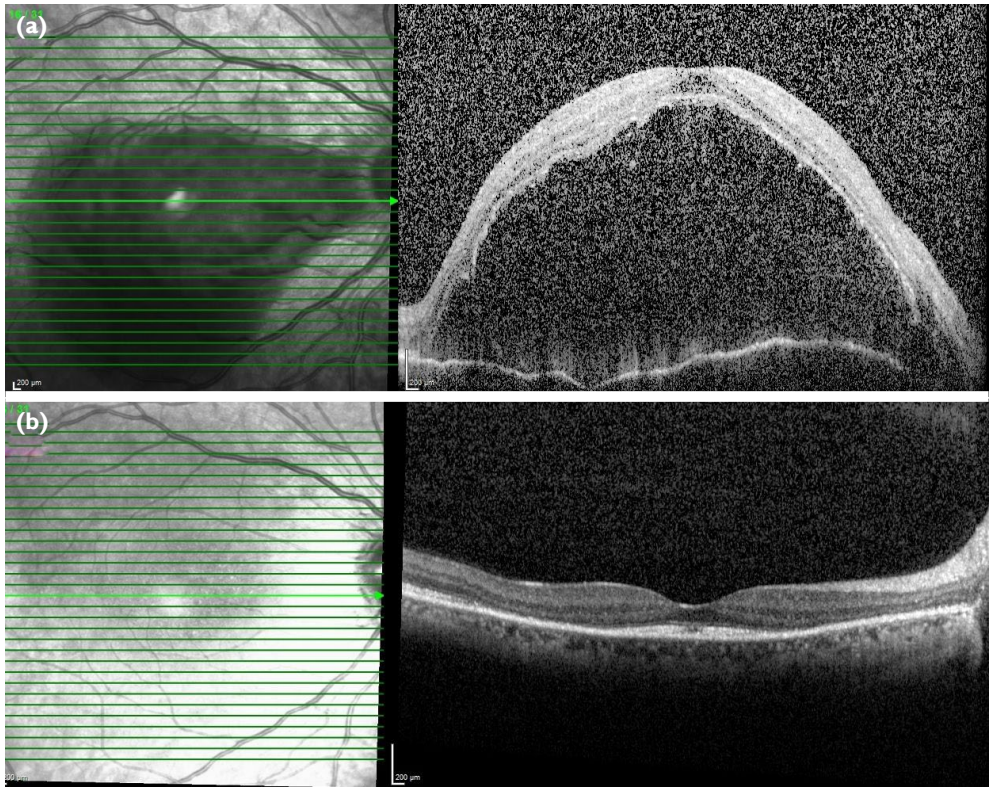

Figure 1. (a) Macular optical coherence tomography (OCT) at patient admission. **(b)** Macular OCT taken in the 1st week of admission.

Discussion

This case illustrates that very extensive and highly serous retinal detachment may accompany idiopathic orbital inflammatory disease attacks without findings of anterior and posterior segment inflammation. Despite poor visual acuity and worrisome OCT findings at admission, dramatic improvement in visual acuity, clinical findings, and imaging could be observed with high-dose steroid treatment in a short time.

There have been two cases reported in the literature describing retinal detachment in the course of idiopathic orbital inflammatory disease. Furthermore, retinal detachment in those cases was associated with intense inflammation in the posterior sclera, as proven by B-scan U/S. Chaudhry et al. reported a young male case of unilateral idiopathic orbital inflammatory disease presented with exudative retinal detachment and optic neuritis attributed to posterior scleritis evidenced by the T-sign on the right-sided U/S (5). Systemic corticosteroid therapy was started, and on the 3rd day, his vision had improved to 20/20, along with exudative retinal detachment as well as other symptoms. Yuen et al. described a 9-year-old Chinese girl with bilateral idiopathic orbital inflammatory disease who presented with a bilateral anterior chamber reaction of 1+ cells, severe bilateral optic disc edema, and exudative retinal detachments involving the macular region (6). In that case, again, B-scan U/S was suggestive of posterior scleritis. Clinical improvement was observed in 24 h with 1 mg/kg oral prednisolone and topical steroid therapy. What made the present adult case with a previous diagnosis of idiopathic orbital inflammatory disease different than these two reported cases was the absence of signs of inflammation in the anterior segment, no scleral inflammation, increased thickness or T-sign in the U/S, but only serous detachment in the fundus examination, and OCT in the patient's current attack. Our case is the first to demonstrate serous retinal detachment without evident anterior and posterior segment inflammation in an idiopathic orbital inflammatory disease attack in the literature. Based on this report, it can be suggested that diffuse inflammation in idiopathic orbital inflammatory disease may cause leakage of choroidal vessels through the interconnection of the retinal pigment epithelium without causing increased choroidal thickness in EDI OCT or T sign in the U/S.

Relapses are common in idiopathic orbital inflammatory diseases, especially bilateral ones. Our patient was a patient with unilateral involvement with previous recurrences. Systemic corticosteroids are the most common treatment modality for idiopathic orbital inflammatory disease. Improvement with corticosteroid therapy is also helpful in the diagnosis because corticosteroid-responsive orbital inflammation is likely to strengthen the diagnosis of idiopathic orbital inflammation (2-4,7). More than half of patients show dramatic improvement within 24–48 h of starting systemic corticosteroids. As a matter of fact, we think that the serous detachment seen without evident anterior or posterior segment inflammation in our present case also responded to corticosteroid treatment, which contributes to the literature. Intraorbital corticosteroid injection has been found to be a useful and effective treatment for idiopathic orbital inflammatory disease and may be administered as first-line therapy in some patients (8). Low-dose radiation may be considered in the elderly, in patients who do not improve with systemic corticosteroids, or in whom steroids are contraindicated (2-4,7). Immunosuppressive agents (Methotrexate, Cyclosporine, Azathioprine, and Mycophenolate Mofetil) and biological agents (Rituximab, Infliximab, and Adalimumab) are used in patients who do not respond to first-line treatments or who have frequent recurrences (2).

Conclusion

Extensive serous retinal detachment accompanying idiopathic orbital inflammatory disease attacks is a rare condition seen in the literature at a rate of 1/140 (9). In general, these detachments accompany inflammation in the anterior and posterior segments. The absence of inflammation in the anterior and posterior segments in this case and the dramatic improvement in clinical findings and imaging in a short time with high-dose steroid treatment make the case even more interesting.

Disclosures

Informed consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Conflict of Interest: None declared.

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