

Idiopathic Orbital Inflammatory Syndrome: Pseudotumor Orbita

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Abstract

The idiopathic orbital inflammatory syndrome (pseudotumor orbita) is an unexplained disease that mimics the complications of acute rhinosinusitis. Intense polymorphic lymphoid infiltration develops with inflammation. The masses in the orbita can be seen by 5% to 8%. It can cause nonspecific orbital symptoms, such as proptosis and swelling on eyelids. In our case, the patients presented to the eye outpatient clinic with swelling complaint and acute atopic conjunctivitis treatment was initiated but the patients who did not regression complaints were submitted. The patient's magnetic resonance imaging (MRI) showed bilateral lateral rectus muscle thickening. Idiopathic orbital inflammatory syndrome (pseudotumor orbita) responded to the steroid treatment. Idiopathic orbital inflammatory syndrome (pseudotumor orbita) is benign, but can be confused with sinusitis complications with high morbidity and mortality.

Keywords: Proptosis, pseudotumor orbita, sinusitis, steroid

INTRODUCTION

Idiopathic orbital inflammatory syndrome or pseudotumor orbita is a disease with unknown etiology, which displays polymorphic lymphoid infiltration (1). It is important for otorhinolaryngologists because it is a rare disease imitating the complications of acute rhinosinusitis. In this study, it was aimed to present a rare case encountered in the practice of otorhinolaryngology.

CASE PRESENTATION

A 31-year-old male was admitted the outpatient clinic of ophthalmology for swelling of the eyes. Medical treatment was arranged for the patient who was suspected to have acute atopic conjunctivitis. Because he did not benefit from medical treatment, he revisited the outpatient clinics of otorhinolaryngology and ophthalmology. Bilateral swelling around the eyes and proptosis were observed in the patient (Figure 1a, b).

His physical examination revealed slight purulent drainage in the bilateral nasal passages. No restriction was observed in visual acuity and eye movements. Paranasal sinus computed tomography (CT) and orbital magnetic resonance imaging (MRI) were ordered for the patient who was hospitalized because of pre-diagnosis with acute sinusitis complication. Elevated white blood cell, CRP, and sedimentation were detected. Because he had thickening in the lateral rectus muscle and contamination/signal changes in the orbital fat tissue in consistency with bilateral pseudotumor orbita in the MRI of the orbita, steroid therapy was initiated (Figure 2a, b). The patient was discharged when his complaints regressed after steroid therapy. Informed consent was signed by the patient.

DISCUSSION

Idiopathic orbital inflammatory syndrome is a benign, rare inflammatory disease of the orbita with unknown etiology and bilateral lymphocytic infiltration. Among orbital pathologies, it is the third most common disease after Graves' orbitopathy and lymphoproliferative disorders (2). It constitutes 5% to 8% of all orbital masses (2). Fat tissue around the orbita, extraocular muscles, optic nerve, and lacrimal gland can be affected by the disease. It can lead proptosis, restricted eye movements, pain in the eyes, swelling in the eyelids, and erythema in the eyes. In case of

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Figure 1. a, b. Patient with bilateral swelling around the eyes and chemosis

delayed diagnosis, diplopia, poor vision, optic atrophy, and dacryoadenitis symptoms can develop. Idiopathic orbital inflammatory syndrome can be accompanied by fibrosis of different degrees (3). Because of these complaints, it can be confused with symptoms associated with sinusitis complications. Neoplastic, infectious, vascular, and trauma-related factors should be ruled out in the differential diagnosis (3).

The diagnosis of idiopathic orbital inflammatory syndrome (pseudotumor orbita) is established with anamnesis, physical examination, and imaging techniques. Magnetic resonance imaging (MRI) is the most appropriate imaging technique for pseudotumor orbita (4). On MRI, fibrosis and sclerosis with different degrees can be observed (4, 5). Histopathological diagnosis cannot be required except for atypical cases. Histopathologically, different grades of fibrosis are observed along with lymphocytic cell infiltration (6).

The first treatment choice for idiopathic orbital inflammatory syndrome is corticosteroids. Corticosteroid therapy should be cut slowly because

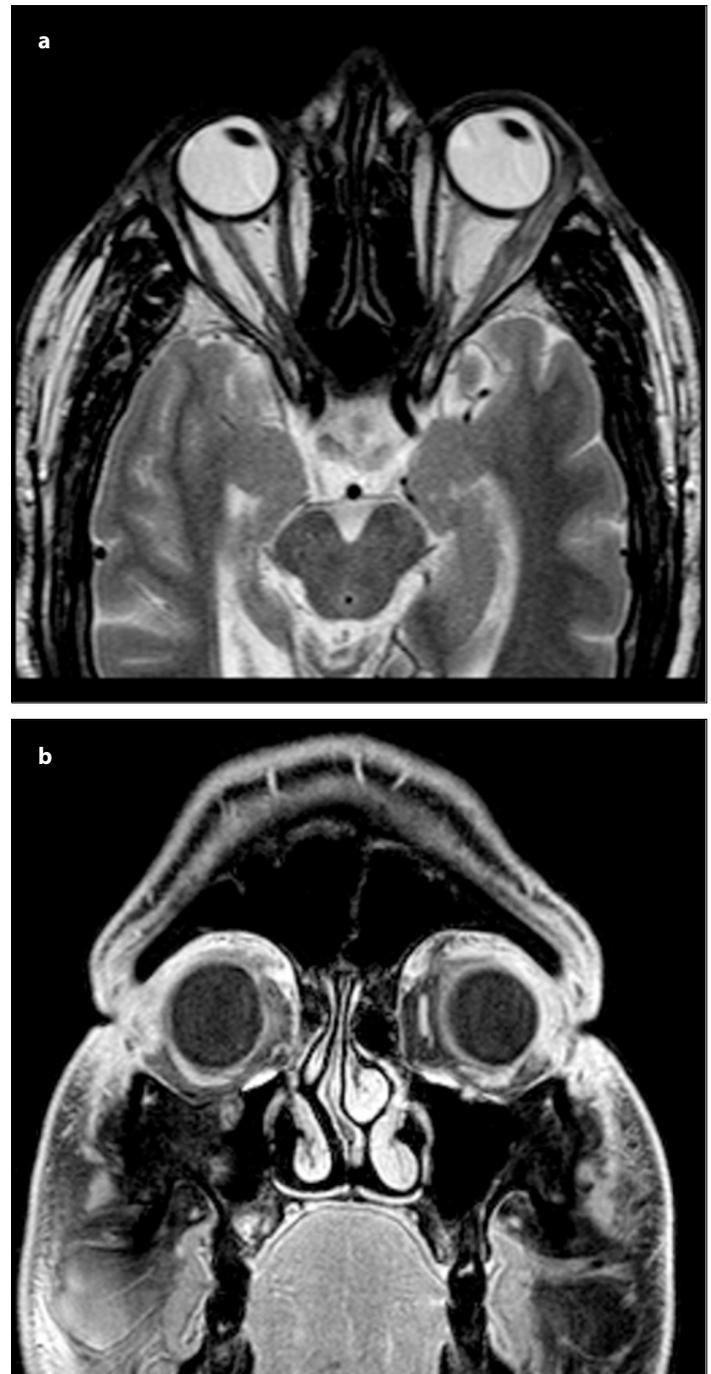


Figure 2. a, b. MRI sections on axial and coronal planes

the disease tends to recur (7). However, cyclosporin, methotrexate, and cyclophosphamide can be applied in atypical cases not responding to steroids (8).

CONCLUSION

Idiopathic orbital inflammatory syndrome (pseudotumor orbita) is a benign disease that is rarely seen but should be considered in differential diagnosis. Besides the clinical findings that can be confused with sinusitis complications such as orbital cellulitis, it is a diagnostic characteristic that is bilateral, a recurrence story and good response to steroid therapy.

Informed Consent: Written informed consent was obtained from patient who participated in this case.

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