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Case Report

A Rare Cause of Diplopia: Idiopathic Orbital Myositis

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ABSTRACT

Orbital myositis is an entity affecting the ocular muscles, especially the medial rectus. These cases are usually referred to clinics with complaints such as diplopia, orbital/periorbital pain, limitation in ocular movements, increased pain with eye movements, proptosis, swelling of the eyelid and/or hyperemia in the conjunctiva. Orbital myositis is usually idiopathic and autoimmunity is often suspected in etiology. In this article, we present a 15-year-old girl who presented with diplopia, pain in both eyes, anomalous head posture, periorbital edema and was diagnosed with idiopathic orbital myositis through history, clinical findings and imaging methods.

Keywords: Orbital myositis, diplopia, anomalous head posture

Introduction

Idiopathic orbital myositis is a syndrome of the acute onset of single or multiple inflammation of the extraocular muscles. This type of myositis is represented in a broad clinical classification of idiopathic orbital inflammatory pseudotumor (1). However, the idiopathic orbital inflammation and non-specific orbital inflammation overlap considerably, and occasionally the two entities can be considered interchangeably. Idiopathic orbital myositis can be described as a clinical syndrome of inflammation (2). In addition to muscle tissue, the disease can also affect other structures of the orbit including fat, lacrimal glands or connective tissues (3). Its etiology is not clear yet. However, it is reported to be associated with polymyositis, thyroid diseases, juvenile idiopathic arthritis, and other rheumatological diseases. It has also been shown that there is an association with several diseases (4,5). Idiopathic orbital myositis accounts for 6-17% of cases in childhood (6,7). It is seen in females from two to four times more often than in males and causes inflammation in extraocular muscles. Due to the inflammation, orbital signs and symptoms, such as pain, proptosis, ptosis, periorbital pain, diplopia, ophthalmoplegia, conjunctival hyperemia or ocular injection can be seen (8). Diplopia is due to the inadequate contraction of the affected eye muscles that can bring about an anomalous head posture characterized by head tilting to the side opposite the inflamed muscles to prevent diplopia (9). Diagnosis of idiopathic orbital myositis can be done after ruling out many diseases that might present with orbital inflammation features. In the magnetic resonance imaging (MRI) of idiopathic orbital myositis cases, there are various patterns of extraocular muscle involvement (3). In this article, a 15-year-old girl with idiopathic orbital myositis, who had no form of chronic systemic illness, presenting with swelling-redness in the left eye lid, diplopia for fifteen days, and anomalous head posture for one week is discussed and a brief review of the literature is also given.

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Case Report

A previously healthy 15-year-old girl presented to our pediatric hospital with a 2-week history of swelling-redness, gradually progressing ptosis in the left eye lid, diplopia, and anomalous head posture. Her ophthalmologic examination revealed painful extraocular movement in all directions with normal visual acuity. The left eye was hyperemic and had alternating exotropia with proptosis. There was a motility restriction of the left eye towards the right side. On her examination of primary view position, she complained of diplopia and, in order to cope with the diplopia, she adopted a mild anomalous head position to the right and downwards with the jaw to the left. (Figures 1 and 2). Direct and indirect light reflexes of the patient were normal in both eyes. Frontal and posterior segment examinations of the eyes were performed. Hemoglobin, biochemical investigations, C-reactive protein and sedimentation rate were normal. Autoimmune tests such as antinuclear antibody, anti-deoxyribonucleic acid, rheumatoid factor, c-anti-neutrophil cytoplasmic antibody were negative. There was no positivity on serological investigations of lyme, *Brucella*, toxoplasma, toxocara and other viral agents. Gadolinium-enhanced T_1 and T_2-weighted MRI of the orbit revealed inflammation in bilateral extraocular muscles which was more pronounced in the left medial rectus suggesting orbital myositis (Figures 3 and 4). Cranial MRI was reported as normal. Thyroid ophthalmopathy, intracranial pathologies, rheumatologic diseases and infections were ruled out and the case was diagnosed with idiopathic orbital myositis. Oral 60 mg/day methylprednisolone therapy was started. On the third day of the treatment, eye edema and proptosis were decreased. Methylprednisolone therapy was stopped by gradually reducing dosages over a four-week period and policlinic follow-up was performed due to the possibility of recurrence. Informed consent was obtained from our patients' parents.

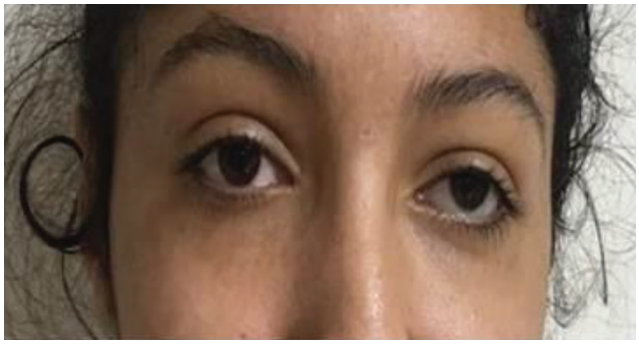

Figure 1. Primary view position of the left eye which was hyperemic and had alternating exotropia with proptosis

Figure 2. The patient's mild anomalous head position to the right and downwards, with the jaw to the left

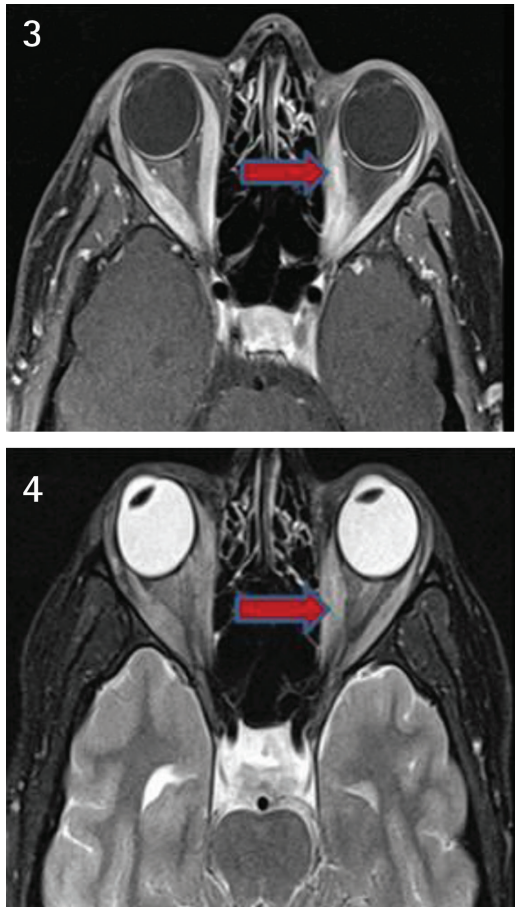

Figure 3, 4. Magnetic resonance imaging of the orbit showing inflammation in the bilateral extraocular muscles which was more pronounced in the left medial rectus

Discussion

This case report describes a young girl who developed idiopathic orbital myositis related to no systemic illness. Idiopathic orbital myositis, also called myositis subtype of idiopathic orbital inflammatory pseudotumor, first described by Gleason (10) in 1903, is a myositis resulting in the inflammation of the extraocular muscles without any local or systemic reason. The disease is represented in a broad clinical classification of idiopathic orbital inflammation termed by Birch-Hirchfield in 1905 as an orbital pseudotumor condition. The most frequent clinical features of idiopathic orbital myositis are acute or subacute exacerbated orbital pain. In childhood, it is also reported to be associated with non-specific signs (3). It is widely seen in the female gender between 18-40 years of age. In studies, it has been reported to be in coexistence with diseases (1,3,11).

Acute, chronic or recurrent forms of idiopathic orbital myositis may be seen. The patients might present with symptoms such as pain in the eyes, strabismus, diplopia, proptosis, conjunctival hyperemia, and swelling-pain in the eyelids. Additionally, patients may suffer from abnormal head position as a result of diplopia as seen in our case. In a study (3), 86.4% of the cases with idiopathic orbital myositis were most frequently referred to as ophthalmologists with oculomotor disorder and strabismus. They reported that proptosis (68.2%), congestion-edema (65.9%) in the conjunctiva, swelling in the eyelids, pain in the eyes, and an increased sensation of pressure may also be seen in idiopathic orbital myositis. In the same study, diplopia was found in 43.2% of the patients.

Diagnosis of idiopathic orbital myositis is based on the history and clinical findings with the detection of one or more extraocular muscular involvement in orbital computerized tomography or orbital MRI. Diagnosis should be confirmed after excluding thyroid disorders, other orbital inflammatory conditions, vasculitis, sarcoidosis, orbital cellulitis and orbital tumors. Some studies indicated that the medial rectus muscular involvement is the most common involvement (9).

In conclusion, although orbital myositis is a rare entity in childhood, it should be considered in patients presenting with complaints of diplopia, proptosis, periorbital edema and anomalous head posture in differential diagnosis.

Ethics

Informed Consent: Informed consent was obtained from our patients' parents.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Concept: Ş.G., Data Collection or Processing: E.Ş., S.K.Y., Analysis or Interpretation: S.A., Literature Search: S.A., Writing: Ş.G., S.A.

Conflict of Interest: No conflict of interest was declared by the authors.

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