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CASE REPORT / OLGU SUNUMU

Orbital muscle involvement in a child with familial Mediterranean fever

Ailevi Akdeniz ateşi tanılı bir çocuk hastada orbital kas tutulumu

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ABSTRACT

Familial Mediterranean fever is an autosomal recessively inherited autoinflammatory disorder characterized by recurrent attacks of fever, serositis and synovitis. Various atypical presentations have been reported in the literature. Herein, we report a case of orbital myositis presenting with acute and devastating ocular symptoms resembling orbital cellulitis in a 12-year-old boy with familial Mediterranean fever.

Keywords: Familial Mediterranean fever, Eye involvement, Orbital myositis

ÖZ

Ailevi Akdeniz ateşi otozomal resesif olarak kalıtılan, tekrarlayan ateş, serözit ve sinovit atakları ile karakterize otoinflamatuar bir hastalıktır. Literatürde hastalığın farklı bulgular ile başlayabileceğini bildiren yayınlar mevcuttur. Bu yazımızda, orbital selülit benzeri ani ve yoğun göz tutulumu bulguları olan ancak takibinde orbital myozit tanısı konulan, ailevi Akdeniz ateşi tanılı 12 yaşında bir erkek hasta sunulmuştur.

Anahtar kelimeler: Ailevi Akdeniz ateşi, Göz tutulumu, Orbital myozit

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Introduction

Familial Mediterranean fever (FMF) is an autosomal recessively inherited autoinflammatory disorder characterized by recurrent attacks of fever, serositis and synovitis [1]. Although the diagnosis is made during childhood based on the clinical features, atypical presentations may cause a delay in the recognition of the disease. Muscle involvement with the symptoms of pain and signs of inflammation may accompany FMF. But as far as we know, orbital myositis has not been reported yet. We present a child with FMF who attended to the emergency clinic by acute onset of redness and swelling around the left eye and pain while moving the eye. Although, he was diagnosed as orbital cellulitis initially, the final diagnosis was orbital myositis (OM) due to FMF.

Case Report

A 12-year-old male patient who was being followed with the diagnosis of FMF and receiving colchicine regularly for 3 years was admitted to outpatient clinic with the complaints of swelling, pain and redness in the left eye. He had overt proptosis in this eye. It was learned that he had the same complaints in the right eye two weeks ago and all complaints had resolved in a few days without therapy. He did not have abdominal pain, fever or arthritis. Neurological examination was normal. In the ophthalmological examination visual acuity was full and retina was normal. Accomodation was normal and diplopia was not detected. He had severe pain while moving the left eye and conjunctival hyperemia was also detected. On laboratory tests C-reactive protein level was 21 mg/L, erythrocyte sedimentation rate was 53 mm/hr and leukocyte count was 13000/mm^3. After obtaining ophthalmological consultation, orbital cellulitis and OM

were thought as preliminary diagnoses and antibiotherapy was started. Orbital computerized tomography (CT) scan showed the enlargement of the medial rectus muscles bilaterally (Fig. 1). Two days after hospitalization, painful movement and redness occurred in the right eye. Symptoms of the left eye started to resolve. The diagnosis of orbital cellulitis was ruled out and antibiotherapy was stopped on the third day. Thyroid function tests were normal and anti-nuclear antibody was negative. Nonsteroidal anti-inflammatory therapy (ibuprofen 30 mg/kg/day) with the diagnosis of OM was started. One week after this therapy all the symptoms regressed and acute phase reactants became normal. He is being followed for 2 years without any complication or recurrence.

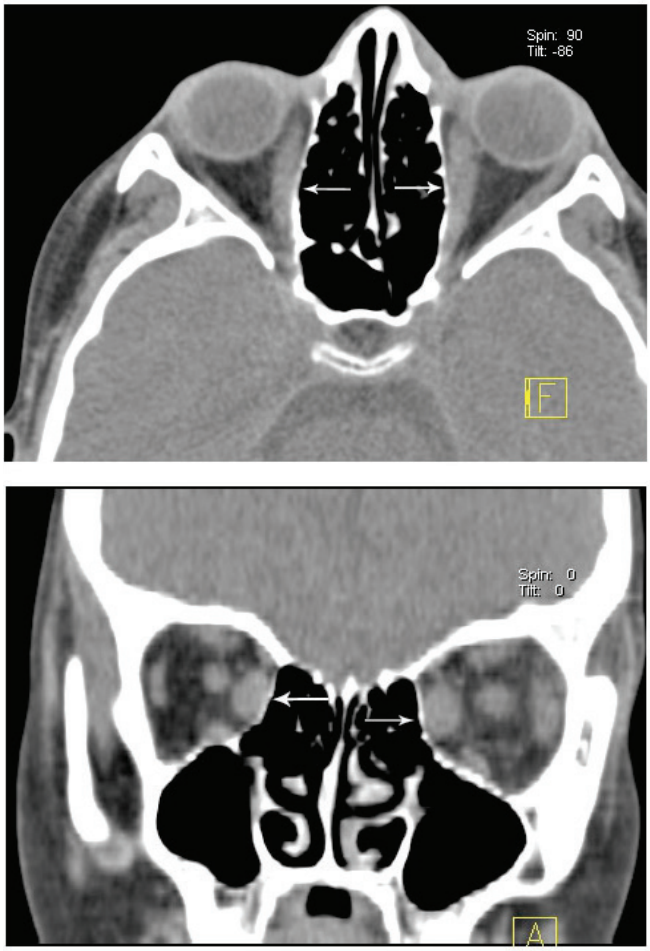

Fig. 1. Bilateral enlargement of the medial rectus muscles on the CT scans (marked with arrow)

Discussion

FMF is a disease with recurrent attacks of sterile inflammation of the peritoneum, pleura and synovia. During these episodes neutrophils migrate into the serosal and synovial spaces. Serous membranes are the main targets of the disease. Recurrent fever during early childhood may be the only manifestation of FMF [2]. Abdominal attacks are reported in 90%, articular symptoms in 75% and pleural involvement in 45% of the affected individuals along with the fever. Various atypical presentations such as recurrent isolated monoarthritis, isolated pleuritis and pericarditis, recurrent urticaria and meningitis have been reported [3-10]. Ocular pathologies such as uveitis, episcleritis and retinal changes have been described rarely in the literature [11-13].

OM is the enlargement of the extraocular muscles of the eye with infiltration of the inflammatory cells. Sign and symptoms are periocular pain, eyelid swelling and redness, restricted ocular motility and strabismus. The etiology can be various including spirochetal, viral and bacterial infections and some systemic diseases such as systemic lupus erythematosus, and rheumatoid arthritis [14,15]. Major differential diagnosis is thyroid ophthalmopathy [16]. In thyroid ophthalmopathy, orbital findings are usually bilateral and more than one muscle group is involved. There is no pain with eye movements, eyelid retraction is obvious, and there is no response to steroidal or nonsteroidal anti-inflammatory treatment [16]. Orbital cellulitis, lymphoproliferative disorders, and metastatic orbital diseases also should be considered in the differential diagnosis of OM. CT scans in OM demonstrate swelling around one or more extraocular muscles and T2-weighted magnetic resonance images show localized inflammation. For idiopathic OM cases steroids are the main treatment option [17].

In our case, OM had been shown radiologically by CT scan. Malignant diseases, infectious diseases and other inflammatory diseases such as thyroid ophthalmopathy were ruled out by clinical and laboratory examinations. We used ibuprofen with good response. To the best of our knowledge, this is the first case of OM associated with FMF. As a conclusion, OM should be kept in mind for children with FMF having ophthalmological signs and symptoms.

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