

Protracted febrile myalgia of familial Mediterranean fever

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ABSTRACT

Protracted febrile myalgia (PFM) includes severe myalgia of the upper and lower extremities accompanied by fever lasting up to 6 weeks, an elevated erythrocyte sedimentation rate and leucocytosis. We report a 13-year-old girl with PFM, and discuss the magnetic resonance imaging findings of the involved calf muscles. To our knowledge these are the only images of the pathology in the literature.

Introduction

The clinical and laboratory characteristics of protracted febrile myalgia (PFM) include severe myalgia of the upper and lower extremities accompanied by fever lasting up to 6 weeks, an elevated erythrocyte sedimentation rate and leucocytosis. Muscle biopsies and muscle enzymes are within normal limits, although autoimmunity has been implicated in the pathogenesis of PFM and responds to steroids (1). We report a case with magnetic resonance imaging findings of the involved calf muscles; to our knowledge these represent the only images in the literature.

Case report

A 13-year-old girl was referred to our hospital with severe muscle pain in her

right leg and knee accompanied by fever of 38°C within the last 3 months. On palpation, the right calf muscles were painful and the skin seemed tight. She was almost unable to walk and there was no any history of trauma. She held her ankle in an equinus position which had led to shortening of the Achilles tendon. Her medical history revealed many attacks of abdominal pain and fever, but since then she had not been diagnosed as having FMF. She had two very similar episodes of muscle pain in 1997 and 1999; small doses of corticosteroids maintained a complete recovery.

MRI images of the leg showed non-specific oedema of the subcutaneous fat tissue and the distal part of the medial gastrocnemius muscle prior to the musculo-cutaneous junction of the Achilles tendon. The characteristics of the signal in the oedema region were found to be highly suggestive of an inflammatory lesion rather than a traumatic contusion (Figs. 1 and 2). Total leucocytes, the erythrocyte sedimentation rate and C reactive protein were raised. Biochemical analysis was normal. Biopsy of the cutaneous and subcutaneous tissue with fascia of the muscle revealed scarce non-specific inflammatory infiltration with leuco-

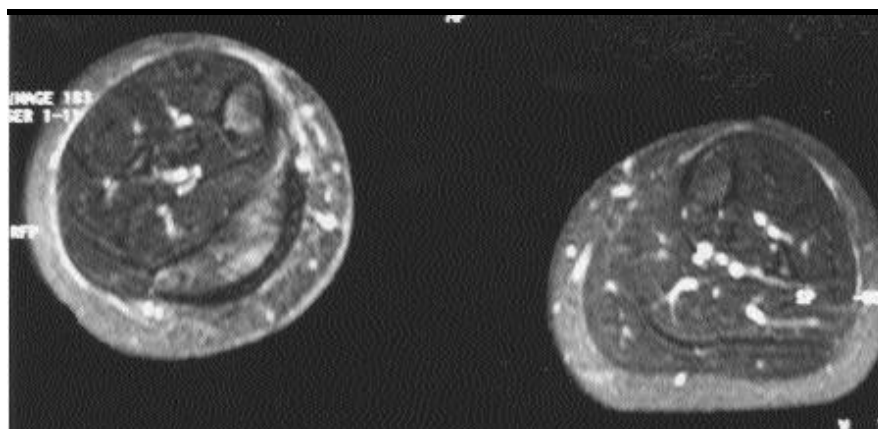


Fig. 1. T2-weighted axial image with fat saturation. TR 4952, TE 99, foV20. Diffuse oedema of the subcutaneous fat tissue. Increased signal intensity of the intermuscular septa and distal lateral part of the medial gastrocnemius muscle.

cytes, lymphocytes and eosinophils. Molecular study proved her to be heterozygote for M694V and V726A mutation. She was diagnosed as having FMF with protracted febrile myalgia. Although PFM is assumed to be a vasculitic disorder and responds to corticosteroids, we put her on just colchicum 0.5 mg tid, because her symptoms resolved spontaneously after her admittance to our hospital. During a follow-up period of 2 years on colchicum at the same dosage, she suffered no complaints. She can walk normally without any sequelae of Achilles tendon.

Discussion

PFM affects some FMF patients and is associated with relatively severe symptoms of FMF (2). The pain of myalgia in PFM is severe and disabling and generally lasts for a few weeks. PFM should be differentiated from dermatomyositis, parasthesia due to hypocalcemia, other myopathies, early juvenile idiopathic arthritis of the systemic onset type, and benign post-infectious childhood myositis. The contrast between the extreme severity of pain and tenderness in patients with PFM and the findings of normal CPK and subtle, non-specific EMG changes is striking (3). In some studies patients suffering



Fig. 2. T1-weighted axial image of the calf. TR 672, TE 15, foV 20. Subcutaneous hypointense linear signal changes due to oedema, limited hyperintensity was noted on the distal part of the medial gastrocnemius muscle.

from PFM have relatively high rate of three mutations: V726A, M694V, and E148Q (4). In a series of 35 children with FMF-associated vasculitis only 3 were found to be homozygotes (5). Three mutations (M694V, V726A, M608I) were found in 85% of Turkish FMF patients in this same study. Since this is the first case with MRI findings of the involved muscles, we felt it valuable to be presented.

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