Magnetic Resonance Imaging of Muscular Involvement in Juvenile Dermatomyositis: Case Report

Alicioglu B.[1], Karakas H. M.[2], Firat A. K.[2]
[1]Department of Radiology, Trakya University Medical Faculty, Edirne, Turkey;
[2]Department of Radiology, Turgut Ozal Medical Center, Inonu University Medical Faculty, Malatya, Turkey

Received September 9, 2008; Accepted December 31, 2008.

Key words: Dermatomyositis – Magnetic resonance imaging

Mailing Address: Assist. Prof. Dr. Banu Alicioglu, Fatih mah. 4.cd 43.sok Ziraatliler sit B Blok No: 8, 22030 Edirne, Turkey; Phone: +902 842 363 089; Fax: +902 842 352 730; e-mails: banualicioglu@trakya.edu.tr, banu_alici@yahoo.com

Abstract: A case of 5 years old juvenile patient with dermatomyositis together with conventional and diffusion-weighted MRI images is reported. The reported case was atypical for the pattern of involvement of the accompanying oedema which was affecting distal parts of the extremities and anterior muscle groups. Electromyography was negative. The case confirmed by a pathologist demonstrates the use of diffusion weighted imaging in determining unequivocal oedema and its exact extent.

Introduction

Juvenile dermatomyositis is a rare childhood disorder. However, it is the main primary inflammatory muscle disease in that age group [1]. There are several studies on clinical, laboratory and radiological findings of that disease [2–5]. However, its diffusion-weighted (DW) magnetic resonance images (MRI) have not yet been reported.

In the present study, a case of juvenile dermatomyositis is reported with a special emphasis on conventional and diffusion-weighted MRI characteristics. The case was atypical for the pattern of involvement of the accompanying oedema affecting distal parts of the extremities and the anterior muscle groups.

Case

A five years old boy was hospitalised with a right calf pain lasting for 10 days. The pain increased throughout the course of the disease. Walking and fatigue caused an aggravation of the symptoms. He was unable to climb stairs, run or even stand-up without assistance. In physical examination a very slight heliotrophic rash over lower extremities was determined. He had difficulty in standing up, which took approximately 3 seconds. However, he was able to walk on the tips of his toes and Babinski reflex and/or clonus were not present.

Laboratory tests showed creatine kinase (CK) and creatine phosphokinase (CPK) significantly increased (1905 UI and 2200 UI, respectively; normal values of CK and CPK: 25–200 U/L). Lactate dehydrogenase was also elevated. However, electromyography was normal. Physical examination of the abdomen and abdominopelvic ultrasonography did not reveal any abnormality.

MRI of thigh was performed to reveal the presence of a possible muscular involvement and its extent. Conventional T1 weighted turbo spin echo (TSE), T2 weighted TSE and short tau inversion recovery (STIR) sequences were used. Additionally, DW images were obtained since they were known to be sensitive to the restriction of random movements of water molecules. These images were acquired to delineate the full extent of oedema. In STIR images, an increase of pathological signal was seen in the lower extremities. This signal was more severe in thigh muscles and more widespread in leg muscles (Figure 1). Radiological findings were indistinct in conventional T2 weighted images. In postcontrast images, mild to moderate enhancement was noticed in muscle groups of the

presented locations (Figure 2). Enhancement was more prominent in dorsal muscles (Figure 3). Pathological muscles had hyperintense imaging characteristics in DW images (Figure 4), whereas they were hypointense in apparent diffusion coefficient (ADC) maps (Figure 5). DW images and ADC maps had shown an additional extension of oedema to ventral muscular groups when compared to conventional sequences.

The presence of heliotrophic rash, symmetrical muscular weakness and prominence of extremity oedema more proximally implicated dermatomyositis. Incisional muscle biopsy was performed to obtain a conclusive diagnosis. Microscopic analysis revealed the presence of degenerated and necrotic fibers that were dispersed between atrophic muscle fibers. Oxidative enzymatic and ATPase staining were not specific.

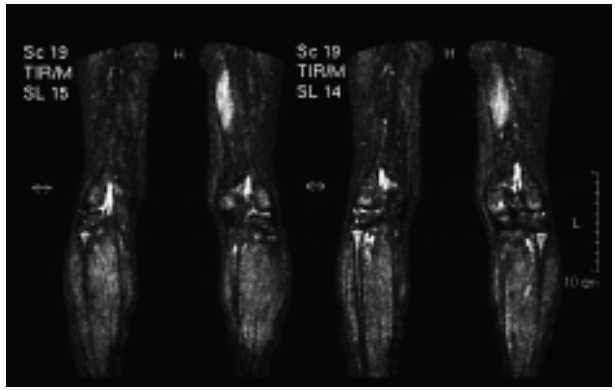

Figure 1 – Coronal STIR images of lower extremities: Increased signal intensity was more severe in thigh muscles whereas it was more widespread in leg muscles.

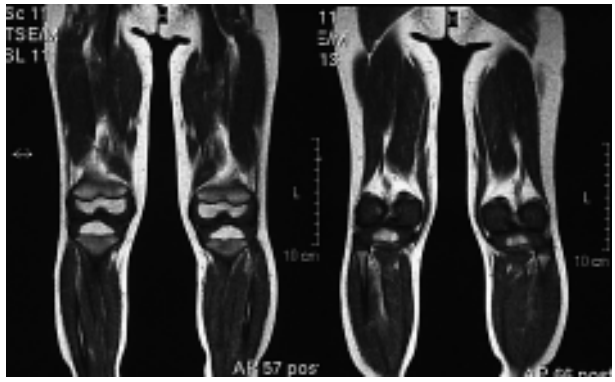

Figure 2 – Patchy enhancement of pathological muscles in coronal native (A) and contrast enhanced (B) T1 weighted images.

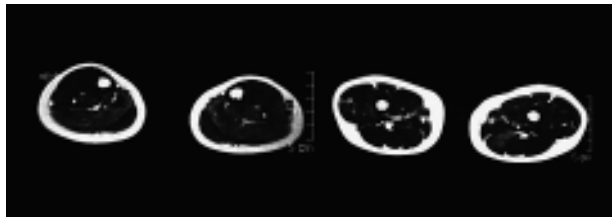

Figure 3 – Patchy enhancement in dorsal muscle groups in axial contrast enhanced T1 weighted images.

In immunohistochemical evaluation, spectrine, merosine, dystrophine, and alpha, beta, delta and gamma sarcoglycans were found to be positive. Additionally, human MHC class II molecules that trigger immune response related cellular processes were increased in cellular membranes. All these findings were compatible with inflammatory myopathies.

Discussion

Inflammatory myopathies represent a spectrum of disorders changing from focal diseases affecting a single muscle or muscle group to diffuse diseases with extensive muscular involvement. These diseases are etiologically classified under two headings named as immune-related or infective myopathies [6, 7].

Dermatomyositis, polymyositis and inclusion body myositis that constitutes inflammatory myopathies have differential clinical and histopathological features. These diseases manifest either as solitary disorders or they accompany systemic connective tissue diseases, other autoimmune disorders, retroviral infections or malignancies [8]. Ooedema is the paramount clinical feature. It is usually confined to proximal part of the extremities [9]. Widespread ooedema, on the other hand, is not very common [10, 11]. The presented case was rare due to the occurrence of widespread oedema in distal parts of the patient's extremities. Myopathies, as seen here, can have atypical radiological presentations.

Autoimmune mechanisms play an important role in the aetiology of infantile dermatomyositis and the disease therefore affects many organ systems. Patients suffer from proximal muscular weakness developing through weeks or months. However, in early childhood patients may not be able to express their problems, and inattentive parents can miss early changes and spatiotemporal evolution of the disease. In some additional cases, dermal inflammation, the main accompanying feature, can be only delicate and may therefore go unnoticed. In those situations, and probably in the reported case, patients can present in a "pseudo-acute" state. Only the characteristic dermatological

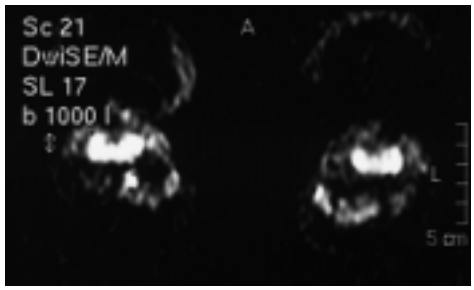

Figure 4 – Extension of muscular oedema to ventral muscles in axial diffusion weighted images. This finding was not noticed in conventional MRI.

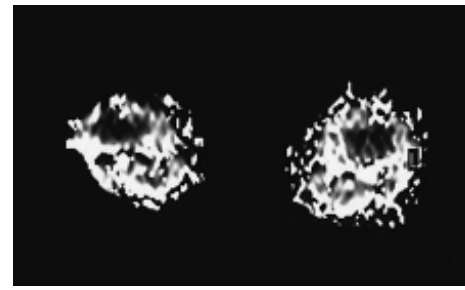

Figure 5 – Corresponding ADC map to Figure 4 shows hypointense changes compatible with pathological oedema.

findings, symmetrical weakness of proximal muscles, and the elevation of muscle enzymes can help to diagnose dermatomyositis. Nevertheless, a conclusive diagnosis requires electromyographical confirmation and a positive muscle biopsy [12–15].

MRI can help to reveal the presence of inflammatory myopathy, to describe its subtype and to define the potentially most useful biopsy site [2]. In patients with polymyositis or dermatomyositis the characteristic finding represent a focally or diffusely increased signal of proximal muscle groups in T2 weighted or fat-suppressed T2 weighted (STIR) images [3–5]. These features were present also in the reported case. However, he had an additional feature in the form of extremity oedema in distal parts. DW imaging, proved the presence of oedema in that case with a normal electromyography. DW images also showed the severity of the oedema in proximal muscle groups and the involvement of ventral muscle groups.

Juvenile dermatomyositis is a rare disease. However, in its classical manifestation it can be recognized according to its clinical and radiological features in the skin and in the proximal parts of extremities. However, the less typical manifestation can cause diagnostic controversies. MRI, assisted with DW images, can reveal the involved sites with high sensitivity, show the degree of severity, and can solve the diagnostic controversy. These images can be also used to determine the most severed site for the MR oriented or guided muscle biopsy.

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