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CASE REPORT

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Occult myopathy of the vastus intermedius muscles detected by magnetic resonance imaging in subclinical dermatomyositis: report of two cases

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Abstract The thigh muscles of two patients with dermatomyositis (DM) without muscle weakness or conspicuous creatine kinase elevations were studied by magnetic resonance imaging (MRI). Myositis limited to the vastus intermedius muscles (VIM) was detected in both patients, and in one, the diagnosis was confirmed by the findings of a biopsy specimen. Focal myositis of the VIM in early-stage DM, which otherwise would remain hidden by the relatively small muscle size and deep location, can be detected by MRI.

Key words Amyopathic dermatomyositis (ADM) · Dermatomyositis (DM) · Magnetic resonance imaging (MRI) · Vastus intermedius muscle (VIM)

Introduction

Dermatomyositis (DM) is diagnosed from the presence of characteristic skin involvement, proximal muscle weakness, muscle enzyme elevation in the serum, findings in electromyograms (EMG) and muscle biopsy specimens, and the detection of serologic markers.[1,2] However, some of these signs are not fully evident in the early stage of the disease because the affected muscles might be small and/or deeply situated, which limits access for EMG or muscle biopsy.

Some recently proposed diagnostic criteria for DM[3] include characteristic findings in magnetic resonance imaging (MRI) as an additional criterion that can be substituted for proximal muscle weakness or elevated muscle enzymes.

We describe the two patients with DM in an early stage who had occult muscle involvement which was limited to the deeply situated vastus intermedius muscle (VIM) in the thighs. This was detected successfully by MRI.

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Case reports

Case 1

On October 11, 2000, a 62-year-old Japanese man presented to our hospital with polyarthralgia, painful small skin ulcers of the fingertips, Gottron's sign, and atypical periorbital heliotrope discolorations. Neither Raynaud's phenomenon nor pulmonary problems were apparent. A physical examination did not demonstrate proximal muscle weakness, pain elicited by grasping the muscles, or sclerodermatous skin changes. Serum creatine kinase was only slightly elevated (265 IU/l; normal 25–200 IU/l). Autoantibodies, including antinuclear antibodies (ANA), anti-Jo-1 antibodies,[4] and anti-Mi-2 antibodies,[5] were all absent. The patient was diagnosed with possible amyopathic DM (ADM), or DM at a subclinical stage. Subsequently, however, high signal intensities in left thigh within the VIM were seen in T2-weighted MRI. No abnormalities were seen in T1-weighted images (Fig. 1). An electromyogram (EMG) recorded from the VIM showed a myopathic pattern. Muscle biopsy specimens showed muscle fiber necrosis, phagocytosis, and regenerating myocytes compatible with DM (Fig. 2) in samples from the VIM, but not in samples from the overlying rectus femoris muscle, which appeared normal by MRI. Steroid therapy (prednisolone, 30 mg/day po) resulted in an immediate improvement of joint and skin abnormalities. During 7 months of postdischarge follow-up, no muscle weakness or muscle enzyme elevation has developed. The patient was recently readmitted for rapidly progressive interstitial pneumonia; this was treated successfully with a combination of simulta-

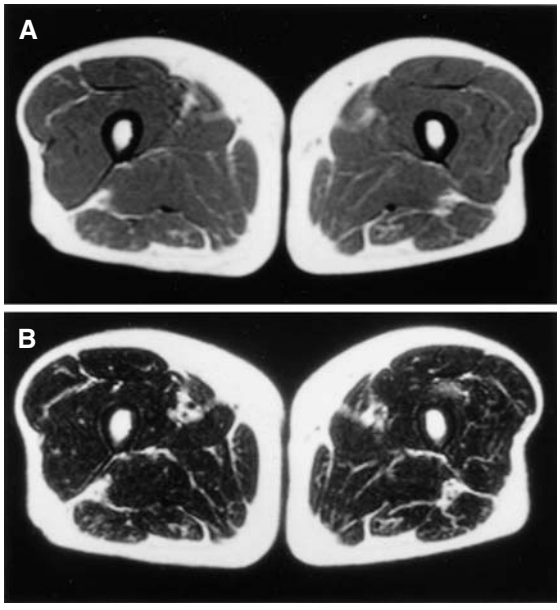

Fig. 1. Magnetic resonance imaging of thigh muscles in patient 1. A normal T1-weighted image (**A**) contrasts with high signal intensities in a T2-weighted image (**B**) of the left vastus intermedius muscles

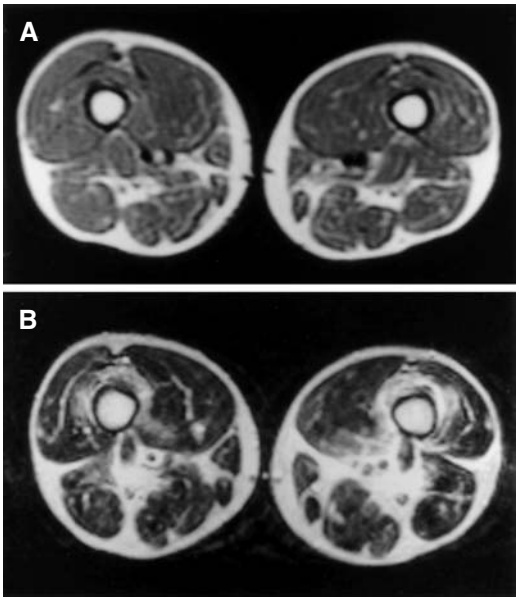

Fig. 3. Magnetic resonance imaging of the thigh muscles in patient 2. A normal T1-weighted image (**A**) and a T2-weighted image (**B**) with high signal intensity in both vastus intermedius muscles are shown

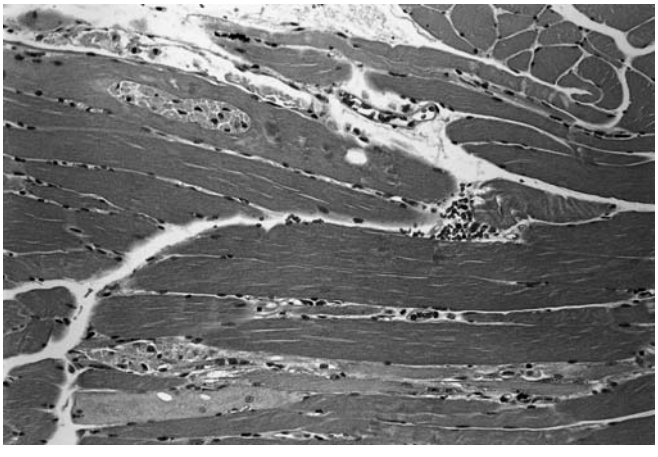

Fig. 2. Findings in a muscle biopsy specimen from the vastus intermedius muscle in patient 1. Muscle fiber necrosis, phagocytosis, and regenerating myocytes can be seen, although lymphocytic infiltration is not seen

neously administered high-dose intravenous γ-globulin, cyclosporine, and pulse steroid therapy.

Case 2

A 40-year-old Japanese man presented in December 1999 with polyarthralgia, Gottron's sign, and atypical heliotrope discolorations with periorbital edema. Raynaud's phenomenon, dysphagia, and dyspnea on exertion were not present. On admission, a physical examination revealed active synovitis involving peripheral joints. In addition, crepitant rales were noted bilaterally in the lung bases. Muscle weakness, pain elicited by grasping the patient's muscles, and sclerodermatous skin changes were all absent. His serum creatine kinase concentration was 151 IU/l. An EMG of the vastus lateralis muscle showed no myopathic abnormalities on either side. Autoantibodies, including ANA, anti-Jo-1, and anti-Mi-2, were all undetectable. Roentgenography, computed tomography, and gallium scintigraphy of the chest all indicated interstitial pneumonia. The patient was diagnosed with possible ADM, or DM at a subclinical stage, including arthritis and interstitial pneumonia. Subsequent examination of the thigh muscles by MRI showed a high signal intensity localized to the VIM, and present only in T2-weighted images (Fig. 3). Pulse steroid therapy was effective in treating all clinical abnormalities, and the patient was discharged on June 15, 2000. No muscle weakness or muscle enzyme elevation had become apparent at the most recent follow-up assessment, which was 14 months after discharge.

Discussion

DM at the early stage considered here has been variously termed, DM without muscle involvement,[6] DM without creatine kinase elevation,[7] and ADM showing typical skin rashes, but no concurrent muscle involvement.[8,9] However, MRI now enables us to detect hidden myopathy in cases such as those described here.

Among the four quadriceps femoris muscles, the vastus lateralis, vastus medialis, and VIM have been reported to be affected in early DM, but not the rectus femoris. Like the rectus, other flexor muscles are not involved at this early stage. VIM involvement appears to be the initial quadriceps component in DM. As both of our DM patients were seronegative men with interstitial pneumonia and arthritis,

selectivity in the muscles affected might depend on disease subsets. Further studies in a larger number of patients might resolve this issue of whether patients with mild or inapparent myopathy, but overt interstitial pneumonia and arthritis, indeed constitute a new subset of DM.

Our findings support the impression that a high signal intensity in T2-weighted images of muscle, referred to as an "edema-like abnormality"[10] and occurring in the absence of any associated T1 abnormality, indicates the presence of acute myositis. Confirmatory evidence includes biopsy specimen findings in case 1. Furthermore, myopathy limited to the VIM could explain why muscle weakness and muscle enzyme elevation are not found in early-stage DM. In the same context, normal EMG and muscle biopsy findings are not infrequent when other muscles are sampled. In contrast to the absence of EMG and muscle biopsy findings in the MRI-negative rectus femoris muscle in case 1, the MRI-positive VIM showed EMG and histologic abnormalities. This site is not routinely examined by EMG or sampled by muscle biopsy.

In conclusion, the involvement of the VIM, which otherwise would remain undetected because of the small muscle size and deep location, may be detectable by MRI in early-stage DM.

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