Recurrent Focal Myositis of the Left Thigh– rare but treatable inflammatory myopathy - A case report

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Abstract

Focal myositis presents as an acute and localized inflammation of the muscle without systemic involvement. The etiology is unknown. Several case reports in the literature have described different presentations of the condition, the majority affecting the limbs with a spectrum of clinical progression, laboratory, and radiological findings.

It is often confused with infection, muscle tumor such as sarcoma, paraneoplastic syndrome or even immune-mediated polymyositis. Delay in diagnosis and inappropriate treatment may lead to recurrence of the condition.

We report a case of recurrent focal myositis of the left thigh in a man in whom optimal treatment commenced six months after the onset led to regression of his symptoms and signs.

Keyword: Focal myositis, muscle biopsy, magnetic resonance imaging.

Introduction

Focal myositis (FM) is a rare benign inflammatory myopathy of unknown etiology, characterized by the presence of pseudotumor in the skeletal muscle of the limbs commonly thighs and calf.1 Other differential diagnosis would be infective causes, deep vein thrombosis and soft tissue sarcomas. 2, 3, 4

Muscle biopsy and magnetic resonance imaging (MRI) are crucial in making the diagnosis of FM.

FM may be self-limiting or resolve spontaneously over time.1 However, it responds well to corticosteroids and non-steroid anti-inflammatory drugs and cases that progress may require immunosuppressive therapy.3

We describe a case with recurrent FM of the vastus medialis muscle of the left thigh. He responded well to treatment with corticosteroids and methotrexate. This has never been reported in the Malaysian literature.

Case report

A 50-year-old man, presented with a history of localized pain and swelling over his left thigh for the past 6 months. Initially he also noted redness of the overlying skin. There was no associated generalized muscle pain or arthritis. He had no history of trauma or injury, fever, rash, or other systemic involvement. He was unable to squat down due to the pain since then.

Systemic examination was unremarkable except for tenderness over the left quadriceps. The overlying skin was not indurated.

Laboratory investigations showed normal creatine-kinase (106 U/L), mildly elevated erythrocyte sedimentation rate (ESR) of 26 mm/hour and negative screening for HIV, hepatitis B and C. Immunological markers (antinuclear antibodies, rheumatoid factor, extra-nuclear antigen – anti-JO1, anti-U1RNP, anti-Ro and anti-La) were negative. Electromyography (EMG) of the left thigh was normal.

Muscle biopsy of the left vastus medialis was performed and showed a disarray of muscle bundles associated with fibrosis and interstitial inflammation. The perimysium was widened and cellular in some areas. The muscle fibers were variable in size but there was no necrosis noted. Scattered atrophy of muscle fibers with atrophic multinucleated giant cells were seen. The interstitial and fibrotic tissue showed increase lymphocytes and plasma cells and occasional eosinophils. There was an absence of immature fibroblasts, hemorrhage, osteocytes or myofibroblasts. Calcification or bony trabecular formation was not seen and there was no sign of malignancy seen. Immunohistochemical test was not performed.

MRI of the left thigh showed increased T2 signals over the lower belly and musculotendinous junction of vastus medialis conforming to a feathery pattern of inflammatory myositis. A smaller area of necrosis or degeneration was noted in the post contrast examination. There was edema of the overlying subcutaneous tissue. Similar signal alteration was also noted in the short head of bicep femoris with areas of necrosis. The overlying soft tissues was edematous. (Figure 1a, b)

He was given prednisolone 5 mg daily with non-steroidal anti-inflammatory medication (celecoxib 200 mg daily). However he showed no improvement and had several admissions for recurrent pain over the same site.

At six months, the pain had still not resolved. MRI was repeated and this showed improvement in the increased signals in the left vastus medialis. There was no mass-like lesion or necrosis seen. The earlier lesion seen in the short head of bicep femoris had resolved. (Figure 2a,b)

In view of recurrent episodes, prednisolone dose was increased to 20

mg daily in combination with methotrexate 10 mg weekly. His symptoms resolved within a few days of commencement of the higher dose of steroid.

Discussion

Focal myositis was first described by Heffner et al in 1977, as a clinical entity describing focal myositis as a benign inflammatory pseudotumor of the skeletal muscle.[1] Focal myositis of the thigh has been described in the literature involving quadricep femoris, distal leg group of muscles.[2,5] Clinical diagnosis of focal myositis is commonly supported by muscle biopsy which is confirmatory.[6] MRI scan is highly sensitive and diagnostic to localize the site of inflammation and mass.[6,7] It is also very useful to evaluate the outcome.

In our patient, non-specific myositis was diagnosed during his first presentation and this was confirmed by MRI scan and muscle biopsy. However, due to its rarity and lack of recognition of the condition, the corticosteroid dosage was suboptimal. The clinical history, muscle biopsy, MRI scan and laboratory finding were consistent with focal myositis. Absence of systemic involvement such as fever, generalized muscle weakness or other constitutional symptoms makes a diagnosis infection and polymyositis to be very unlikely. Typically in polymyositis ESR, CRP or CK will be raised and immunology markers such as the anti-JO1 autoantibody may be positive. However, there are cases of polymyositis reported in the literature involving distal muscles or with normal CK levels.[8] Immunohistochemical test of the muscle biopsy may show presence of CD4+ T cells in focal myositis[9] and dermatomyositis[10], but not in polymyositis.[11]

Despite a very low dose of prednisolone 5 mg daily, the repeat MRI scan after 6 months showed the lesion has resolved partially. In many previous case reports, focal myositis may achieve spontaneous remission over time with corticosteroid and NSAIDs, even in relapsed cases.[12] The use of immunosuppressive agents such as methotrexate for a steroid sparing effect has been shown to be effective in refractory cases.[13, 14]

It is unlikely for the focal myositis in this patient to progress to polymyositis based on normal laboratory markers (ESR, CRP, CPK) and regression of the symptoms. The MRI scan has showed only single muscle group involvement. However, long-term follow-up is needed to monitor any progression of the condition.

In conclusion, patients with focal myositis have a good prognosis and relapse can be prevented if diagnosed early and appropriate treatment is given.

Conflict of interest: Authors has no conflict of interest.

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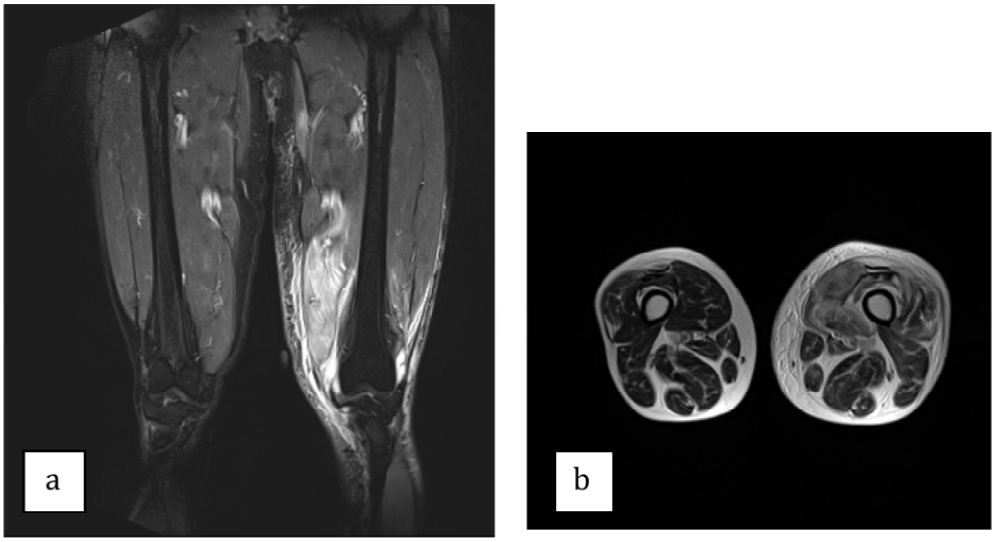

Figure 1. A coronal and axial view of T2 weighted magnetic resonance imaging – high signal intensity in vastus medialis of the left thigh suggestive of muscle inflammation.

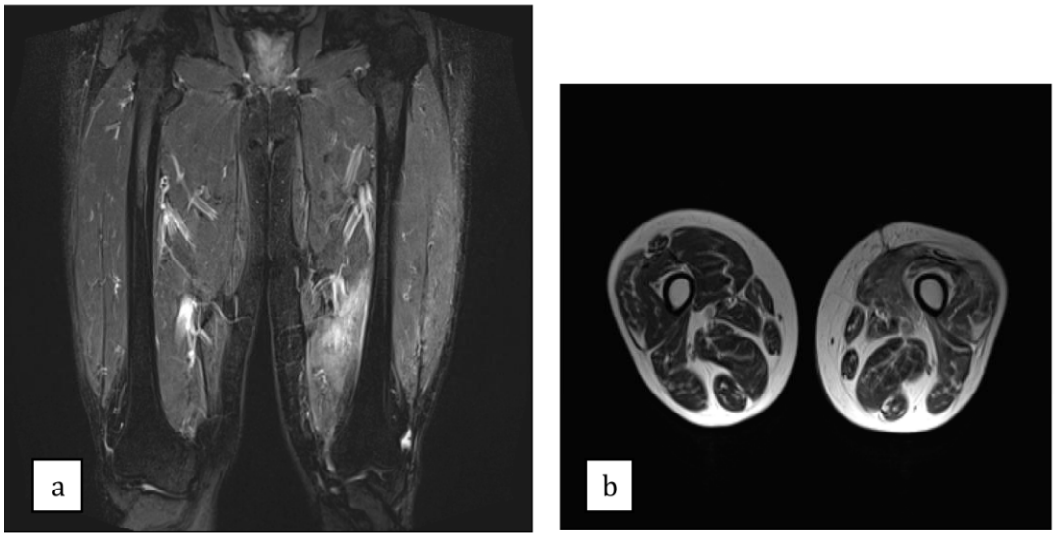

Figure 2. A repeated T2 weighted magnetic resonance imaging 6 months later showing resolution of the signal intensity in vastus medialis of the left thigh.