Utility of MRI Findings in Inflammatory Myopathies

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Introduction

MR imaging can provide important information for establishing a diagnosis in inflammatory myopathies, and routine MRI sequences can help exclude alternative diagnoses. With MRI, muscle pathologic findings typically fall into three categories: mass lesions, fatty atrophy, and edema [1]. Masses can include muscle injury with hematoma or myositis ossificans, sarcoma or pseudosarcoma, abscess, parasitic infection, or sarcoidosis. Fatty infiltration and atrophy is the end-stage result of many muscle pathologies, including disuse, myopathy, tendon injury, or denervation. Muscle edema is the third muscle pathology commonly encountered with clinical MR imaging. Muscular edema can be caused by muscle injury, denervation, drug-induced myopathy, or inflammatory myopathies including paraneoplastic syndromes. The focus of this article is to present several cases of muscle edema to demonstrate the utility of MR imaging in simplifying the differential diagnosis based on the distribution of findings and clinical history.

Case 1

A 37-year-old male rancher developed pain, weakness, and slight stiffness in his left thigh one month previously and noticed a mass. He denied any injury or constitutional symptoms. On his initial examination, he was afebrile with mild swelling of his left thigh with tenderness and a palpable mass. There was no redness or warmth to touch. He showed 4/5 strength with extension, but there was no atrophy of the left thigh. His initial radiographs were negative. His initial MRI showed extensive edema within the vastus medialis and intermedius muscles with a central mass with low intensity rim (Fig. 1A-C). Follow-up MRI three months later, showed decreased muscular edema and development of a discreet mass with low signal intensity rim (Fig. 1D-F).

Diagnosis: Myositis Ossificans

Myositis ossificans is a benign, self-limited condition that presents following an injury to muscle [2]. The presenting clinical scenario may mimic muscle denervation or myopathy and since patients frequently do not recall an inciting event. The initial imaging may also be confusing, especially on MRI, given the intramuscular inflammatory mass-like appearance and lack of zonal peripheral mineralization. In this particular case, there is no initial muscle abnormality visible on the T1 sequence (Fig. 1A), but rim of low signal intensity on the fluid sensitive sequence indicates an underlying mass (Fig. 1C), which in some cases is only appreciated after subsequent imaging [2]. A mass usually develops within 6-8 weeks and extensive edema has been described in lesions that are imaged within 8 weeks of the inciting event [2]. Clinical and imaging follow-up can confirm the diagnosis (Figure 1D-F). The ossific rim usually appears after 4-6 weeks, but early in its formation may only be visible by CT [2]. Post-contrast imaging will demonstrate enhancement due to its extensive vascularity [2]. Biopsy should be avoided, especially early (within 1-2 weeks) when the mass could potentially be confused upon histology with extraskeletal osteosarcoma resulting in aggressive therapies for an inherently benign condition [3].

Case 2

A 56-year-old female with renal cell carcinoma developed right lower extremity weakness, primarily with extension following radical right nephrectomy, retroperitoneal lymph

Key Points

Inflammatory myopathies encompass several rare diseases that affect the lower extremities greater than the upper extremities and include inclusion body myositis, polymyositis, and dermatomyositis. The presence of inflammatory myopathy must be distinguished from other potential causes of muscle pathology, including muscle injury, nerve injury, rhabdomyolysis, tumor and diabetic ischemic myopathy. MR imaging can help to exclude other pathologies and can provide important information about the location and extent of inflammation. For cases in which a specific diagnosis by MRI alone may not always be possible, MRI may provide value in localizing the greatest areas of inflammation to focus muscle biopsy, since treatments may differ depending on the underlying etiology.

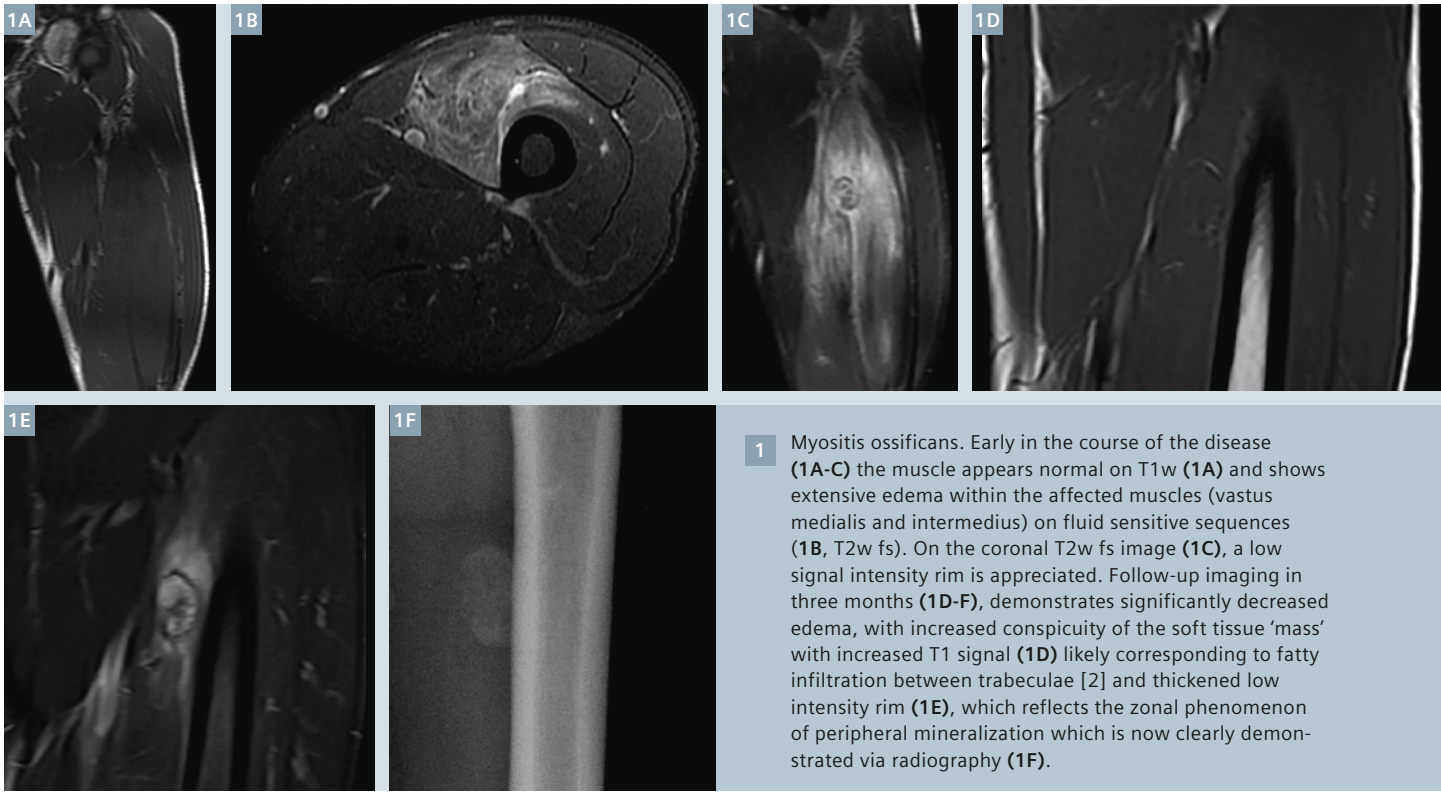
node dissection, and inferior vena cava thrombectomy, which was complicated by iliopsoas and paraspinal hematoma. Five months following surgery she was evaluated and found to have 0 out of 5 right quadriceps muscle strength compared to 5 out of 5 strength within the extensor hallucis longus, tibialis anterior, gastrocnemius, and soleus muscles. Pelvic MR imaging was performed and demonstrates unilateral muscle edema within the quadriceps musculature, specifically the rectus femoris and vastus lateralis muscles (Fig. 2).

Diagnosis: Femoral Nerve Injury
MR imaging can help distinguish a nerve injury edema pattern from myopathy. Most commonly, MR imaging of denervation is unilateral and demonstrates edema and possibly atrophy of the musculature within a specific and typical distribution of the affected nerve. Although electromyography (EMG), nerve conduction studies, or ultrasound can be used to localize nerve injury, MR imaging has the advantage of greater sensitivity, non-invasiveness compared to EMG, superior anatomic resolution, and in some cases allows identification of aberrant nerve supply [4].

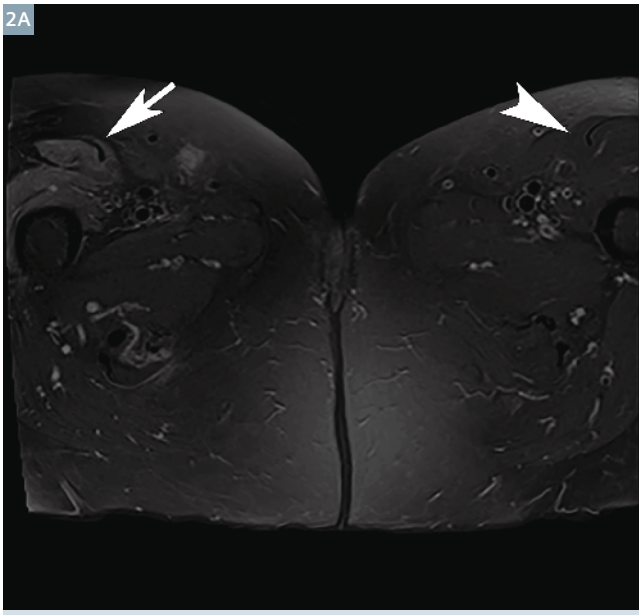
Case 3

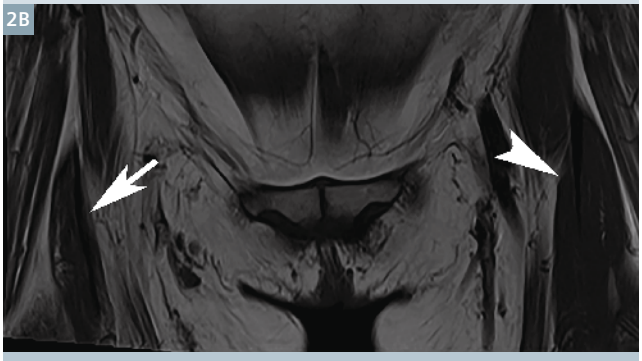
A 58-year-old female who presented with a 12-day rash that initially started on her chest and back and spread to her neck, face and forehead. She revealed progressive muscle weakness over the course of the past year primarily within her shoulders and hips manifest by increasing difficulty brushing her teeth and climbing stairs. She had been treated with a steroid taper without significant improvement of her rash or her muscle weakness. On examination, she was afebrile with an erythematous rash over her upper, back, chest, and neck with macules and papules over her lateral thighs and elbows, but no rash over her eyelids. She was unable to lift her arms off the bed and had 3 out of 5 muscle strength with knee extension and mild decreased grip strength. She had an elevated creatine kinase at 678 (units/liter; reference range 20-180 U/l) with a normal erythrocyte sedimentation rate and C-reactive protein. Tests for anti-nuclear antibody, lyme disease, and coccidiomycosis were negative. An MRI of her thighs demonstrated relatively symmetric muscle edema within her knee extensors (Fig. 3). Initial muscle biopsy of her left vastus lateralis was negative, while subsequent biopsy of her right semitendinosus muscle revealed perivascular inflammation.

Diagnosis: Dematomyositis
Dermatomyositis is an autoimmune inflammatory myopathy that results in subacute symmetric proximal muscle weakness. There is a predilection for women and age can vary, but has a peak between 30 and 50 years of age [5]. The differential diagnosis of muscle weakness includes other inflammatory myopathies, such as polymyositis and inclusion body myositis. In this particular patient the presence of a rash favors dermatomyositis over polymyositis and inclusion body myositis, however it is important to exclude inclusion body myositis, as this disease does not respond to most therapies, including steroids [6, 7], as reportedly occurred in the short term with this patient. The disease is characterized by an erythematous rash and often has Gottron's papules over extensor surfaces as in this patient over the elbows. They frequently also have a heliotrope rash characterized by erythematous eyelids, which this patient did not have.

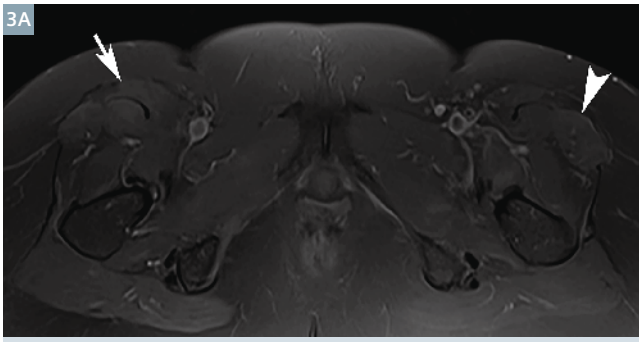


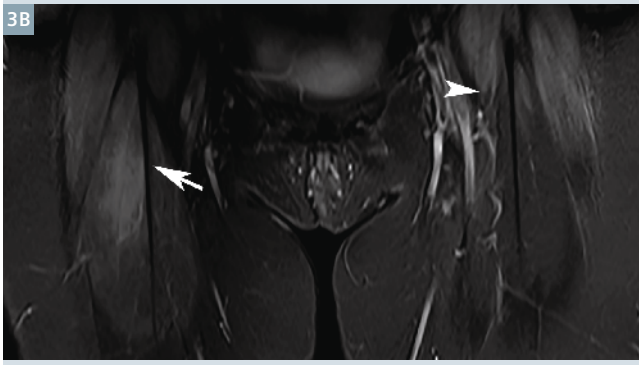
1 Myositis ossificans. Early in the course of the disease **(1A-C)** the muscle appears normal on T1w **(1A)** and shows extensive edema within the affected muscles (vastus medialis and intermedius) on fluid sensitive sequences (**1B**, T2w fs). On the coronal T2w fs image **(1C)**, a low signal intensity rim is appreciated. Follow-up imaging in three months **(1D-F)**, demonstrates significantly decreased edema, with increased conspicuity of the soft tissue 'mass' with increased T1 signal **(1D)** likely corresponding to fatty infiltration between trabeculae [2] and thickened low intensity rim **(1E)**, which reflects the zonal phenomenon of peripheral mineralization which is now clearly demonstrated via radiography **(1F)**.

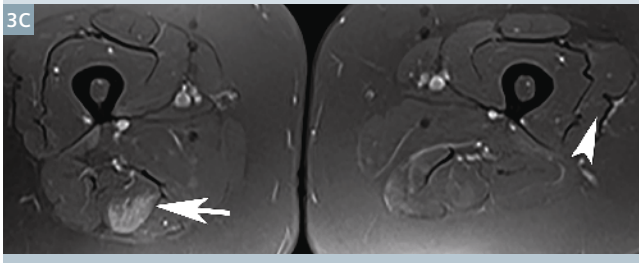




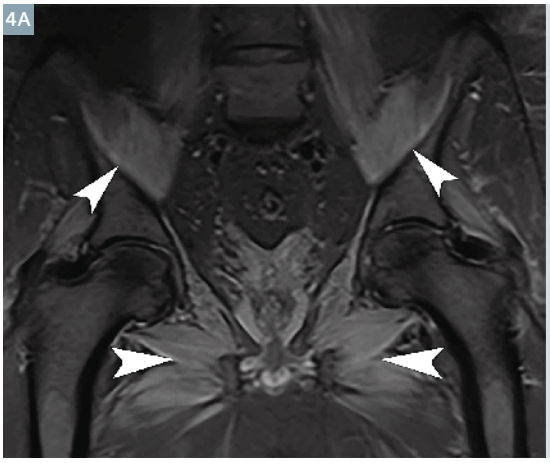
2 Right femoral distribution denervation. Five months following right nephrectomy for renal cell carcinoma, the patient has edema within the rectus femoris and vastus lateralis muscles (arrow in **2A**; T2w fat suppressed axial image) as well as mild atrophy (arrow in **2B**; T1w coronal image).

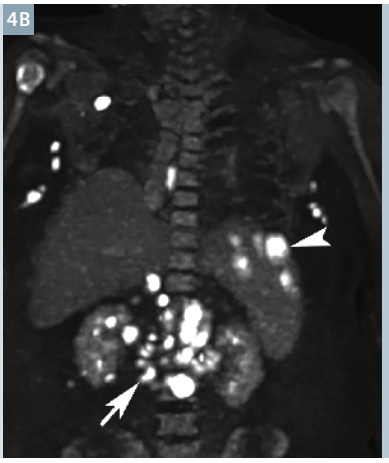






3 Dermatomyositis. Axial proton density with fat suppression **(3A)** and coronal STIR **(3B)** images demonstrate bilateral muscle edema in the rectus femoris and tensor fascia lata muscles (arrow on right side and arrowhead left side). Distal axial proton density-weighted image with fat suppression demonstrates asymmetric involvement of the right greater than left semitendinosus muscle (arrow) and no involvement of the left vastus lateralis muscle (arrowhead), which is consistent with the negative biopsy result performed at this location.





4 Polymyositis with lymphoma. Coronal STIR image **(4A)** of the pelvis demonstrates symmetric edema in multiple muscle groups (arrowheads). Maximum intensity projection (MIP) image from subsequent PET/CT scan **(4B)** demonstrates FDG uptake in extensive retroperitoneal lymphadenopathy (arrow) and splenic uptake (arrowhead) from lymphomatous infiltration.

MR imaging

MRI can be helpful in confirming the presence of myopathy, excluding non-inflammatory causes of muscle disease and, in some cases, differentiation between inflammatory myopathies. Ultimately, the definitive diagnosis is made through muscle biopsy. The distinguishing features of the inflammatory myopathies by histology include perivascular inflammation with dermatomyositis, endomysial inflammation with polymyositis, and both the presence of inclusion bodies and endomysial inflammation in inclusion body myositis [5, 7-10]. Theoretically, this would translate into differences in MR imaging, but in practice the edema and enhancement patterns in individual muscles overlap, especially between dermatomyositis and polymyositis. Dermatomyositis, with its perivascular inflammation, tends to have more perifascial muscle edema than polymyositis, which occurs more frequently in the juvenile form of the disease, but nevertheless is not diagnostic [11-13]. However, one distinguishing feature of inclusion body myositis by MR imaging is the relative sparing of the rectus femoris muscle early in the disease process and the predominant involvement of flexor digitorum profundus in the forearm [14, 15]. In this case, which is relatively early in the disease process based on the relative paucity of muscle atrophy, there is involvement of the rectus femoris muscle suggesting that this is not inclusion body myositis, despite a history of lack of response to steroids.

Case 4

A 59-year-old male presented with a one-month history of proximal muscle weakness, arthralgia, fevers, weight loss, and truncal rash, along with progressive shortness of breath. He underwent a laboratory workup and was found to be anemic with elevated c-reactive protein and ESR and a positive ANA. His SPEP was negative. He was initially put on steroids with a presumed lupus diagnosis, which helped his weakness. A subsequent elevated LDH and lymphadenopathy by computed tomography as well as a negative bone marrow biopsy prompted a referral to an oncologist. He then underwent a lymph node biopsy and MRI of the pelvis for muscle weakness (Fig. 4).

Diagnosis: Polymyositis with lymphoma

MR imaging revealed a highly symmetrical myopathy without significant perifascial edema, which was confirmed polymyositis by biopsy. Whenever the diagnosis of polymyositis or dermatomyositis is made, an occult malignancy should be considered [10, 16]. Malignancy associated with these inflammatory myopathies is defined as a malignancy diagnosed within 3 months of the diagnosis of inflammatory myopathy and occurs in 3-6% of polymyostis and 13-29% of dermatomyositis [10, 16]. Despite the relatively high association with underlying malignancy and likely paraneoplastic etiology, there is no current consensus regarding a standard malignancy workup for this patient population.

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