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IMAGING (D MINTZ, SECTION EDITOR)

Diagnostic Imaging of Inflammatory Myopathies: New Concepts and a Radiological Approach

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Abstract

Purpose of Review The purpose of this review article is to highlight the current role of diagnostic imaging in the assessment of inflammatory myopathies.

Recent Findings Recent research demonstrates that imaging plays an important role in evaluating patients with symptoms of an inflammatory myopathy. In general, MRI is the pivotal imaging modality for assessing inflammatory myopathies, revealing precise anatomic details because of changes in the signal intensity of the muscles. Whole-body MR imaging has become increasingly important over the last several years. US is also a valuable imaging modality for scanning muscles.

Summary Together with the clinical history, familiarity with the imaging features of inflammatory myopathies is essential for formulating an accurate diagnosis.

Keywords Inflammatory myopathy · Magnetic resonance imaging · Whole-body magnetic resonance imaging · Ultrasound

Introduction

Inflammatory myopathies (IM) are the largest group of potentially treatable myopathies in both children and adults. These myopathies constitute a heterogeneous group of disorders that are best classified on the basis of distinct clinicopathological features and according to clinical, laboratory, and histological data. The most recognized types of IM are dermatomyositis (DM), polymyositis (PM), immune-mediated necrotizing myopathy (INM), sporadic inclusion body myositis (sIBM), and overlap myositis (including antisynthetase syndrome) [1••].

This review will discuss these, plus toxic and drug-induced myopathies and eosinophilic fasciitis.

Its purpose is to clarify the current role of imaging studies in the assessment of inflammatory myopathies, focusing on the typical imaging findings.

This article is part of the Topical Collection on *Imaging*

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Role of Imaging Studies in Myopathies

MRI is the pivotal imaging modality for assessing inflammatory myopathies. It is the most sensitive imaging test for detecting their musculoskeletal abnormalities and is able to define the extent and distribution of disease [2, 3].

The MR protocols should always include both T1 and a fluid-sensitive sequence, such as T2-weighted fat-suppressed sequences or short-tau inversion recovery (STIR) [4, 5] and MR imaging also helps to localize the best site for initial muscle biopsy. Longitudinal studies evaluate disease evolution and assess treatment response [6].

Muscle edema pattern and fatty infiltration are nonspecific manifestations of myopathy and neuromuscular disorders [7, 8] and can be identified in many pathologies [9]. The distribution and patterns of muscle involvement may suggest specific diseases (Table 1).

In order to assess the entire muscle anatomy, whole-body MR imaging (WBMRI) has become an important tool over the last years [10–12], revealing specific global distribution patterns of certain subtypes of myopathies and also detecting clinically silent involvement of certain muscle compartments [13, 14].

Whereas generally, MRI imaging is the most appropriate modality for evaluating patients suspected of myopathies [7, 15••], ultrasound (US) is also a valuable imaging modality for studying the musculoskeletal system with some advances of a shorter examination time, cost effectiveness, and the ability to directly correlate with the patient's clinical history and perform dynamic maneuvers [16].

Some limitations of US are that it can underestimate the extent of soft tissue edema, including muscle edema, identified on MRI; and it does not assess the entire musculoskeletal system, as can total body MRI exams. Additionally, US falls short of evaluating the deep musculature, and it is generally accepted that the soft tissue contrast of US is inferior to that of MRI [17].

Polymyositis

Clinical and Histological Aspects

PM occurs more frequently in women and in adulthood, being rare in childhood. Clinically, PM manifests with symmetrical and progressive proximal muscle weakness of variable intensity, an increase serum creatine phosphokinase (CK), and electromyographic changes, which develop in weeks to months. PM remains a diagnosis of exclusion, and other conditions, such as DM, IMNM, overlap myositis, and sIBM, should be excluded [18]. Oropharyngeal involvement and dysphagia are commonly observed. CK levels tend to increase by more than 10 times the reference value during periods of disease activity. A muscle biopsy demonstrates the presence of endomysial inflammatory infiltrate with the invasion of non-necrotic fibers in association with fibers in necrosis, as well as the presence of CD8 T cells and non-necrotic muscle fibers, which express major histocompatibility complex (MHC) class I antigen [19].

Imaging Findings

The most common pattern of magnetic resonance imaging is bilateral and symmetrical muscular edema in the pelvic girdle and thighs with preservation of the muscular architecture [8, 20] (Fig. 1). The signal intensity of the edema is directly proportional to the inflammatory status and the severity of the disease [8, 21].

Table 1 Main clinical and imaging characteristics inflammatory myopathies

Disease	Age	CK	Main muscles	Main finding	Symmetry	Edema pattern	Fat infiltration
Sporadic inclusion body myositis	> 50 years	+/++	Medial gastrocnemius Quadriceps Flexor digitorum profundus	Fat infiltration	Asymmetric	+/++	++++
Dermatomyositis and polymyositis	5–10 and ≈ 50 years	++++	Pelvic Girdle Thighs	Edema pattern	Symmetric	++++/along fascia	++/late stage
Immune-mediated necrotizing myopathy	Any	+++/++++	Lateral rotators Gluteus Medial and posterior thighs and Legs compartments	Edema pattern	Asymmetric	+++	+++
Toxic and drug-induced myopathy	Any	+++/++++	Gluteus Quadriceps Adductors Deep calf muscles	Edema pattern	Symmetric	++++	++

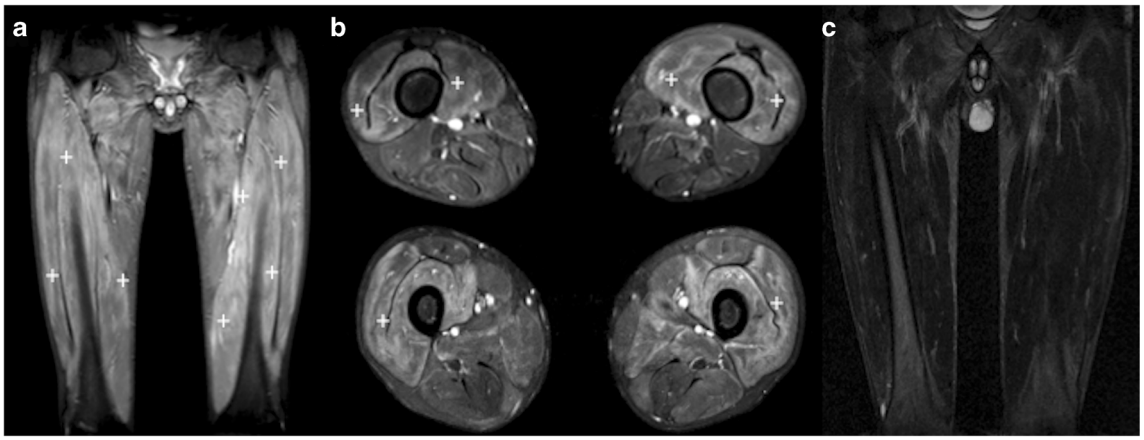

Fig. 1 Coronal and axial images STIR from the thighs of a 42-year-old woman with polymyositis before (**a**, **b**) and after treatment (**c**), showing response to interval therapy. In **a**, **b** shows extensive and diffuse edema pattern (+) in the thighs muscles, predominantly in the anterior compartments. In **c**, 6 months after treatment, no edema pattern are identified, showing a good treatment response

Because the edema in polymyositis most commonly affects the musculature of the thighs, local MRI has good sensitivity symmetrically. However, the whole-body magnetic resonance image (Fig. 2) is a more efficient method by performing an overall evaluation and detecting subclinical changes in different muscle groups [21]. Huang et al. [21] concluded that WBMRI had a higher positive rate than the serum creatine kinase test and EMG in the role of diagnosis.

In addition to corroborating with the diagnosis and excluding any other causes of myopathy, MRI may guide the best site for the biopsy [8, 22–24]. Nonguided muscle biopsies have false negative results in 10 to 25% of cases, and when the site with the highest inflammatory activity is found by muscle edema in STIR, MRI reduces this rate and avoids performing a new invasive procedure [23].

MRI still plays an important role in the follow-up of disease progression and in the analysis of response to treatment. In cases where there is a good response to drug therapy, muscle edema decreases with a reduction in the signal intensity of the affected muscle groups [23, 24] (Fig. 1). Tomasová et al. [24] showed that despite clinical and radiological improvement, the histological score does not change.

Dermatomyositis

Clinical and Histological Aspects

Patients with DM typically present with proximal muscle weakness, an increase in CK and cutaneous manifestations that develop over weeks to months [1••]. Some patients can develop cutaneous manifestations but little or no muscle involvement (hypomyopathic or amyopathic form). DM is more frequent in women and has a peak incidence in childhood (juvenile DM) and another in adulthood [1••]. Cutaneous involvement differentiates DM from other forms of inflammatory myopathies. The most characteristic skin features are heliotrope (violaceous and edematous periorbital rash) and Gottron's papules (erythematous lesions on the extensor surfaces of the joints) [25]. Other signs include Raynaud's phenomenon, ulcers, vasculitis, palmar hyperkeratosis, calcinosis, and periungual hyperemia. Some patients develop cardiac and respiratory involvement (interstitial lung disease) [25].

The muscle biopsy exam demonstrates the presence of perimysial and perivascular inflammatory infiltrate in association with atrophy and degenerative signs of the perifascicular muscle fibers. Cellular infiltration is predominantly plasmacytoid dendritic cells, B cells, CD4 T cells, and macrophages [26]. Class-1 major histocompatibility complex expression is increased preferentially at the perifascicular muscle fibers. Autoantibodies against Mi2 is frequently associated with DM [27].

Imaging Findings

In general, the MRI findings are similar in DM and PM. The most frequent findings are hypersignal areas in the STIR in the proximal musculature symmetrically, representing edema and inflammation [1••], and the muscular architecture preserved in the T1-weighted sequences during the early phases of the disease [8, 28].

However, there are macroscopic differences between DM and PM. Huang et al. [21] demonstrated that DM patients had patchy muscle edema and higher subcutaneous tissue edema, which typically mirrors the distribution of skin involvement; in contrast, PM patients had more diffuse muscle edema and the absence of subcutaneous involvement. Additionally, DM is more associated with the presence of calcinosis or calcifications of non-articular tissues (Fig. 3).

Regarding the distribution of muscle edema, DM often and symmetrically involves the femoral quadriceps, while

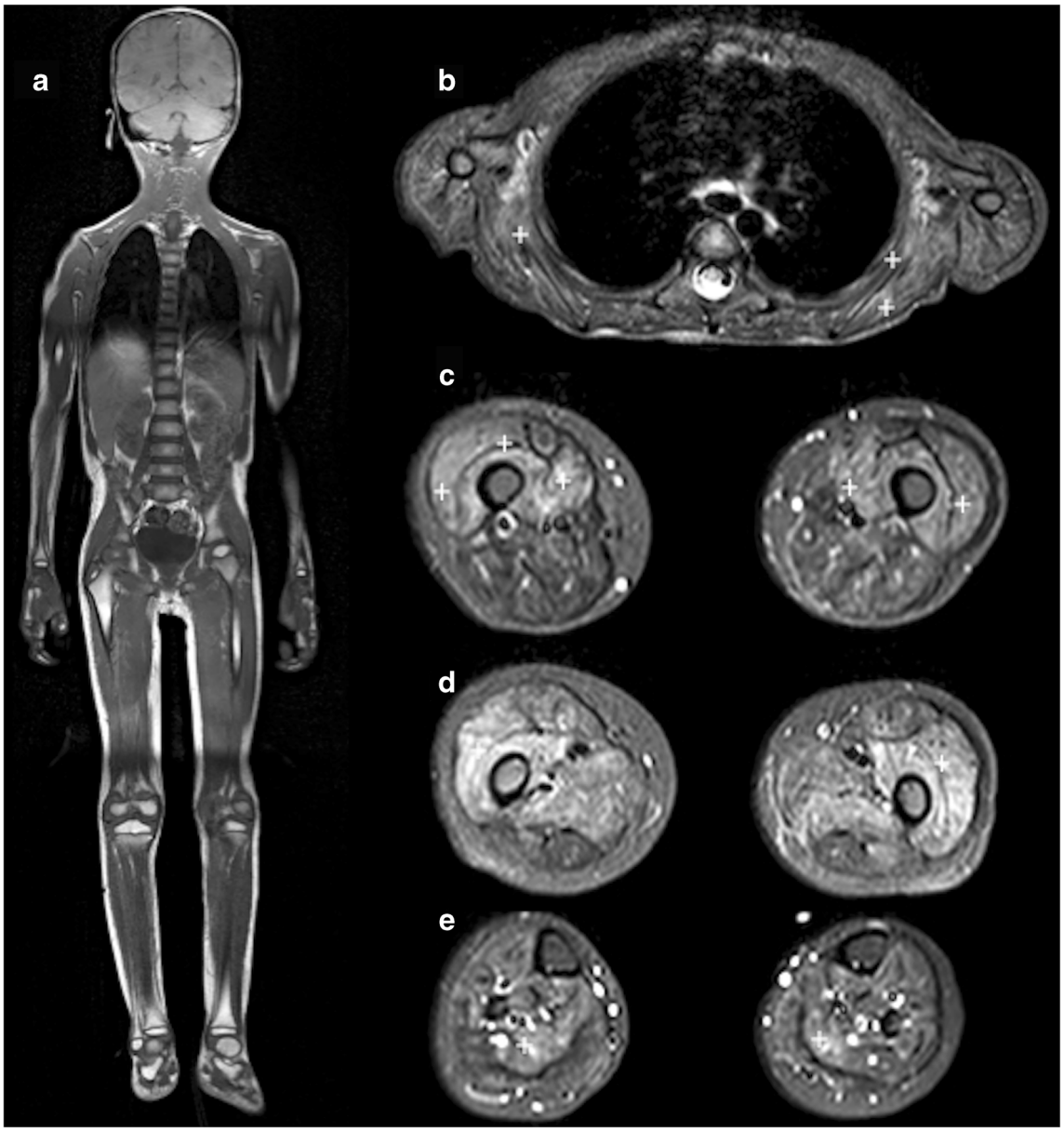

Fig. 2 Total body MRI with coronal FSE T1 (**a**) and axial images STIR from the thoracic girdle (**b**); proximal (**c**) and distal (**d**) thighs; and proximal leg (**e**) of a 7-year-old male with definite diagnosis of polymyositis. Showing in **a** no muscle atrophy or fat infiltrations. In **b**–**e,** diffuse edema pattern (+) in the subscapularis and supraspinatus muscles (**b**), in the anterior thighs (**c**, **d**) compartments, and in the soleus muscle in the leg (**e**)

in PM, edema is more common in the adductors [15••, 29]. Pipitone et al. [30] also compared the MRI findings of the thighs of patients with PM and DM and concluded that there are muscle groups more frequently affected in dermatomyositis, such as the quadratus femoris, the tensor fasciae latae, and the gracilis. However, larger studies are needed to confirm these findings.

A common MR finding in patients with dermatomyositis is edema along the muscle fascia (Fig. 4) and in the subcutaneous fat, which is less commonly seen in patients with polymyositis [7], where the WBMRI has the same importance for muscle pathology as in PM [21].

"Milk of calcium" is a rare form of calcinosis found in patients with DM (Fig. 3). The recognition of "milk of calcium" and its differential diagnosis, especially with soft tissue infections and deep abscesses, are needed, and the exact mechanism of the formation of collections of "milk of calcium" in DM is unknown [31].

Radiograph shows the calcifications, and ultrasound and MRI can easily characterize the fluid collections.

Milk of calcium collections are excellent demonstrated by MR imaging, with the signal intensity of the collections reflecting the differences in the fluid content and fluid/calcium levels [32].

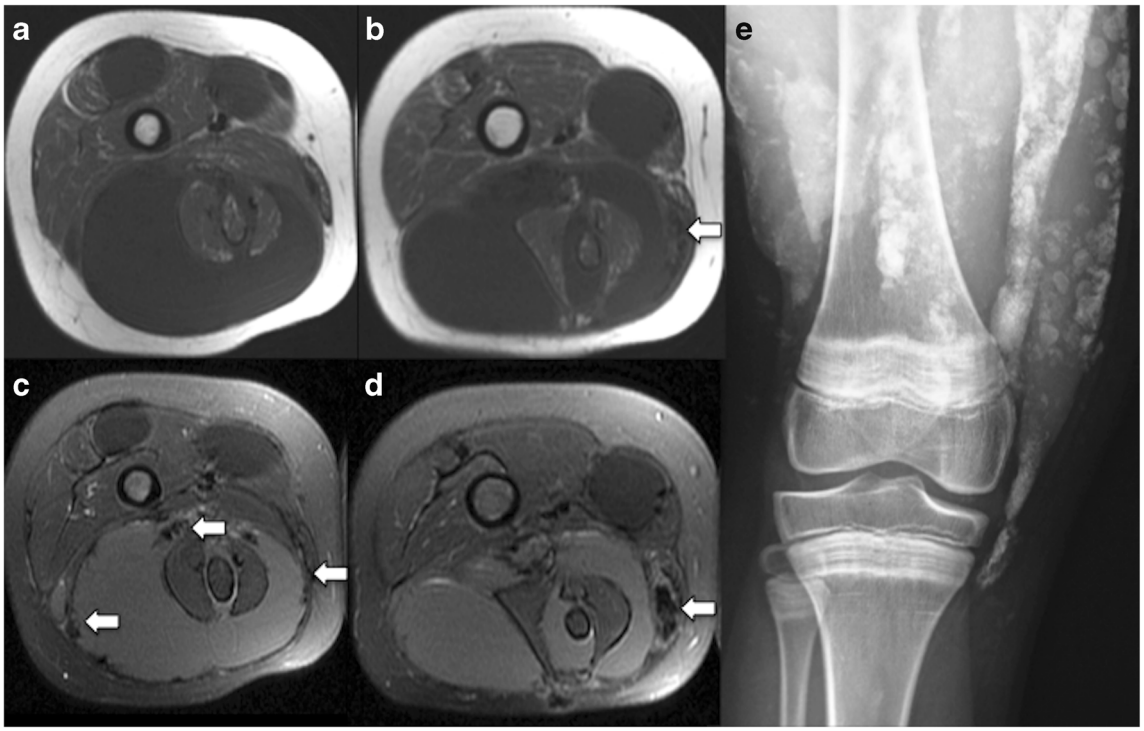

Fig. 3 Eleven-year-old girl with clinical diagnoses of dermatomyositis. Axial FSE T1 (**a**, **b**) and fat-saturated T2 weighted (**c**, **d**) images from the thighs demonstrate symmetric thigh muscle involvement, predominantly in the posterior muscular compartment, with fluid collections (milk of calcium) in the intermuscular septa with low signal calcifications in the periferic region (arrows). Frontal radiograph (**e**) shows the classic soft tissue sheet-like calcification

Immune-Mediated Necrotizing Myopathy

Clinical and Histological Aspects

INM is characterized by an acute or subacute course of proximal muscle weakness with an increase in serum muscle enzymes and minimum extramuscular manifestations [1••]. The disease can affect both adult and children patients. A muscle biopsy will show necrotic and regenerative fibers with a strong macrophage reaction, minimal lymphocytes infiltration, and class-1 major histocompatibility complex upregulation. INM can be classified according to the presence of autoantibodies against 3-hydroxy-3-methylglutaryl coenzyme-A reductase (HMGCR) and a signal recognition particle (SRP) [33, 34]. Up to two thirds of patients with anti-HMGCR myopathy are associated with previous exposure to statins. INM can also occur after viral infections in association with cancer and in patients with connective tissue disorders, such as scleroderma [1••].

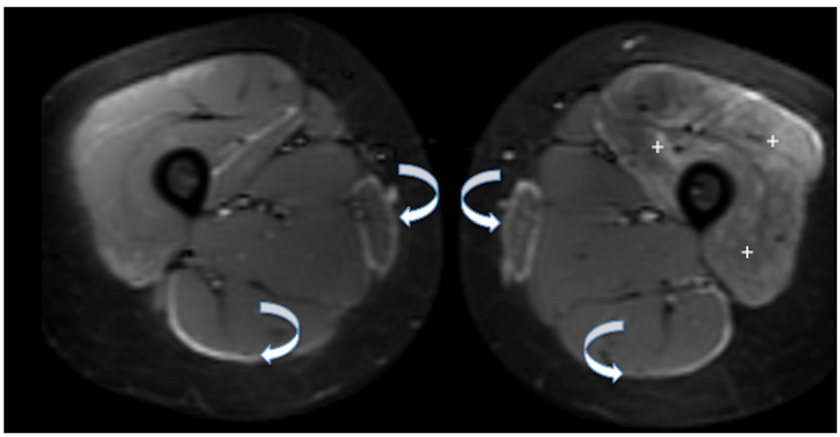

Fig. 4 Twenty-eight-year-old woman with a known history of dermatomyositis. **a** Axial inversion recovery image demonstrates patchy intramuscular edema within the vastus lateralis and vastus intermedius muscles in the left (+) as well as edema within the deep fascia surrounding the hamstrings, gracilis, and sartorius muscles (curved arrows). Case contributed by Dr. Douglas Mintz, HSS, NY, USA

Imaging Findings

To date, few studies have been published regarding the imaging characteristics of IMN [29, 35, 36], with most of them using MR imaging.

In the setting of IMN, muscle MRI can be useful in identifying areas of muscle edema for biopsy, increasing the diagnostic accuracy [9], and MRI may be a useful tool for monitoring the evolution of muscle disease over time [37].

The muscle MRIs of patients with active IMN show generalized muscle edema, muscle atrophy, and fatty replacement of muscle with minimal fascial edema. Muscle atrophy and fatty replacement occur preferentially in the lateral rotators, gluteus, medial compartment, and posterior compartment in the thighs and legs (Fig. 5) [35].

In parallel with the clinical differences in severity, muscle MRI of anti-SRP myopathy patients demonstrates higher rates of atrophy and fatty replacement compared to anti-HMGCR myopathy patients [35].

The muscle MRI characteristics of autoantibody-negative IMN have not yet been described [37].

The only study that compared MR imaging features in IMN patients with other inflammatory myopathies patients showed that the extent of muscle involvement is higher in IMN patients than in those with DM or polymyositis, that asymmetry is more common in IMN and that IMN patients have relatively less involvement of the anterior compartment of the thighs compared with sIBM patients [35].

Sporadic Inclusion Body Myositis

Clinical and Histological Aspects

sIBM is considered the most common form of myopathy in patients over 50 years of age with a prevalence of 3.5 in 100,000 individuals and a male/female ratio of 3:1 [38]. Clinically, the disease progresses slowly over the years and predominantly affects the quadriceps and the gastrocnemius muscles in the lower limbs and the finger flexors in the upper limbs [1••, 39]. Dysphagia is a common complaint, but respiratory muscles are usually spared [40]. The level of CK is slightly increased or even normal, and mild sensory neuropathy can occur in some patients, which can be detected in an electrophysiological study [40].

The skeletal muscle abnormalities include an endomysial inflammatory reaction in association with degenerative changes, characterized by the presence of rimmed vacuoles, intracytoplasmic inclusions formed by the accumulation of abnormal proteins, β-amyloid deposits, and mitochondrial

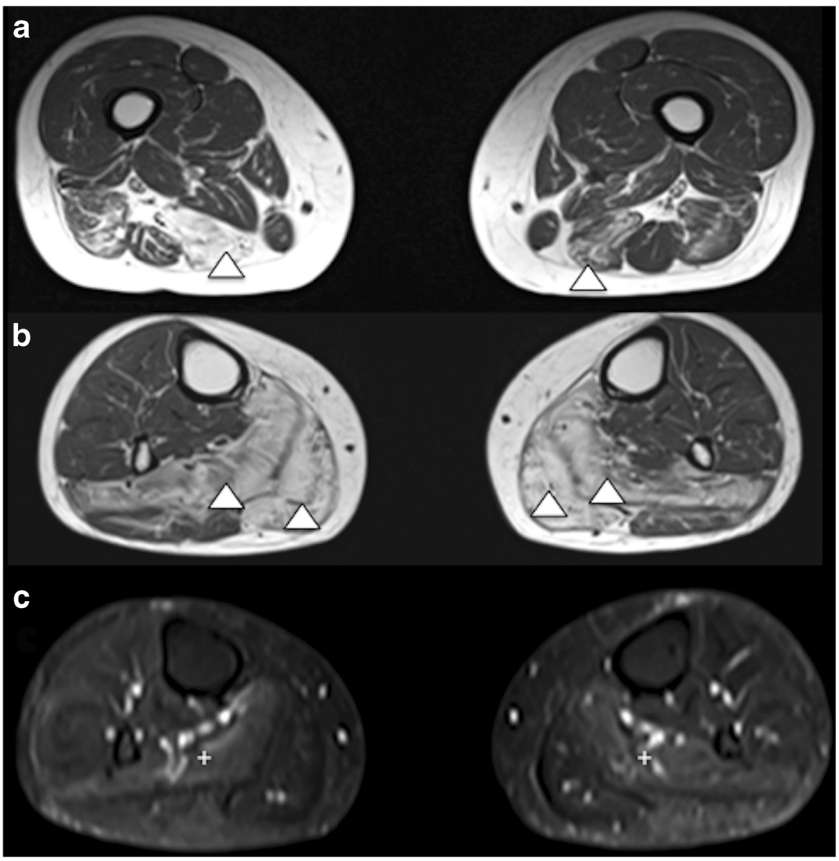

Fig. 5 Axial images FSE T1 (**a**, **b**) and STIR (**c**) from the thighs and leg of a 62-year-old woman with biopsy proven chronic anti-HMGCoA-reductase (HMGCR) myopathy (immune-mediated necrotizing myopathies), related to chronic statin use. **a** and **b** show asymmetric atrophy and fat infiltrations (arrow head) more extensive in posterior thighs compartment (**a**) and posterior legs compartments. In **c**, the edema pattern (+) is present only in the soleus bilateral

changes [39, 40]. The presence of such degenerative alterations suggests that the disease might actually be in the form of muscular degeneration with an associated inflammatory reaction, which could induce or even worsen the degeneration [41]. The occurrence of a degenerative process might explain the failure to immunosuppressive therapy in these patients [39, 40]. sIBM is commonly suspected when a patient with a polymyositis diagnosis does not respond to therapy.

Imaging Findings

A recent published study from our group [42••] showed that sIBM changes assessed by MRI (Fig. 6) more extensively affect the lower extremities, usually asymmetrically, and with more severe changes in the distal muscles. The most frequent pathologic muscle MRI finding was fat infiltration, followed by atrophy and lastly an edema pattern. The characteristic fat infiltration pattern was assessed by MRI, mostly affecting the lower extremities, with more pronounced changes in the distal muscles and with predominant involvement in the thigh of the anterior muscular compartment, which could help differentiate sporadic IBM from other myopathies [43].

The most affected muscles were the medial gastrocnemius, the vastus lateralis, and the flexor digitorum profundus (Fig. 7). Most of the patients presented a peculiar pattern of quadriceps muscle involvement, characterized by severe changes in the vastus muscles associated with significantly less severe involvement of the rectus femoris (Fig. 6). The undulating fascia sign (Fig. 7) is a common imaging pattern found in patients with sporadic IBM, with previous studies showing this sign is present in 75% of patients. It is defined by the presence of a wavy fascia between the severe atrophic and fat-infiltrated vastus intermedius and vastus lateralis muscles [42••].

Patients with sIBM also presented with less severe changes of the other tight muscular compartments, with the sartorius being the most frequently involved. The association of quadriceps and sartorius muscles involvement is a useful hint for the diagnosis of sIBM because the sartorius is usually spared in other adult-onset myopathies [42••].

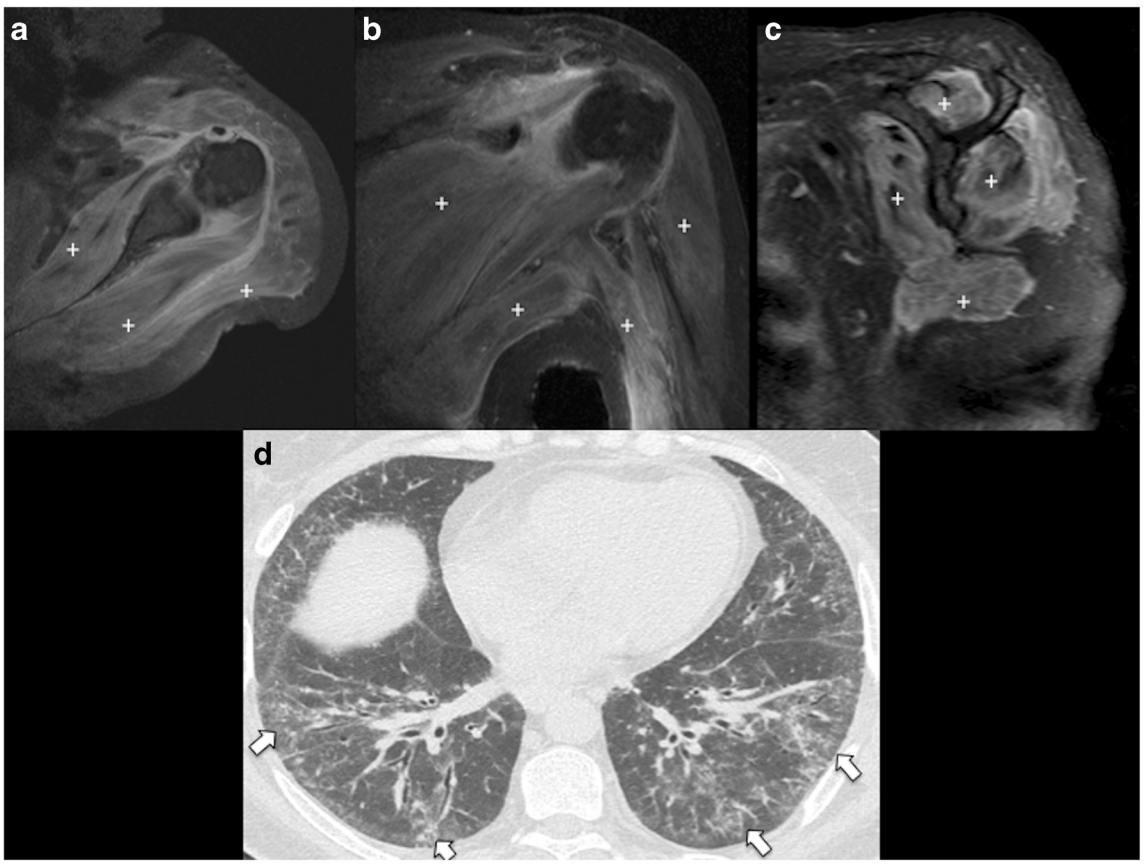

Fig. 6 Axial images FSE T1 (**a**, **b**) and STIR (**c**, **d**) from the thighs of a 68-year-old woman with sporadic inclusion body myositis. **a**, **b** show asymmetric atrophy and fat infiltrations more extensive in the distal segments, with the characteristic pattern of involvement of the quadriceps muscles. The rectus femoralis (arrows) are relatively preserved, while there is significant involvement of the vastus lateralis (arrowheads), intermedium, and medialis. In **b**, the undulating fascia sign (*) is shown. In **c**, **d**, the edema pattern (+) is present only in the vastus lateralis of the right thigh

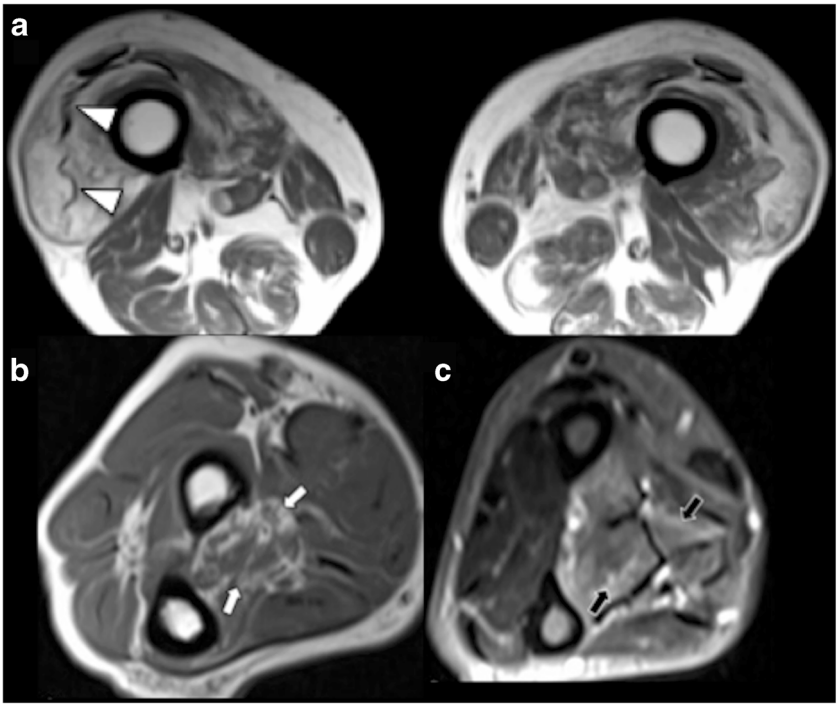

Fig. 7 Axial FSE T1 images of the distal thighs (**a**) of a 76-year-old male with sporadic inclusion body myositis. Bilateral vastus lateralis shows severe fat infiltration and more severe atrophy of the anterior compartment of the thigh compared to the posterior compartment. In the right thigh, the undulating fascia sign is present (white arrowheads). Axial FSE T1 from the proximal third forearm (**b**) and STIR from the middle third forearm (**c**). **b** shows fat infiltration (white arrows) of the flexor digitorum profundus. The finger extensors are spared by the fat infiltration. In **c**, the flexor digitorum profundus shows a high T2 signal consistent with the edema pattern (black arrows)

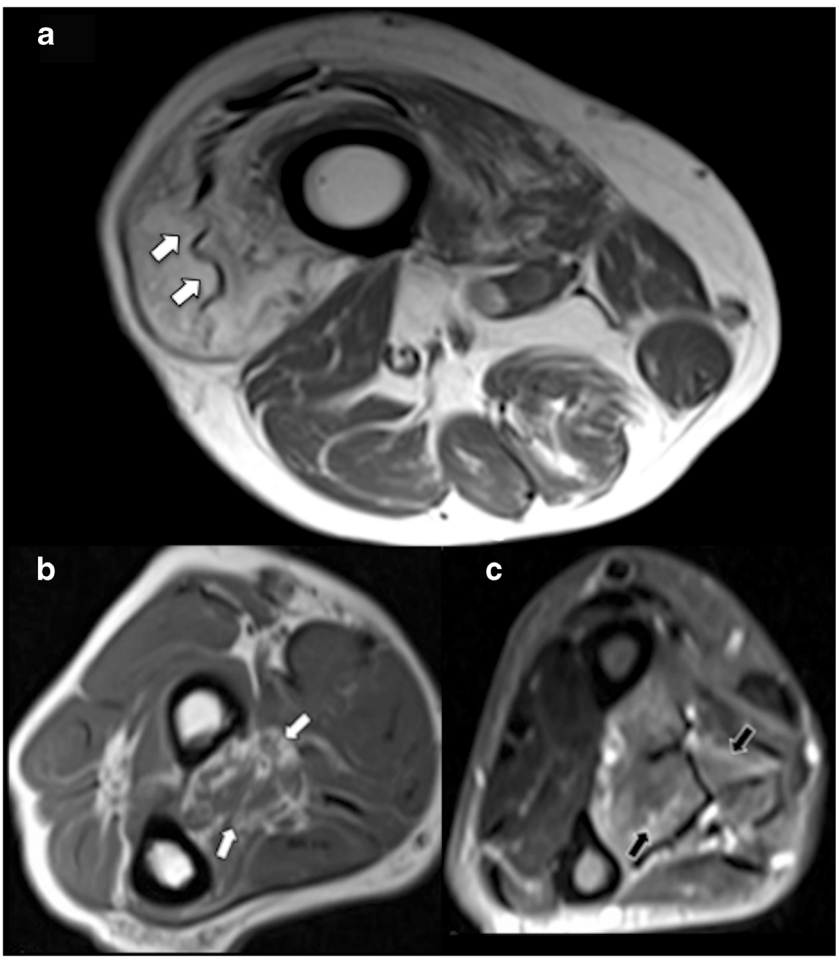

Fig. 8 Forty-two-year-old female with clinical features including interstitial lung disease, non-erosive arthritis, myositis, Raynaud's phenomenon, unexplained fever, and mechanic's hands, with anti-Jo-1-positive and definitive diagnosis of antisynthetase syndrome. Axial (**a**), coronal (**b**), and sagittal (**c**) left shoulder and arm STIR images show a diffuse and extensive muscle edema pattern (+). The chest CT shows a typical nonspecific interstitial pneumonia (NSIP) pattern (arrows)

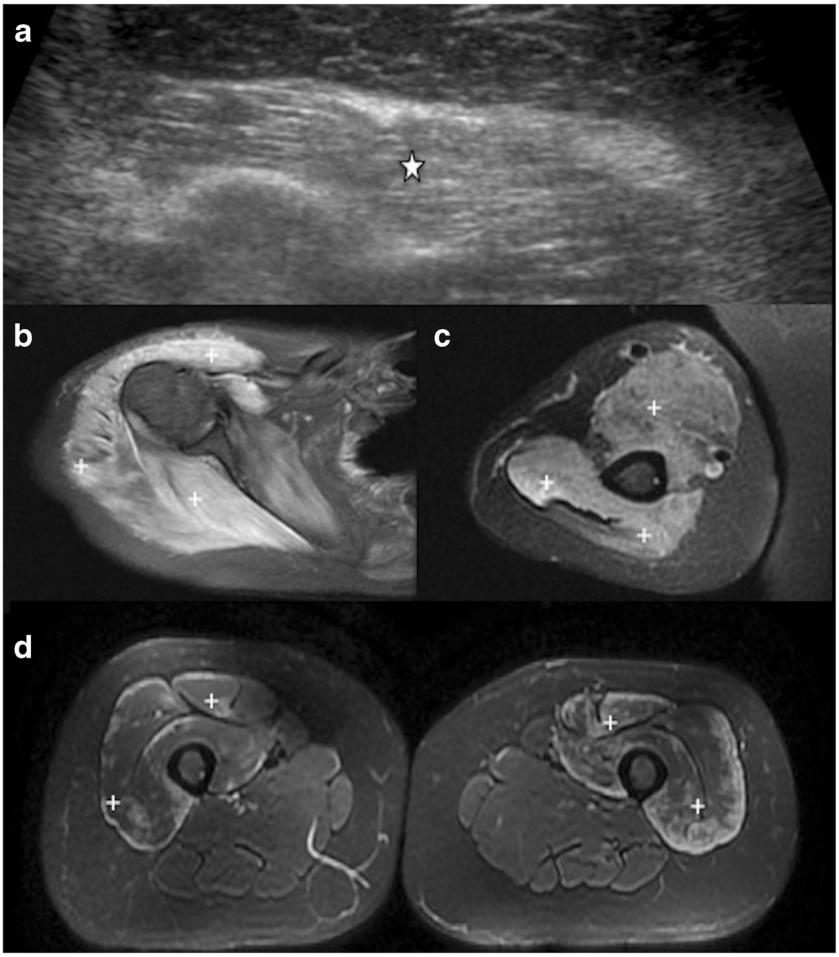

Fig. 9 47-year-old female patient in treatment for breast cancer presented with proximal myalgia and weakness 3 months after initiation of tamoxifen. In **a**, a US exam show diffuse increase ecogenicity (star) in the triceps muscle. In **b**–**d**, axial STIR images show muscle edema pattern (+) in the anterior compartment of the thighs and in the upper right arm/shoulder. There are elevated CK levels, biopsy compatible with inflammatory myopathy, and negative PET-CT. There was significant clinical and laboratorial improvement after the discontinuation of tamoxifen (drug-induced myopathy)

Overlap Myositis and Antisynthetase Syndrome

Clinical and Histological Aspects

Myositis may occur as part of other connective tissue diseases (overlap myositis), such as rheumatoid arthritis, scleroderma, mixed connective tissue disease, Sjögren's syndrome, and systemic lupus erythematosus [44]. The most representative form of overlap myositis is antisynthetase syndrome (ASS) [45, 46]. It is characterized by the presence of myositis, interstitial lung disease, and joint involvement. Patients present proximal muscle weakness, elevated muscle enzymes, and myopathic NCS/EMG [47]. Other manifestations include fever, Raynaud's phenomenon, and mechanic's hand (hyperkeratosis, peeling, and palmar fissures). It is characterized by the presence of serum anti-aminoacyl-tRNA synthetase autoantibodies, the most common of which are histidyl tRNA synthetase (anti-Jo1), threonyl tRNA synthetase (anti-PL7), and alanyl tRNA synthetase (anti-PL12) [44, 45].

Imaging Findings

Defined MRI abnormalities were identified in the majority of patients with ASS (65%) with an edema pattern, fat infiltration, and atrophy (Fig. 8). Thigh muscles MRI changes were most common in the posterior compartment followed by the anterior and medial compartments. Fascial edema was distributed almost equally in the three compartments in approximately 21% of the patients [48].

Toxic and Drug-Induced Myopathies

Clinical and Histological Aspects

Toxic and drug-induced myopathies as a group represent the most common form of myopathy with an estimated prevalence of more than 2000 per 100,000 people in the Western Hemisphere [49]. Toxicity to the muscle tissue can be caused by a variety of substances, including therapeutic drugs, resulting in a wide spectrum of symptoms that range from mild discomfort to permanent damage and disability [50].

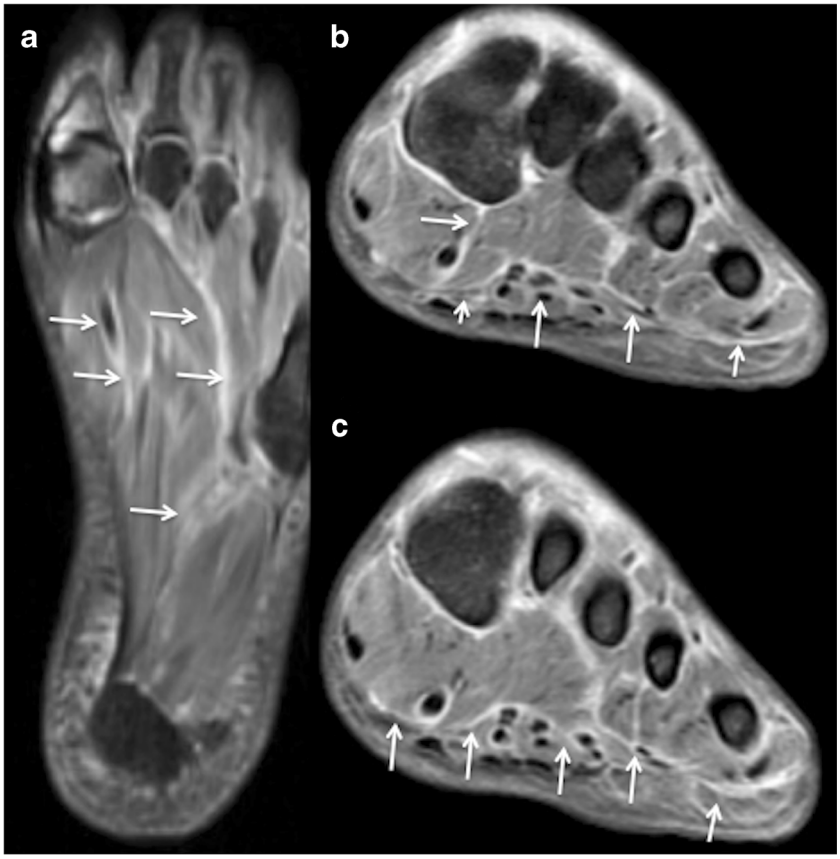

Fig. 10 Fifty-two-year-old man with acute eosinophilic fasciitis in the foot. Axial (**a**) and coronal (**b**, **c**) enhanced fat-suppressed, T1-weighted spin-echo MR image shows intense fascial enhancement

Cholesterol-lowering drugs, particularly statins, have been the most commonly prescribed class of potentially myotoxic medication in recent years [51].

Substances can have direct or indirect adverse effects on muscle. Direct effects can be focal (secondary to drug being

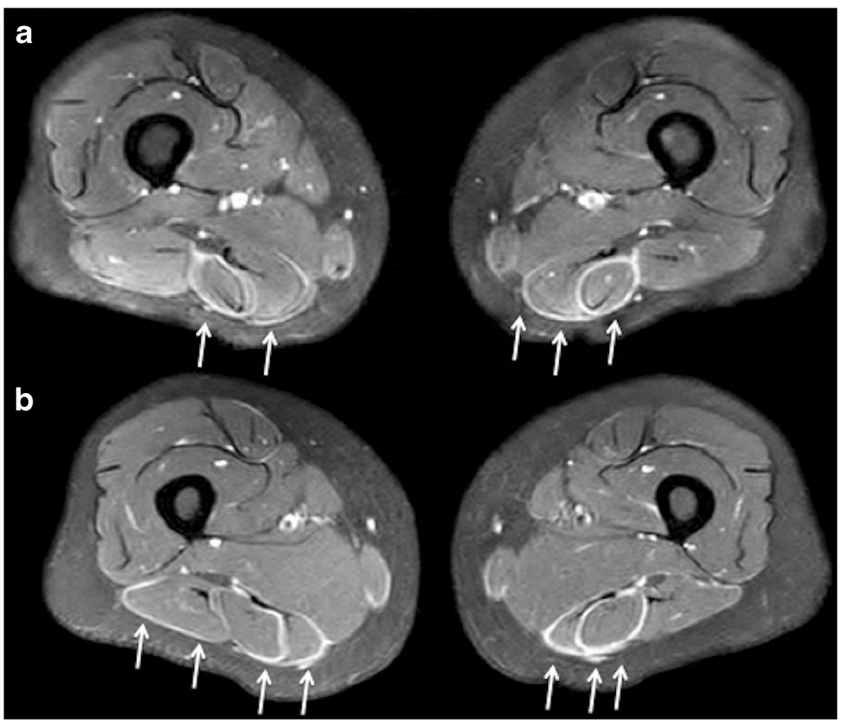

Fig. 11 Thirty-six-year-old man with subacute eosinophilic fasciitis in the thighs. Axial (**a**, **b**) and enhanced fat-suppressed, T1-weighted spin-echo MR image shows intense fascial enhancement in the posterior compartment

injected into tissue) or generalized. Indirect toxic effects may result from an electrolyte imbalance or an immunological reaction [52]. Although some categories of drugs are associated with specific forms of myopathies, a drug can cause more than one type of myopathy.

A drug-induced myopathy is defined as the subacute onset of muscle weakness, fatigue, myalgia, creatine kinase (CK) elevation, or myoglobinuria in patients who were previously without muscle disease when exposed to therapeutic doses of certain drugs. After the discontinuation of the suspected agent, the clinical or biochemical signs of muscle involvement usually improve, supporting the causative effect of the myotoxic drug [50].

A good medical history, including current and previous medication history, is essential to correlate the time of exposure to the onset of neuromuscular deficit.

The pathophysiological mechanisms are diverse and in many cases unclear. Eight main categories of toxic myopathies have been described [51], and some of the most important related agents are listed below:

1. Necrotizing myopathy: statins, other cholesterol-lowering drugs, and immunophilins.
2. Amphiphilic myopathies: chloroquine, hydroxychloroquine, and amiodarone.
3. Antimicrotubular myopathy: colchicine and vincristine.
4. Mitochondrial myopathy: nucleoside reverse-transcriptase inhibitors.
5. Inflammatory myopathy: cimetidine, levodopa, and imatinib mesylate.
6. Hypokalemic myopathy: diuretics, laxatives, amphotericin, toluene abuse, licorice, corticosteroids, and alcohol abuse.
7. Steroid myopathy/critical illness myopathy: high-dose IV steroids and nondepolarizing neuromuscular blocking agents.
8. Unknown: omeprazole, isotretinoin, finasteride, and emetine.

Mild symptoms such as myalgia or cramps occur in one fifth of statin users, while severe myotoxicity in the form of rhabdomyolysis only occurs at a rate of 0.44 per 10,000 patient-years. However, considering that close to 30 million Americans are currently prescribed a statin medication, patients with significant myotoxicity are regularly encountered in clinical practice. Both mild and severe side effects are self-limiting, with the resolution of symptoms after an average of 2 months after discontinuation of the offending medication [53]. However, in a subset of patients, statins can trigger an immune-mediated myopathy that progresses even after the medication is discontinued and responds to steroid/immunosuppressant therapy (previously described in the IMN section).

Imaging Findings

MR imaging findings (Fig. 9) are nonspecific, consisting of symmetrically increased muscle size and edema, typically affecting the buttocks, quadriceps, adductors, and deep calf muscles [20]; if there is no edema on STIR imaging, then there is no active myopathy [49]. In statin-induced myopathy, however, posterior compartment muscles of the thighs and the legs tend to be more prone to pathological changes [49].

Eosinophlic Fasciitis

Clinical and Histological Aspects

Eosinophilic fasciitis is a relatively rare, scleroderma-like disorder that typically presents with marked extremity edema followed by skin induration. Symptoms of stiffness, swelling, and pain often present suddenly, usually after strenuous activity. First described by Shulman [54] in 1975, eosinophilic fasciitis is poorly understood and clinically characterized by the presence of peripheral eosinophilia, hypergammaglobulinemia, elevated sedimentation rate, and scleroderma-like skin findings primarily involving the extremities [54, 55]. An important feature of this disease is its responsiveness to corticosteroid therapy, underscoring the need for early diagnosis and treatment [56].

Imaging Findings

MRI findings in active eosinophilic fasciitis (Figs. 10 and 11) should be highly characteristic [57] and include fascial thickening, hyperintense signal within the fascia on fluid-sensitive sequences, and fascial enhancement after IV contrast administration.

Summary

Imaging studies play an important role in the evaluation of patients presenting with symptoms of inflammatory myopathy. Together with the clinical history, familiarity with the imaging features is essential for formulating an accurate differential diagnosis. Furthermore, imaging plays a central role in guiding muscle biopsy, which remains the gold standard for the diagnosis of myopathy and could be used to the assessment of treatment response.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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