Specialty area:

Multifarious Manifestations of Muscle Disease

Speaker:

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Title:

Multifarious Manifestations of Muscle Disease: Metabolic Conditions & Genetic Disorders.

Target audience:

Radiologists interpreting clinical MRI of the musculoskeletal system including whole body MRI.

Highlights

- Diabetic Muscle Ischemia (DMI) occurs in patients with long-standing, poorly controlled type 1 or type 2 diabetes.
- DMI may be unilateral or bilateral and often affects noncontiguous muscles in the thighs and calves.
- The imaging features of drug-induced myopathy are nonspecific and should be interpreted in clinical context.
- Neuromuscular diseases have different, sometimes highly characteristic, distribution and involvement patterns in skeletal muscles. The forms of involvement within individual muscles are nonspecific and typically include edema-like pattern and fatty infiltration pattern.
- Whole body magnetic resonance imaging (WBMRI) is particularly useful in the evaluation of neuromuscular disorders, allowing assessment of the entire musculoskeletal system.

OBJECTIVES

Review clinical and imaging findings of metabolic conditions and genetic disorders affecting skeletal muscle with an emphasis on diabetes, drug-induced myopathies and the most common forms of muscular dystrophy.

DISCUSSION

Diabetic Myopathy

Diabetic myopathy encompasses various muscle disorders, including diabetic muscle ischemia, pyomyositis, inflammatory myositis and muscle denervation. Historically, diabetic myopathy has been described more frequently in patients with type 1 diabetes. However it may occur more frequently in patients with type 2 diabetes than previously reported, which may be explained in part by an increasing prevalence of type 2 diabetes. Diabetic Muscle Ischemia (DMI), also known as diabetic muscle infarction or diabetic myonecrosis, characteristically occurs in patients with long-standing, poorly controlled diabetes and associated complications such as retinopathy, nephropathy, and neuropathy. Patients typically present with abrupt onset of severe pain localized to the thigh or calf, accompanied with swelling, and sometimes a palpable, painful mass, that evolves over days or weeks. Absence of leukocytosis and fever is characteristic. MR imaging features of DMI include muscle enlargement as well as muscle and fascial edema. The findings may be unilateral or bilateral and often are seen in noncontiguous muscles in the thighs and calves. In the thigh, the anterior compartment is typically involved, while the posterior compartment is most commonly affected in the calves. Following intravenous gadolinium administration, muscle enhancement is seen. In contrast to pyomyositis, areas of muscle ischemia or necrosis in DMI tend to appear heterogeneous, with linear streaks of enhancement crossing central nonenhancing regions surrounded by large regions of enhancing muscle. When the clinical presentation and imaging findings are characteristic for DMI, biopsy may be avoided, because the symptoms tend to resolve with conservative management.

Drug-induced and toxic myopathies

A wide range of therapeutic and recreational drugs can cause myopathy. The spectrum of drug-induced myopathies includes painless myopathies (without or with neuropathy, or with abnormal neuromuscular transmission, aka myasthenic syndrome) and painful myopathies (polymyositis, other painful myopathies, or painful myopathy with neuropathy). Overall, drug-induced myopathy and rhabdomyolysis are relatively uncommon adverse drug reactions, except those caused by cholesterol-lowering agents

and glucocorticoids. In clinical practice 5–10% of patients receiving statins develop myopathy which is associated with the use of all statins (class effect) and is dose dependent. The pathophysiologic mechanism of statin-associated myopathy is unknown and probably multifactorial. 4 clinical drug-induced syndromes are defined: statin myopathy (any muscle complaint related to statin usage); myalgia (muscle complaints without serum creatinine kinase (CK) elevation), myositis (muscle complaints with serum CK elevation) and rhabdomyolysis (muscle complaints with elevated CK levels 10 times the normal upper limit, associated with nephropathy). Patients typically present with muscle pain which is usually symmetrical, involving proximal muscles without CK elevation or less frequently with mild CK elevation. Clinically significant rhabdomyolysis is extremely rare. The risk of statin-associated myopathy is increased in patients with impaired renal or liver function, diabetes, hypothyroidism and advanced age. The imaging features of drug-induced myopathy are nonspecific, and limited to edema-like pattern, representative of acute myositis or myonecrois, and fatty infiltration or atrophy pattern. Aside from direct effect on a muscle, certain drugs such as antibiotics (most notably, fluoroquinolones), and corticosteroids can cause tendinopathy and tendon rupture, thus secondarily causing muscle atrophy.

Neuromuscular disorders

Muscular dystrophies are characterized by permanent and progressive muscle weakness. There are various clinical, pathological and genetic classifications of neuromuscular disorders. Most commonly neuromuscular disorders are classified according to the age of onset (first, second or third decade of life) and the pattern of muscle involvement (e.g. facial, proximal, distal). The final diagnosis of a neuromuscular disease can be established through a comprehensive work-up involving a physical and neurological examination, hematological and biochemical tests, muscle power testing, electromyography, muscle biopsy, mode of inheritance and genetic analysis. Neuromuscular diseases have different distribution and involvement patterns in skeletal muscles, and the atrophy and fatty infiltration can be highly selective and characteristic for certain diseases. Whole body magnetic resonance imaging (WBMRI) is particularly useful because it allows assessment of the entire musculoskeletal system. It plays an important role in defining a pattern of muscle involvement and thus guiding further selection of appropriate genetic and biochemical diagnostic tests, monitoring the

progression of disease as well as facilitating the planning of muscle biopsy. Within the individually affected muscle, many neuromuscular disorders share common characteristics on MRI in terms of muscle signal abnormality and morphology, which typically include edema-like pattern and fatty infiltration pattern. The degree of muscular dystrophy is rated according to standardized rating scales which are based on the amount of fatty degeneration ranging from normal appearance to complete fatty degeneration.

Duchenne's muscular dystrophy (DMD), X-linked recessive muscular disorder, is the most frequent variant of muscular dystrophy, characterized by initial symmetric and selective involvement of the proximal pelvic girdle muscles followed by involvement of the calf and proximal shoulder girdle where muscles are progressively replaced by adipose and fibrous tissue. In the early stages of DMD, MRI displays abnormal signal in the gluteus maximus and adductor magnus, followed by involvement of the quadriceps, rectus femoris, and biceps femoris, with selective sparing of the gracilis, sartorius, semitendinosus, and semimembranosus. In the lower legs, the gastrocnemii are affected earlier and more severely than other muscle groups. Pseudohypertrophy of the calves is present in 80% of patients.

References/Suggested reading

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