13. IDIOPATHIC INFLAMMATORY MYOPATHY, EVERYTHING IT SHOULD NOT BE: ASYMMETRICAL, NORMAL CK, HIGH FEVER

Emer Gates[1], Ben Faber[1], Steve Hepple[1], and Harsha Gunawardena[1]

[1]Rheumatology, North Bristol Trust, Bristol, United Kingdom

Introduction: Myositis can be infective, metabolic or immune-mediated. Idiopathic inflammatory myopathy, which is immune-mediated, tends to be subacute, with symmetrical symptoms, overlap clinical features and positive autoimmune serology. We present a case of acute onset lower limb seronegative inflammatory myopathy with a normal creatinine kinase (CK), a marked acute phase response that responded promptly to immunomodulatory therapy.

Case description: A 68-year-old man presented with a 2-week history of lower limb pain with subjective weakness, on a background of well-controlled type 2 diabetes mellitus and hypertension. He was admitted with worsening symptoms of marked left thigh pain, night sweats, and fevers. On admission, he had swinging pyrexia (above $>39\,^{\circ}C$) while remaining haemodynamically stable. He had focal tenderness over the left anterolateral thigh, with a good range of movement, normal power and no signs of focal collection or cellulitis. There were no extra-muscular features to suggest systemic infection or overlap connective tissue disease.

Bloods showed C-reactive protein (CRP) 225, normal CK 212 and negative blood cultures. X-rays knee, femur and pelvis were normal. Magnetic resonance imaging (MRI) on T2, fat-suppressed STIR sequences demonstrated increased signal/oedema both thighs throughout the anterior muscle compartment and along the fascial plane, notably most severe in the left vastus lateralis. He was treated empirically for infective myositis. Despite 14 days of broad-spectrum antibiotics, he remained febrile with persistently elevated CRP. There was no focal collection, lymphadenopathy or occult malignancy on CT abdomen and pelvis. Trans-oesophageal echocardiogram showed no evidence of infective endocarditis but revealed incidental moderate aortic stenosis.

The patient described persistent now bilateral thigh pain with continued normal CK and high CRP. Full autoimmune screen (ANA, ANCA, ACE and complement studies) was negative. Despite negative nuclear and cytoplasmic HEp-2 immunofluorence, extended myositis immunoblot was negative. Muscle biopsy from the left vastus lateralis demonstrated inflammation within the perimysium and perivasculature. In view of biopsy findings and no response to anti-microbial therapy, prednisolone (0.5mg/kg) with significant clinical response (resolution of fever and pain) with concurrent normalisation in CRP. The patient remains in remission following steroid reduction with no additional immunomodulatory therapy required.
Discussion: We report a case of idiopathic inflammatory myopathy presenting with predominantly asymmetrical symptoms, normal CK, marked inflammatory response and negative myositis autoantibodies. Diagnosis was confirmed on MRI and muscle biopsy. The normal CK can be explained by the histology demonstrating inflammation in perivascular regions and around muscle fibres, rather than inflammation or necrosis in the muscle fascicles and fibres themselves. Idiopathic inflammatory myopathy including sporadic inclusion-body myositis, dermatomyositis, overlap CTD myositis and polymyositis/necrotising myopathy subsets are distinguishable based on clinical features, autoantibodies, MRI and biopsy features. The table below summarises the atypical aspects of this case. Differential diagnoses for this case include atypical infection, sarcoid myopathy and amyloid myonecrosis secondary to diabetes.

TABLE: Features of typical idiopathic inflammatory myopathy compared with this atypical case.

	Idiopathic inflammatory myopathies	Our patient-typical features	Our patient-atypical features
Symptoms	Pain, fever, weakness.	Pain and fever.	Normal power.
Clinical distribution	Symmetrical, proximal muscle groups.		Predominantly asymmetrical (worse on left), only in thighs.
Antibodies	Myositis associated autoantibodies		Seronegative
Muscle enzymes	Elevated CK		Normal CK.
Inflammatory markers	Normal to slightly elevated CRP		Markedly raised CRP and WCC.
MR imaging results	Focal muscle oedema in affected muscles		Diffuse and speckled muscle oedema

Key learning points: Early idiopathic inflammatory myopathy can have inflammation around the muscle fascicles in the perimysium. Normal CK does not rule out a diagnosis of idiopathic inflammatory myopathy.
Idiopathic inflammatory myopathies can present atypically with fevers $>39\,^{\circ}C$, significantly raised inflammatory markers, and asymmetrical symptoms and MRI findings.
In the absence of overlap features, normal CK and negative serology, MRI and biopsy can delineate the type of myositis and direct management.
Conflicts of interest: The authors have declared no conflicts of interest.