Case 10632

A case of juvenile dermatomyositis
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Section: Musculoskeletal system
Area of Interest: Extremities Musculoskeletal system
Procedure: Imaging sequences
Imaging Technique: MR
Special Focus: Inflammation Case Type: Clinical Cases
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Patient: 8 years, male

Clinical History:

An 8-year-old boy was referred with 5 weeks history of generally feeling unwell with lethargy, intermittent fevers, rash on chest, face, knees and elbows and loss of appetite. Neurological examination revealed proximal muscle weakness in the lower-limbs. Laboratory tests demonstrated leucopenia, elevated transaminases and creatine kinases. Abdominal ultrasonography was normal.

Imaging Findings:

MRI of pelvis and upper thigh was performed. T1 axial, STIR axial and STIR coronal imaging sequences were used. STIR sequences showed diffuse bilateral patchy high signal intensity involving the skeletal muscles.

Discussion:

Juvenile Dermatomyositis is a rare multisystem disorder of children. It is the most common idiopathic inflammatory myopathy accounting for 85 percent of the cases [1]. Autoimmune reaction in response to environmental triggers like a viral infection in genetically susceptible individuals has been proposed [1]. Timely diagnosis and management is of paramount importance to avoid complications and functional disability in children.

Symmetrical proximal muscle weakness is the hallmark clinical manifestation of Juvenile Dermatomyositis. The most common symptom at the disease onset is skin rash (Gottron's and Heliotrope rashes). In a classical presentation like in this case, diagnosis can be made based on the clinical presentation. MRI findings in proximal musculature and skin is a sensitive tool in confirming the diagnosis.

MRI demonstrates inflammatory changes in proximal muscles in the form of high signal intensities on T2 weighted or fat suppressed T2 (STIR) images like in this case. Though clinically Juvenile Dermatomyositis causes symmetric proximal muscle weakness, the muscle involvement can be patchy or asymmetric [2]. In more severe cases, there will be involvement of skin, subcutaneous connective tissue and muscle fascia. T1 weighted images detect muscle atrophy and fatty infiltration in the setting of chronic disease [2]. MRI thereby can accurately demonstrate the intensity and extent of the disease and defines an appropriate biopsy site. Literature review also shows a decline in high signal intensities in patients responding to immunosuppressive treatment. Hence, MRI also demonstrates a role in monitoring disease process [3].

Abnormal MRI signals in muscles characterise myopathies. MRI is a sensitive tool in detecting muscle inflammation, although not specific. The signal changes on imaging needs to be interpreted on the background of the clinical
Differential Diagnosis List: Juvenile Dermatomyositis, Viral infections, Polymyositis, Muscular dystrophy

Final Diagnosis: Juvenile Dermatomyositis

References:

C. Hutchison, B. M. Feldman (2012) Pathogenesis and clinical manifestations of juvenile dermatomyositis and polymyositis. UpToDate
Figure 1

Description: High signal intensity on upper thigh muscles on STIR axial image. Origin: Department of Radiology, Townsville Hospital, Australia.
Figure 2

Description: High signal intensity on STIR coronal image
Origin: Department of Radiology, Townsville Hospital
Description: Involvement of muscles of all compartments of thigh. Origin: Department of Radiology, Townsville Hospital