Juvenile Dermatomyositis - role of MRI

Clinical History:

A 2 year old girl presented with a violaceous rash around the eyes and the knee joints and irritability. A month later she complained of difficulty in climbing stairs and getting up. Clinical examination revealed papules over the MCP and PIP joints and an abnormal Gower maneuver.

Imaging Findings:

Laboratory investigations revealed raised CPK enzyme. X-ray examination obtained was normal. MRI was performed of both the thighs. MRI revealed diffuse increase in signal intensity on water-sensitive sequences within the muscles and the superficial and deep fascia, skin, and subcutaneous fat. Proximal muscles were more involved. The abnormalities were best seen on STIR and T2WI with fat saturation. T1WI and Proton Density appeared normal.

Discussion:

Juvenile dermatomyositis (JDM) is a disease marked by muscle weakness and skin rash. JDM affects females more often than males, with a ratio of 2.3:1 in the United States. In the United Kingdom, the ratio is even higher at 5:1. The median age of onset of JDM is 6.8 years in girls and 7.3 years in boys, with a median delay to diagnosis of 3-4 months. One fourth of patients are aged 4 years or younger at diagnosis. The first sign of JM is usually a skin rash. Characteristic reddish-purple rashes are Gottron’s papules (bumps found over the knuckles, elbows and knees) and heliotrope rash (purplish rash around the eyes). JDM patients can have weak muscles at the same time they see the skin rash, or the weakness may come after the rash over days, weeks or months. The weaker muscles are usually closer to the body and the child may have trouble climbing or standing from a seated position. The skin rash and weak muscles are caused by inflammation or swelling in the blood vessels under the skin and in the muscles. Diagnosis is confirmed by muscle enzyme values followed by an MRI or an EMG or muscle biopsy. The differential diagnosis for the muscle and fascial inflammation is broad, and clinical correlation is generally required. The MRI features of dermatomyositis may mimic necrotizing fasciitis, strenuous exercise, pyomyositis, viral myositis, myositis ossificans, acute muscle denervation, metabolic myopathies, graft versus host disease following organ transplant, medication-related myositis (e.g., diuretics), other rheumatologic conditions such as lupus, polyarteritis nodosa, and juvenile rheumatoid arthritis as well as undifferentiated inflammatory myopathy. Inflammation of a single muscle or muscle group should not raise the diagnostic possibility of dermatomyositis, since this entity is only a consideration if there is a typical bilateral, symmetric, and proximal appendicular distribution of muscle inflammation. The best sequence for diagnosis on MRI is T2WI with Fat Saturation or a STIR. T1WI and PD fail to reveal the
The diagnosis of dermatomyositis in this case was made based on clinical history, rash, MRI features with a typical distribution of myositis, and abnormal muscle enzyme (CPK) values. The MRI and clinical features of dermatomyositis made muscle biopsy unnecessary in this case.

**Differential Diagnosis List:**  Juvenile Dermatomyositis

**Final Diagnosis:**  Juvenile Dermatomyositis

**References:**

Diagnosis of bone and joint disorders by Donald Resnick.
Pediatric and adolescent musculoskeletal mri (case based approach) (springer) by J.Herman Kan, Paul K. Kleinman.
Figure 1

Description: Coronal STIR image showing diffuse increase in signal intensity of all thigh muscles
Origin:
Description: Coronal STIR image showing diffuse increase in signal intensity of gluteal muscles along with hyperintensity in the subcutaneous tissues Origin:
Figure 2

a

Description: Axial T2WI with fat saturation showing diffuse signal intensity in the muscles and the gluteal subcutaneous fat and fascia Origin:

b

Description: Axial T2WI with fat saturation showing diffuse signal intensity in the muscles Origin:
Description: Axial T2WI with fat saturation at the level of the calf showing diffuse signal intensity in most muscles with relative sparing of the medial muscles Origin:
**Figure 3 a**

**Description:** T1WI fails to reveal any abnormality **Origin:**
**Figure 4 a**

**Description:** T2WI showing subtle abnormality in the muscles however fat saturation makes the signal abnormality more profound. **Origin:**
Description: PD image-no obvious abnormality discernable Origin: