A Case of Myositis in Sickle Cell Anemia

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CASE HISTORY

- 39-year-old male with sickle cell anemia requiring monthly hospital admission for pain crises, hypertension secondary to ESRD requiring dialysis s/p failed renal transplant
- He presented with atraumatic swelling and pain out of proportion to exam over the inner right thigh
- He could not bear weight
- This focal pain contrasted with the generalized, bilateral arm and leg pain typical of his vaso-occlusive crises
- He denied fever, loss of sensation, motor weakness, paresthesias, recent viral illness, rash.

PHYSICAL EXAM

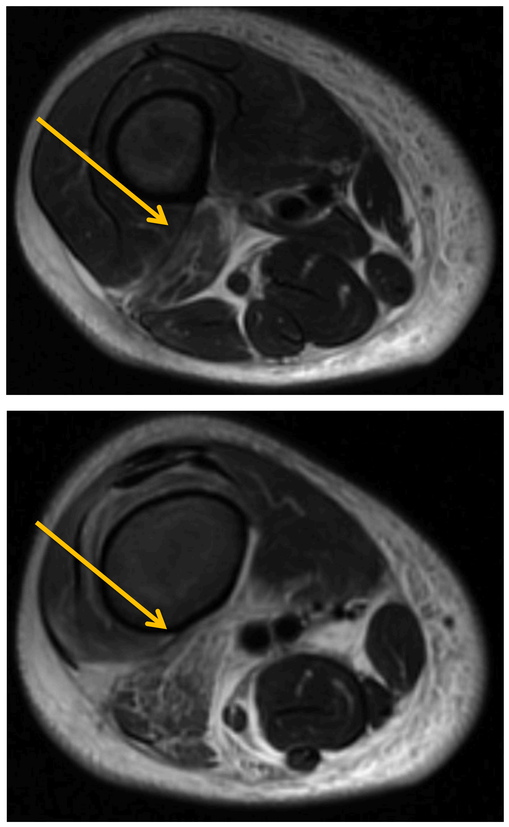
- *Extremities*: asymmetrical, homogenous 1+ non-pitting edema over the anteromedial aspect of the right thigh. Exquisite, diffuse tenderness to palpation. No surface erythema. No exacerbation of pain with resisted hip adduction with knees in flexion or extension.
- *Neurologic*: CN II-XII intact. 5/5 strength at flexion and extension of hip, knee, ankles. 5/5 strength of hip abduction and adduction. Sensation to light touch intact.
- *Skin*: no malar, discoid, V-neck, Shawl, heliotrope rash. No Gottron papules

DIFFERENTIAL DIAGNOSIS

- Polymyositis, dermatomyositis, focal myositis
- Idiopathic inflammatory myopathy
- Avascular necrosis, myonecrosis
- Necrotizing fasciitis, osteomyelitis, cellulitis
- Compartment syndrome

TESTS & RESULTS

- *WBC* 13, *ESR* 114, *CRP* 2.4, *CPK* 49
- *MRI*: patchy intramuscular edema scattered throughout the anterior, posterior, and medial compartments of the thigh. A small amount of fluid is noted about the sartorius and gracilis muscles



DIAGNOSIS

Myositis

TREATMENT AND OUTCOME

- Methylprednisolone for 5 days
- Doxycycline for 7 days
- Edema and pain resolved after the steroid + antibiotic course
- He was discharged several days later

FOLLOW-UP

- He was re-admitted three weeks later for vaso-occlusive crisis but did not complain of the focal right lower extremity pain

DISCUSSION

- He had no clinical evidence of connective tissue disorders such as lupus, SLE, Sjogren's. Myonecrosis was less likely without a fluid collection and a normal CPK. By exclusion, this could be focal myositis for which they recommended EMG as well as muscle biopsy if the EMG was abnormal.
- Idiopathic inflammatory myopathies include polymyositis, dermatomyositis, inclusion body myositis, among others
- All are characterized by progressive worsening of multiple myalgias with histopathologic evidence of inflammatory infiltrate in muscle tissue
- Gottron papules, Shawl sign are pathognomonic for dermatomyositis
- Distal muscles are more affected than proximal muscles in inclusion body myositis
- Diagnosis made with positive muscle enzyme markers, CK, aldolase, muscle biopsy, EMG, MRI
- Mainstay of therapy is immunosuppression: corticosteroids, DMARDs, IVIG
- 13 case reports of acute onset, proximal muscle weakness, swelling, and pain in patients with sickle cell disease
- Myonecrosis secondary to infarction during sickle cell crisis is thought to occur more commonly than it is diagnosed