**Polymyositis and dermatomyositis**

**Key points:**
1. There is a huge differential for muscle weakness. Polymyositis and dermatomyositis are rare.
2. Muscle biopsy is key for diagnosis of PM/DM.
3. Steroids are for now the best known treatment.

**Intro:** Polymyositis and dermatomyositis are rare idiopathic myopathies. Occur 1/100,000, have a female predominance, and peak incidence in 5<sup>th</sup> decade of life.

**Pathophys:** DM has immune complex deposition injury and PM has T-cell mediated injury.

5 criteria to diagnose DM/PM
1. proximal muscle weakness
2. rash of DM
3. increased CK
4. myopathic changes on electromyography
5. muscle biopsy abnormalities

Clarifications of above criteria

**Rash:**
- Malar rash: like SLE rash but this does involve the nasolabial folds
- Gottron’s sign: symmetric, non-scaling violaceous erythema on extensor surfaces of hands, can also see on elbows. May look like psoriasis.
- Heliotrope rash: red-violaceous rash on upper eyelids
- Periungual erythema and hemorrhages, cracked skin on fingertips “mechanic’s hands”

**Muscle biopsy abnormalities:**
DM: primary lesion in blood vessels. See cellular infiltrate in the perifascicular or perivascular areas.
PM: infiltrative cells in fascicles with inflammatory cells invading muscle fibers

Other organ systems involved: polyarthritis, CHF, interstitial lung disease, dysphagia

**Labs:**
- CK, LDH, aldolase, AST, ALT are often elevated
- ANA positive in over 80%
- Anti-RNP
- Anti-RNA positive in over 30%

**Differential Diagnosis (broad!)
- Motor neuron disease (ALS): see more distal muscle weakness than PM/DM and no myopathic changes on EMG
- Neuromuscular disorders (myasthenia gravis, Eaton-Lambert): see normal enzymes, facial weakness, anti-acetylcholine antibodies
- Muscular dystrophies (Duchenne’s, limb-girdle, Emery-Dreifuss, etc)
- Other collagen vascular diseases
• Infectious: septic (staph, strep, leprosy), viral (ie mono, influenza), bacterial, parasitic involvement (ie trichinosis, toxo)
• HIV inflammatory myopathies: like PM. AZT can cause mitochondrial myopathy but symptoms should resolve once drug is stopped
• Endocrine: thyroid, adrenal, parathyroid, hypocalcemia, hypokalemia
• Drugs: penicillamine, colchicine, HMG-CoA reductase inhibitors, hydroxychloroquine
• Toxins: EtOH/cocaine/heroin
• Metabolic myopathies (uremia, liver failure)
• Inherited metabolic disorders (glycogen and liped storage diseases): usually see post exertional pain and tenderness
• Inclusion body myositis: insidious, more prominent distally, inclusion bodies on biopsy
• Hypereosinophilic syndrome
• Sarcoid or amyloid myopathy

**Treatment:**
Only 2 published RCTs. Steroids are mainstay of therapy. One study showed 39% had normalization of CKs and 25% regained full strength after steroid treatment (better for those treated as outpatients).
Other therapies that have shown some success in small studies (most have under 20 patients): azathioprine, methotrexate, IVIG, cyclosporine, tacrolimus, cyclophosphamide, chlorambucil, tumor necrosis factors… more to come in the future.

**Prognosis:** Survival rate over 90% at 5 years if treated. Worse if --severe weakness --duration of disease present for longer period prior to treatment --dysphagia.

**Other things to consider:**
• Osteoporosis prevention: encourage exercise and screen for osteoporosis, treating early if present
• Use sunscreen
• Dysphagia precautions

**Is there an association of PM/DM with myocardial infarction??**

Reviewing the literature, there are some cases but the association is rare.
1. In 1996, a case report in Mexico named a patient who had an MI during an exacerbation of disease. Literature review in abstract quotes a “rare association”.
2. In 1979 a patient had changes on ECG consistent with acute MI but normal coronaries on angiogram.
3. Other study sites vasoconstriction caused by impaired vascular tone leading to a type of vasospastic infarct.

**Sources:**
UpToDate on polymyositis and dermatomyositis. 2002
Practical Rheumatology by Klippel