Polymyositis and Dermatomyositis

Symptoms and Physical Signs

- **MUSCLE WEAKNESS**, - **PROXIMAL**
- Symmetrical
- **Of insidious onset** (usually takes several months)
- **Sometimes painful** (myalgia and arthralgia)
  - DEFINITELY painful on palpation:
  - May even be ATROPHIC after a while
- **DYSPHAGIA +/- Aspiration**
- **FINE MOTOR CONTROL** is lost late in polymyositis
  - BUT is lost EARLY in INCLUSION BODY MYOSITIS

**CHARACTERISTIC RASH:** face, trunk, hands:
- **The heliotrope rash:** symmetric, confluent, purple-red, macular eruption
  Mainly face, the eyelids and periorbital tissue.
- **Gottron’s rash:** erythematous nail beds and scaly purple papular eruptions
  over the dorsum of the hands, especially MCP and interphalangeal joints.

Extramusculoskeletal manifestations:
- Interstitial lung disease or aspiration pneumonia
- Cardiac arrhythmia or congestive heart failure

Differentials to exclude:
- Vasculitis
- Progressive systemic sclerosis
- Infectious myositis
- Muscular dystrophy
- Eaton-Lambert syndrome
- Drug-induced myopathies - Corticosteroids, statins,
- Electrolyte disorders
- Inherited myopathies

INVESTIGATIONS:

POLYMYSITIS can be produced by a PARANEUROPOLYCLASTIC SYNDROME!
Especially ovaries, gastrointestinal tract, lung, and breast and NHL. 10-20% of patients with
dermatomyositis have neoplasms

The only specific antibody: **anti-Jo-1**
- antibody against histidyl-tRNA synthetase;
  associated with DM, PM, Raynauds’, interstitial pneumonitis, and so forth…

FBC may show some leucocytosis
EUC – are they in acute renal failure 2ndary to rhabdomyolysis?
Urinalysis may show blood++++ as myoglobinuria
ESR will be moderately elevated
Rheumatoid factor (RF) present in 50%
Antinuclear Antibodies (ANA) present in 50%
Acetylcholine Receptor Antibody for Mysathenia…
Creatine Kinase (CK) WILL BE RIDICULOUSLY HIGH
**ELECTROMYOGRAPHY:** excludes neurogenic cause
MUSCLE BIOPSY

MANAGEMENT:

1. Prednisone: monitor response with CK levels and clinically: no functional improvement?
2. ADD AZATHIOPRINE and watch for side-effects
   - bone marrow depression, hepatotoxicity, skin rashes, nausea and vomiting.
3. PHYSIO and EXERCISE; taper immunosuppressives, repeat electromyography in 3 months
4. Other options for second line therapy would be methotreaxate, chlorambucil, cyclosporin and intravenous immunoglobulin