



Polymyositis and Dermatomyositis

Symptoms and Physical Signs

All members of the same evil family, the Idiopathic Inflammatory Myopathies

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

Difficulty kneeling, climbing or descending stairs, raising arms, and arising from a seated position; difficulty holding the head up

SENSATION AND REFLEXES WILL BE PRESERVED

IS IT MYASTHENIA ?
Myasthenia gravis affects the EYE MUSCLES early; The myopathies NEVER affect the eye muscles.

- BUT is lost EARLY in INCLUSION BODY MYOSITIS

- **CHARACTERISTIC RASH: face, trunk, hands:** The "shawl" sign

These qualify your myositis as a **DERMATOmyositis** (commonest variety)

- 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

Need to ask about **FAMILY HISTORY** of muscle disease ...and **MEDICATIONS**

DIFFERENTIALS TO EXCLUDE:

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

INVESTIGATIONS:

POLYMYOSITIS can be produced by a **PARANEOPLASTIC 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...
THESE are the only 3 tests you really need

- 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** Will give accurate diagnosis; IF you manage to get a biopsy of an affected patch of muscle.

dermatomyositis is the only disease that shows "perifascicular atrophy" whereas polymyositis shows necrosis and inflammation

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 methotrexate, chlorambucil, cyclosporin and intravenous immunoglobulin