Inflammatory Myopathies
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- Polymyositis (PM)
- Dermatomyositis (DM)
- Inclusion body myositis (IBM)
Inflammatory Myopathies

- **General clinical picture:**
  
  i. Progressive, Symmetric, Proximal muscle weakness (sensations n DTRs preserved)……
  
  ii. Often presents with recurrent falls (knee buckling d/t Quadriceps weakness) and dysphagia (pharyngeal musculature)
  
  iii. Ocular n facial muscles spared
Inflammatory Myopathies

- Extramuscular features of Infl. Myopathies
  - Constitutional symp
  - Raynaud’s phenomena (if asso CTDs in PM)
  - CMP n Conduction defects
  - Subcutaneous nodules (DM)
  - RLD (resp mus weakness)
Polymyositis (PM)

- Usually in asso with CTDs
- Defined as Subacute infl myopathy of adults who do not have -
  1. Skin rash
  2. Facial or ocular muscle involv
  3. Muscle dystrophies on biochem muscle dr (metabolic dr/enzyme def/genetic dr)
  4. Endocrinopathy (thyroid/parathyroid, DM, Cushing’s ds, vit D def.)
# Polymyositis (PM)

## v. Exposure to myotoxic drugs or toxins

<table>
<thead>
<tr>
<th>Hypolipidemics</th>
<th>Alcohol</th>
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<tbody>
<tr>
<td>Glucocorticoids</td>
<td>Amphetamine</td>
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<tr>
<td>HAART (Zido)</td>
<td>Cocaine</td>
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<tr>
<td>HCQS</td>
<td>Phencyclidin</td>
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<td>D-penicillamine</td>
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<td>Colchicine</td>
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<td>Amiodarone</td>
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## vi. IBM (ruled out by muscle bx)
Dermatomyositis (DM)

- Proximal mus weakness + Skin Rash
- Skin rash:
  - Helitrope rash
  - Gottron rash
  - Shawl sign
  - Mechanic’s hands
  - Capillary dilatation at nail beds
- Asso malignancies- Breast, Ovary, Colon and Skin
Inflammatory body myositis: IBM

- Age above 50 yrs
- Early involv of distal mus (wasting) esp finger extensors n deep flexors
- Supf resemblance to MND or peripheral neuropathy (hand muscle wasting)
Pathogenesis:

- **Auto-immune:**
  1. Asso with other CTDs
  2. Auto-Abs. Anti-Jo-1 against RNA synthetase
  3. Asso with MHC genes (DR3)
  4. Response to immunosuppressive therapy

- Asso with some viral infxns..HIV, HTLV-I, Coxsackie
Differential diagnoses

- Ac. Muscle weakness
- GBS, Poliomyelitis
- Glycogen storage dr (myoglobinuria)
- Tropical myositis/pyomyositis (staph, strep, yersinia)
- Periodic paralysis (hyper- n hypokalemic)
- Parasitic myositis (toxoplasma, cysticercosis)
Differential diagnoses

- Subacute/Chronic muscle weakness
  - SMA, ALS
  - Muscular dystrophies (progression over yrs)
  - Glycogen storage disorders
    - (Myophosphorylase def, Acid maltase def)
  - Lipid storage dr (Carnitine def)
  - Endocrine myopathies
  - Drug induced myopathies
  - Macrophagic (PAS +) myofascitis
  - Chr. Fatigue synd/Polymyalgia rheumatica
Diagnosis

- Serum muscle enzymes: CK, LDH
- EMG: low amp, polyphasic units on voluntary activity
  : complex repetitive discharges & spontaneous activity with fibrillation
  : mixed (short n long duration) polyphasic units-indicating chronic process with muscle fiber regeneration
Diagnosis

**Muscle bx:**

1. **PM:** endomysial T-cell infilt surrounding healthy muscle fibers
2. **DM:** interfascicular perivascular infilt with perifascicular muscle fiber atrophy
3. **IBM:** endomysial T-cell infilt with vacuolar degeneration of muscle fibers, ragged-red fibers
Treatment

- **Glucocorticoids:**
  - Tab Pred 1mg/Kg/d X 1 mth
  - f/b tapering (over 2 mths) to 1mg/Kg alt day
  - f/b reaction by 5-10mg every 2-4 weeks till lowest effective dose
  - Second-line immunosuppressive drugs if no response by 3 mths......n accelerate steroid tapering
  - Î muscle weakness after initial improvement ....suspect Steroid Myopathy
Treatment

- Second-line immunosuppressive therapy:
  - 75% cases require these agents bcs of Steroid dependance, Steroid resistance, Steroid toxicity, Progressive disease
  - Options .. AZT (3mg/Kg/d)
    MTX (7.5-25 mg/wk)
    ...issue-pulm fibrosis (MTX or MCTD)
    Cyclophos (0.5-1 g iv mthly X 6 mth)
    IV Ig (2g/Kg divided over 2-5 days)
    .....issue- repeated infusions required