Inflammatory Myopathies

Inflammatory Myopathies

- 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 –

i. Skin rash
ii. Facial or ocular muscle involv
iii. Muscle dystrophies on biochem muscle dr (metabolic dr/enzyme def/genetic dr)
iv. Endocrinopathy (thyroid/parathyroid, DM, Cushing's ds, vit D def.)

Polymyositis (PM)

v. Exposure to myotoxic drugs or toxins
   - Hypolipidemics
   - Glucocorticoids
   - HAART (Zido)
   - HCQS
   - D-penicillamine
   - Colchicine
   - Amiodarone
   - Alcohol
   - Amphetamine
   - Cocaine
   - Phencyclidin

vi. IBM (ruled out by muscle bx)

Dermatomyositis (DM)

- Proximal mus weakness + Skin Rash
- Skin rash:

i. Helitrope rash
ii. Gottron rash
iii. Shawl sign
iv. Mechanic's hands
v. 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)
  - Endocirne 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 reuction by 5-10mg every 2-4 weeks till lowest effective dose

- Second-line immmunosuppressive drugs if no response by 3 mths……n acclelerate steroid tapering
- Î muscle weakness after initial improvement ….suspect Steroid Myopathy

Treament

- 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