INCLUSION BODY MYOSITIS

KEY CLUES:

- **Male**
- **>50 years old**
- **Isolated quadriceps myopathy - weakness and atrophy**
- **Early distal weakness - especially long finger flexors**
- **Asymmetrical, diffuse muscle weakness**

- inflammatory myopathy - but resistant to immunosuppressant therapies and IVIg
- most cases are sporadic
- much more common in males (5:1)
- some patients have 'related' autoimmune disorder
- late onset - most cases in those over 50 years old and not unusual in those in their 70's and 80's

- Muscle weakness - most commonly:
  - Quadriceps weakness - difficulty squatting, rising from a chair, climbing stairs, etc.
  - Weakness of forearm muscles (often brachioradialis) - weakened grip, weakened elbow flexion
  - Ankle dorsiflexion weakness - foot drop and tripping
  - Cricopharyngeal weakness - Dysphagia (+/- choking) can occur at any point in the disease.

SPECIFIC:
LONG FINGER FLEXION WEAKNESS

O/E the patient can bend at MCP joint but not DIP

- asymmetrical muscle involvement
- myalgia
- progression over months-years

Major Differential Diagnosis:
MOTOR NEURONE DISEASE!

DIAGNOSIS - clinical suspicion
- raised CK (non-specific)
- EMG changes (non-specific)
- various autoantibodies in ~20% (non-specific)
- Biopsy is needed to confirm diagnosis (characteristic microscopic changes - vacuoles and filamentous inclusions)

Presentation is variable!
However all cases eventually become a syndrome of diffuse, progressive, asymmetric, muscle weakness refractory to immunosuppressive treatment.