Inclusion Body Myositis – Treatment and Symptom Management

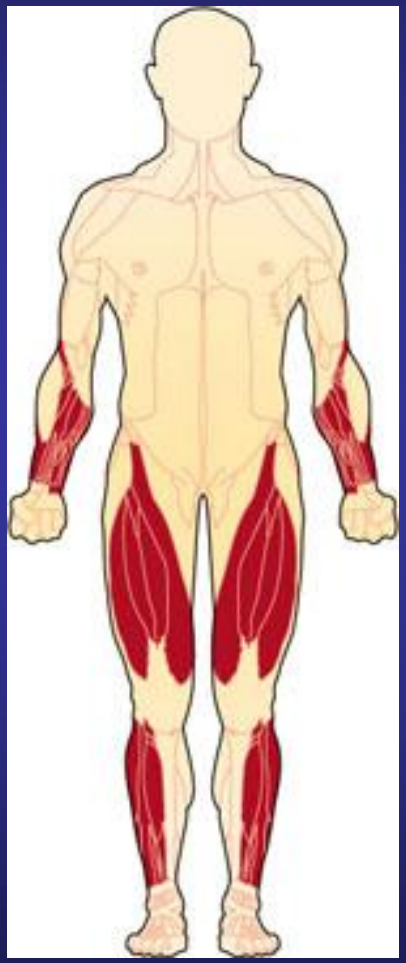
Inclusion Body Myositis (IBM)

Demographics,Epidemiology & Natural History

- Rare: prevalence is ~15 per 100,000
- Primarily a disease of later life – typically begins at age 50 or later (20% have symptoms at earlier age)
- The most common acquired muscle disease in those over 50
- Affects men slightly more than women
- Progressive – assistive devices typically needed within 10 years of onset
- No significant impact on life-span, i.e. not a fatal illness

Inclusion Body Myositis (IBM)
Clinical features

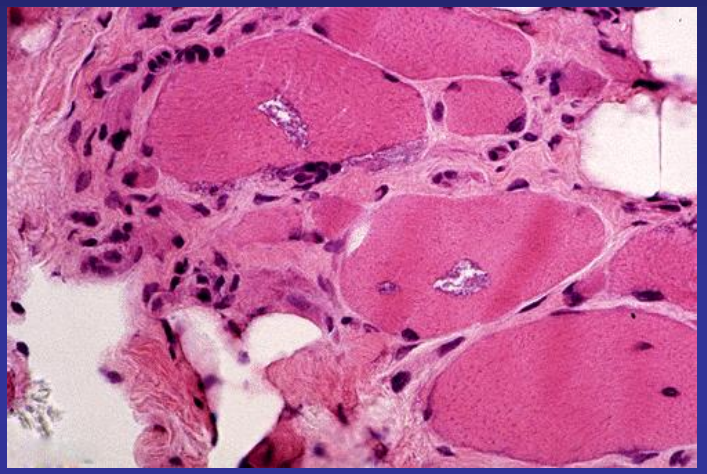
- Typical pattern of weakness:
  - Quadriceps
  - Finger & wrist flexors
- Eventual progression to other muscles
- Some muscles spared – eg Deltoids
- Pattern of muscle weakness can vary
- Possibly more rapid progression with onset >60 years
- Swallowing affected in 40-85%

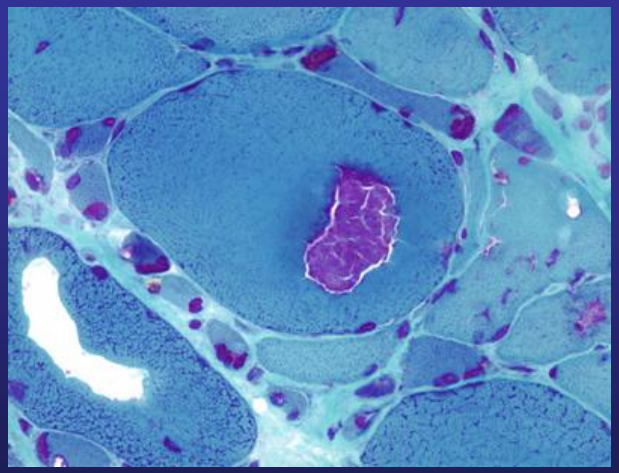


Inclusion Body Myositis (IBM)

Muscle Pathology

- Inflammation in muscle
- Rimmed vacuoles
- Inclusions

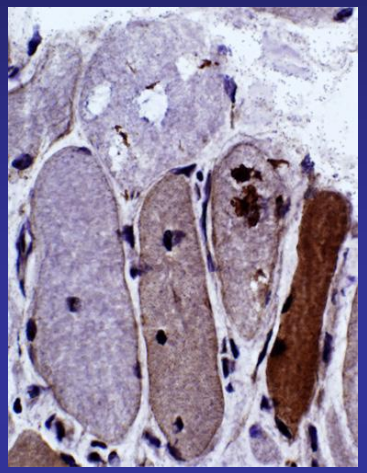


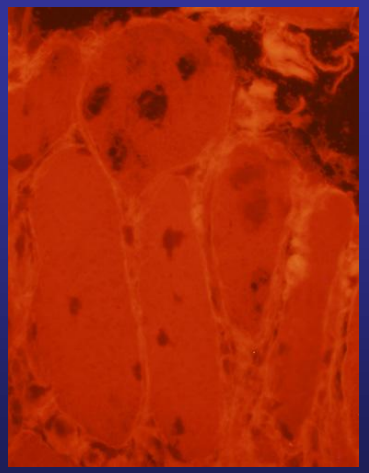


Inclusion Body Myositis

Amyloid deposition

- Protein aggregates found in muscle
  - Characteristics of amyloid proteins
- Similar to proteins found in brains of Alzheimer disease
- Possibly may be source of inflammation





Inclusion Body Myositis

Treatment

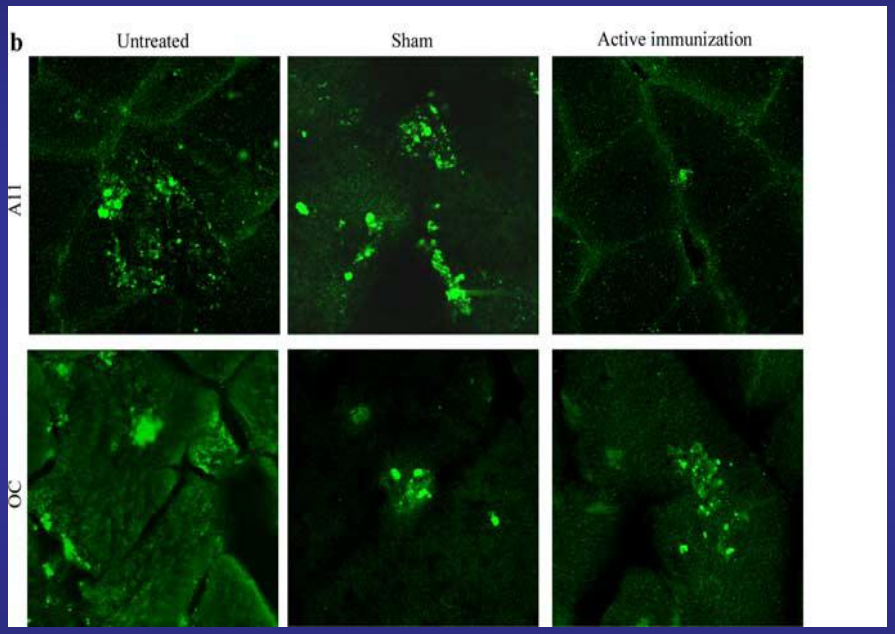
- Historically, treatment like PM or DM – largely unsuccessful
  - Corticosteroids
  - Immune suppression
- Can reverse inflammation, but no change in strength
- Anecdotal reports of efficacy
  - "Data is not the plural of anecdote"

Immune therapy in IBM

- Corticosteroids
  - Reduce inflammatory infiltrates in muscle, but no increase in strength
- IVIG
  - Limited efficacy in small numbers of patients
- Other immune suppressants
  - Only anecdotal reports of disease stabilization, no clear evidence for improvement in strength
- More directed immune therapy?
  - Etanercept, et.al. – anti-TNF-alpha therapy
  - May act by reducing inflammation related to amyloid deposits in muscle

Immunotherapy for IBM

- Mouse model for IBM
- Mice immunized with a protein derived from A-beta1-33 (a fragment of APP)
- After 3 mo immunization, mice had improved rotorod performance.
- Muscle bx showed less A-beta, less vacuoles & expressed fewer stress-related proteins
- Not directly practical for humans – potential toxicity, question about mouse model



Kitazawa, et.al. *J Neurosci* 2009;29:6132-6141

Inclusion Body Myositis

Treatment – Other Agents

- Co-Q10
- L-carnitine
- Creatine
- Vitamins – B2, B-6, B-12, E
  - Caution with toxic effects – B-6, E
- Testosterone, androgenic steroids

Inclusion Body Myositis

Supportive & Rehabilitative Strategies

- Reduction/discontinuation of corticosteroids
- Exercise: may require analgesics
- Weight control
- Appropriate rest
  - Role of sleep disorders, esp OSA
- Swallowing dysfunction
  - Cricopharyngeal myotomy
  - PE
- Appropriate bracing
  - AFO
  - Dynamic knee brace
- Mobility issues, fall prevention
- Prevent ankle swelling
- ROM, stretching
- Adaptive devices for arm/hand weakness