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

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