Question 8

Which one of the following clinical features best differentiates inclusion body myositis from polymyositis?

A. Quadriceps wasting
B. Deltoid wasting
C. Weakness of long finger flexors
D. Bilateral ptosis
E. Truncal Weakness

Both inclusion body myositis and polymyositis share the features:

- Proximal lower extremity weakness is usually the first sign with subsequent involvement of upper extremity and distal muscle groups.
- Proximal muscle weakness is typically more pronounced although distal weakness is predominant in some patients.
- The facial muscles may be involved,
- Oculomotor muscles are spared.

It just seems a random fact that weakness of long finger flexors is more suggestive of IBM – however there are some more definitive differentiators between the two that would make a better question

Answer C

Inflammatory myopathy
Polyomyositis and dermatomyositis
Dalakas MC, Hohlfeld R

The inflammatory myopathies are a heterogeneous group of subacute, chronic or acute acquired disease of skeletal muscle. They have in common the presence of moderate to severe muscle weakness and inflammation in the muscle.

Clinical presentation
Onset is acute or subacute over a period of several weeks and may follow systemic infection. Systemic symptoms prevail at onset eg lassitude and are followed by muscle weakness. Extensive oedema of skin and subcutaneous tissues is common (especially in the periorbital region)

Polyomyositis

- Muscles may be painful and tender in 60% of cases though onset is often painless
- Proximal muscles are first involved and initially weakness may be asymmetrical eg one quad only
- Weakness of posterior neck muscles will result in the head ‘lolling’ forwards
- Occasionally weakness may spread into distal limb muscle groups
- Pharyngeal and laryngeal involvement results in dysphagia nad dysphonia
- Cardiac muscles may also be involved
- Respiratory muscle weakness causes respiratory failure ( this may be disproportionately severe)
- The eye muscles are not involved unless there is coexistent myasthenia gravis
- Reflexes are retained (if absent, consider underlying carcinoma and added neuropathy)
- Immunopathology: in polymyositis and inclusion-body myositis, CD8 positive cells invade MCH-1 antigen expressing muscle fibres

Dermatomyositis

- Often more severe and acute
- Characterised by skin rash. Violet discoloration of light exposed skin including heliotropic discolouration of eyelids; raised scaly erythematous rash involving nose and cheeks, shoulders, extensor surfaces of limbs and knuckles
- Telangiectasia and tightening of skin are common and small ulcerated vasculitis lesions develop over bony prominences
- Childhood form – multisystem involvement. Calcification develops in skin and muscle with extrusion through skin. Muscle contractures develop – tip-toe gait. Gastrointestinal ulceration occurs
Immunopathology: the primary antigenic target in dermatomyositis is the endothelium of the endomysial capillaries. The disease begins when putative antibodies directed against endothelial cells activate complement C3.

**Differential diagnosis**
- Inclusion body myositis
- Acid maltase deficiency (presenting as respiratory failure)
- Limb girdle dystrophy
- Drug induced, toxic and metabolic myopathies

**Investigation**
- CK elevated – indicates disease activity and severity
- Electromyography – shows typical myopathic pattern
- Muscle biopsy shows necrosis of muscle fibres with inflammatory cells – lymphocytes. Plasma cells and leucocytes
- Antibodies RA and ANA present in 40%
- ESR elevated in most patients

**Treatment**
- Steroids

**Inclusion body myositis**
Sporadic inclusion body myositis (IBM) is classified along with polymyositis and dermatomyositis as one of the idiopathic inflammatory myopathies. However, despite some histologic similarities, the clinicopathologic manifestations of IBM are clearly distinct from the other two disorders.

- Presents after age 50yrs.
- Patchy and asymmetric in distribution.
- Muscle biopsy shows basophilic inclusion granules.
- Often clinically confused with polymyositis but response to immunotherapy is poor

Physical examination — The physical examination helps to determine the distribution of muscle weakness and atrophy. Findings of quadriceps, and finger flexor weakness and wasting and of foot drop due to tibialis anterior weakness is often noted.

**Differentiation between inclusion body myositis and polymyositis**

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<thead>
<tr>
<th></th>
<th>Inclusion body myositis</th>
<th>Polymyositis</th>
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<tbody>
<tr>
<td><strong>Sex</strong></td>
<td>Male&gt;female</td>
<td>Female &gt; male</td>
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<tr>
<td><strong>Age</strong></td>
<td>Rare before 50</td>
<td>Common before 50</td>
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<tr>
<td><strong>Onset</strong></td>
<td>Insidious</td>
<td>Acute or subacute</td>
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<tr>
<td><strong>Course</strong></td>
<td>Slowly progressive</td>
<td>rapid</td>
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<td><strong>Distribution of weakness</strong></td>
<td>Variable may be primarily distal, proximal symmetric</td>
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<tr>
<td><strong>CK</strong></td>
<td>Normal or &lt;10x normal</td>
<td>Often &gt;10x normal</td>
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<tr>
<td><strong>EMG</strong></td>
<td>Myopathic or mixed myopathic and neurogenic</td>
<td>myopathic</td>
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<tr>
<td><strong>Muscle biopsy</strong></td>
<td>Inflammation, rimmed vacuoles Inclusions</td>
<td>inflammation, fiber necrosis</td>
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<td><strong>Response to therapy</strong></td>
<td>Generally poor</td>
<td>expected</td>
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