

# image of the moment

**David Hilton-Jones**

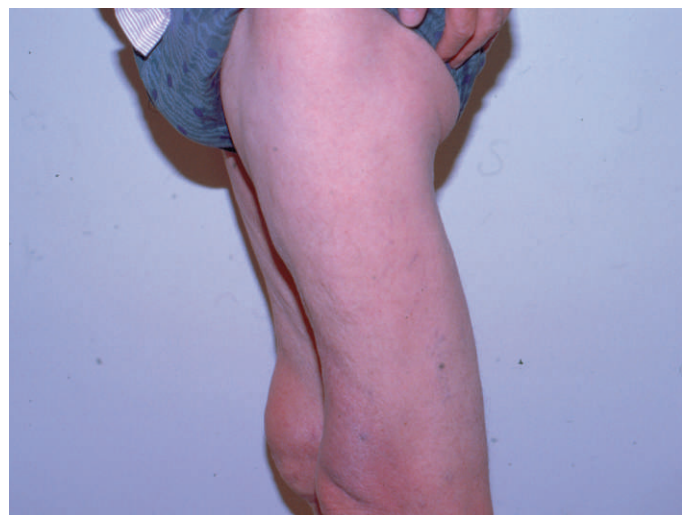
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# Inclusion body myositis



**Figure 1** Selective quadriceps wasting.



**Figure 2** Selective quadriceps wasting. Inclusion body myositis is the most common cause of 'isolated quadriceps myopathy'.

Since its delineation in the early 1970s, it has become apparent that inclusion body myositis (IBM) is the most commonly-acquired myopathy in middle-aged and older people. Although classified as one of the inflammatory myopathies, with polymyositis and dermatomyositis, several features set it apart, not least its resistance to immunosuppressant therapies. The diagnosis may be strongly suspected on the basis of clinical features alone, but confirmation is by the demonstration, in a muscle biopsy specimen, of characteristic light and electron microscopic changes

Patients typically present with symptoms relating to quadriceps weakness

## CLINICAL ASPECTS

### Presentation

Most cases present after the age of 50 years. IBM is substantially commoner in men, by at least 5 : 1. The overall incidence is in the order of 1/100 000 population/annum.

Patients typically present with symptoms relating to quadriceps weakness (Figs 1–3): 'knees giving-way', difficulty in rising from a chair and squatting. Observers may comment on quad-

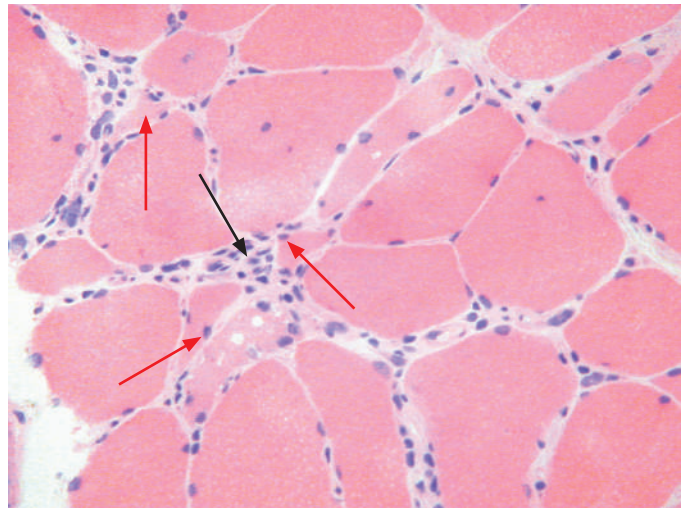


**Figure 3** Another example of selective quadriceps wasting.

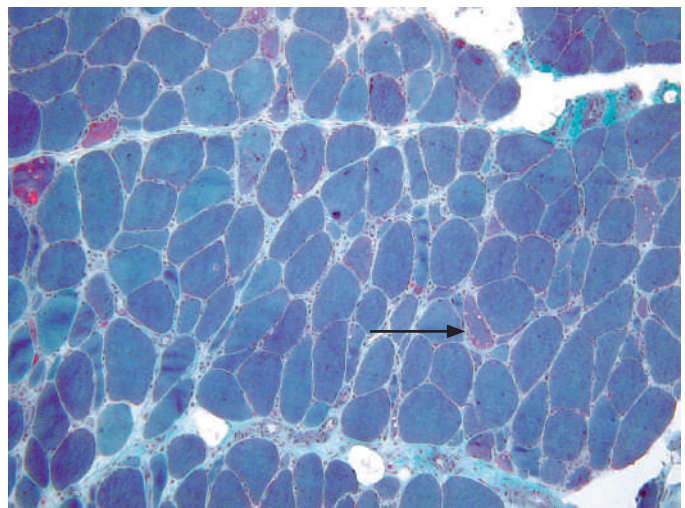
Most patients show little or no response to immunosuppressant drugs or to intravenous immunoglobulin



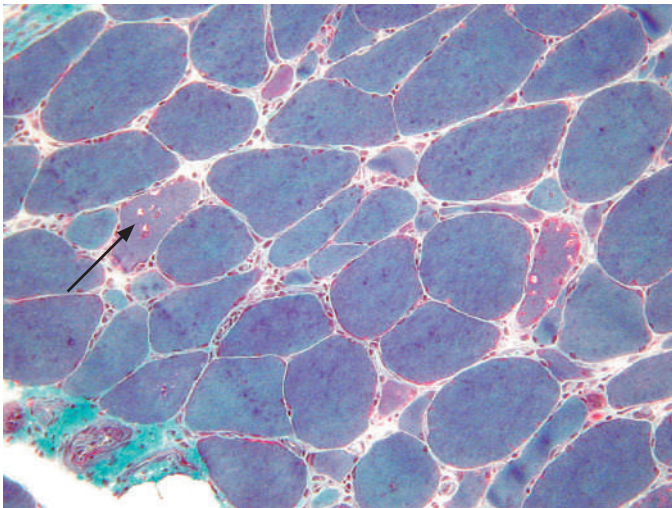
**Figure 4** Profound weakness of finger flexion – proximal upper limb strength nearly normal.



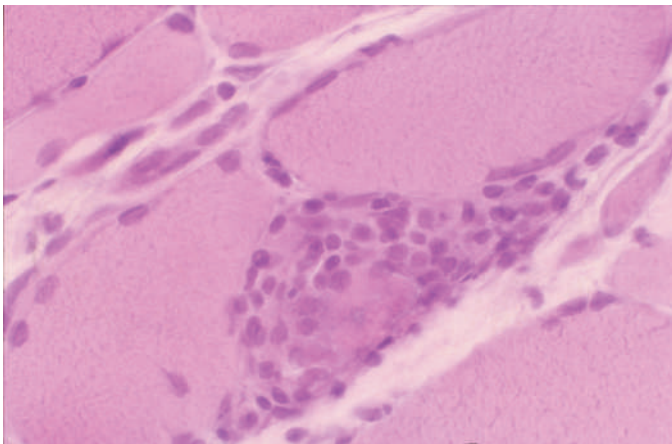
**Figure 5** (Haematoxylin & Eosin) Inflammatory infiltrate (black arrow) Atrophic fibres (red arrows).



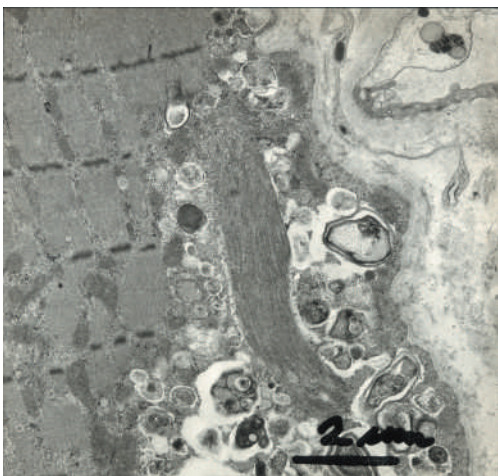
**Figure 6** (Gomori trichrome) Rimmed vacuoles.



**Figure 7** (Gomori trichrome) Rimmed vacuoles.



**Figure 8** (Haematoxylin & Eosin) Partial invasion by cytotoxic T cells.



**Figure 9** Electron microscopy 15 nm filaments.

riceps atrophy. It is the most common cause of so-called 'isolated quadriceps myopathy'. Unlike dermatomyositis (DM) and polymyositis (PM), distal weakness is an early feature and particularly affects the finger flexors (Fig. 4). Ankle dorsiflexion weakness presents as tripping. Also unlike DM and PM, the muscle involvement in IBM is often asymmetric.

Dysphagia may be an early symptom, or develop late in the course of the disease.

Some patients have an apparently related auto-immune disorder such as Sjogren's syndrome, and various nonspecific autoantibodies may be seen in up to 20% of cases of IBM.

### Diagnosis

The serum creatine kinase is usually moderately elevated. Electromyography shows 'myopathic' changes and 'irritative' phenomena (fibrillation potentials and positive sharp waves). None of these findings are specific and confirmation of diagnosis requires muscle biopsy.

### Treatment

Most patients show little or no response to immunosuppressant drugs or to intravenous immunoglobulin. Indeed, many cases of IBM were first considered to be 'steroid-resistant polymyositis'. A decision as to whether or not to attempt such treatment must be based on detailed discussion between the patient and a physician knowledgeable about the disease.

### The histological hallmarks of IBM

- Endomysial inflammatory infiltrates (predominantly cytotoxic T cells) (Fig. 5).
- Scattered or grouped round and angular fibres.
- Rimmed vacuoles (basophilic granular deposits around slit-like vacuoles) (Figs 6 and 7).
- Partial invasion of non-necrotic muscle fibres by cytotoxic T cells (Fig. 8).
- Eosinophilic cytoplasmic inclusions.
- Ragged red fibres (and mitochondrial DNA studies show multiple deletions).
- Congo red positive amyloid within or next to vacuoles.
- Bundles of 15 nm filaments in the cytoplasm or nuclei (Fig. 9).

### ACKNOWLEDGEMENTS

Pathological figures courtesy of Dr Waney Squier.