



Classification (Old)

- Idiopathic polymyositis (PM)
- Idiopathic dermatomyositis (DM)
- Dermatomyositis of childhood
- Myositis with neoplasm
- Myositis with other CTD

(Bohan & Peter, *NEJM* 1985)



Classification (New)

- Idiopathic polymyositis
- Idiopathic dermatomyositis
- Inclusion body myositis (IBM)

(Dalakas MC, *NEJM* 1993)



Incidence 1:100,000

DM

Age
children

PM

adults
> 18 years

IBM

> 50 years



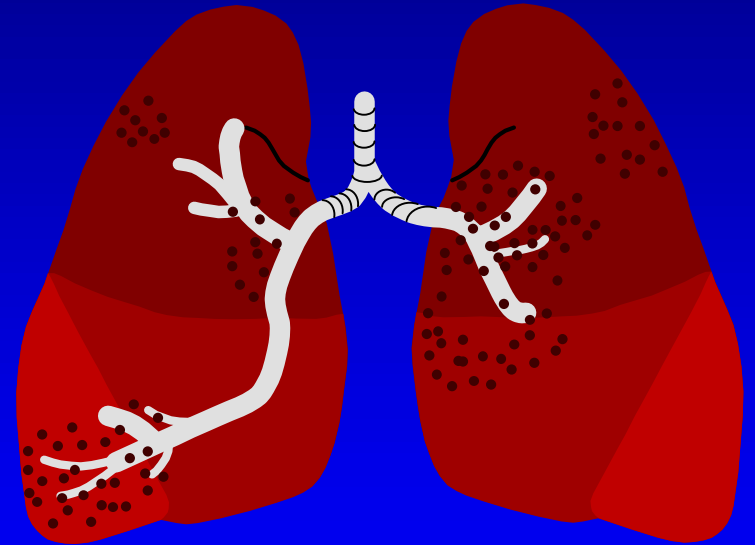


Clinical Features

- Slowly progressive proximal muscle weakness
- Myalgia rare
- Ocular and facial muscles spared
- Dysphagia
- Hypoventilation
- Cardiac abnormalities (carditis, conduction defects)

Pulmonary Involvement

- Interstitial lung disease
- Aspiration pneumonia
- Drug-induced pneumonitis
- Opportunistic infection
- Thoracic muscle weakness





Other Extramuscular Features

- Constitutional symptoms
- Raynaud's
- Subcutaneous calcifications (DM)



Dermatomyositis



Polymyositis

“Diagnosis of exclusion”

- No unique clinical features



Proximal Muscle Weakness

- Collagen vascular diseases
- Metabolic myopathies
- Drugs
- Infections
- Neurologic Diseases



Diagnosis

- Elevated muscle enzymes
 - CK
 - SGOT, LDH
- Myopathic EMG
 - Short duration, low amplitude polyphasic units
 - Increased spontaneous activity with fibrillation
 - Positive sharp waves



Histology

- Inflammatory cell infiltrate
- Phagocytosis, necrosis and regeneration of muscle fibres
- Scattered atrophic fibres



Dermatomyositis is *not*
Polymyositis with a rash



Polymyositis

- Endomysial infiltrate of inflammatory cells
- Necrosis of individual muscle fibres
- Cellular invasion of non-necrotic muscle fibres



Dermatomyositis

- Perivascular or interfascicular inflammatory infiltrates
- Endothelial hyperplasia and fibrin thrombi
- Capillary obliteration
- Perifascicular atrophy
- Necrosis of a group of muscle fibres
- Micro-infarcts



Inclusion Body Myositis (IBM)

- ▶ Myopathy
- ▶ Age >55
- ▶ Males > Females



IBM: Clinical Features

- Slowly progressive (asymmetric)
- Distal and proximal muscle weakness
- Dysphagia common and early
- Mixed myopathic and neuropathic EMG features
- CK - minimally elevated
- Poor response to immunosuppressive therapy

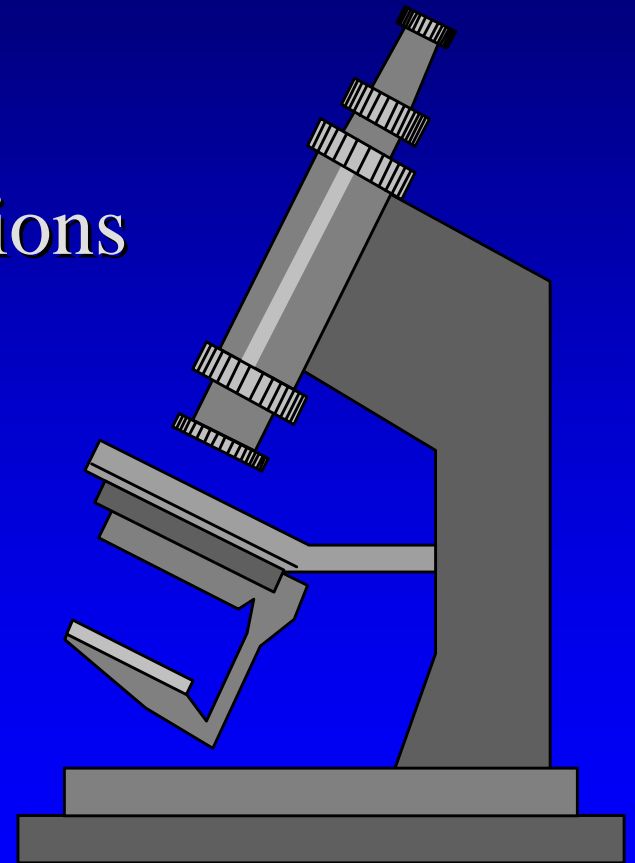


IBM: Histology

- Endomysial inflammation (CD8+ cells)
- Basophilic granular inclusions distributed around the edge of vacuoles
 - “rimmed vacuoles”

IBM: Electron Microscopy

- ▶ cytoplasmic filamentous inclusions





Muscle fibres contain:

- ▶ Ubiquitin
 - ▶ regulates protein degradation in cells
 - ▶ common in neurodegenerative disorders
- ▶ B-amyloid protein
 - ▶ similar to Alzheimer's



Malignancy and Inflammatory Muscle Disease

- Increased risk in DM (Callen, 1980)
- Increased risk in DM and PM (Gordon, 1985)
- Increased risk in DM and PM (Sigurgiersson, 1993)
- Increased risk in DM only (Airio, 1995)



Summary

- Dermatomyositis and polymyositis are different diseases
- Suspect inclusion body myositis in the older, atypical, or patient who is not responding as expected to treatment
- Malignancy is associated with dermatomyositis (? with PM)



Malignancy and Inflammatory Muscle Disease

MALES

FEMALES

DM

Relative risk	2.4 (CI 1.6-3.6)	3.4 (CI 2.4-4.7)
Mortality ratio	3.8 (CI 2.9-4.8)	

PM

Relative risk	1.8 (CI 1.1-2.7)	1.7(CI 1.0-2.5)
Mortality ratio	0.9 (CI 0.6-1.4)	



DM Malignancy Screen

- CXR
- Routine bloodwork
- Mammogram
- Abdominal/pelvic ultrasound
- Directed by symptoms, signs or abnormal tests



Case History

62 year old woman

- Dysphagia for 2-3 years
- Difficulty standing from a seated position for 1 year
- Hoarse voice
- 15 pound weight loss



Multiple Investigations

- ▶ EMG - February 1995
 - ▶ Peripheral neuropathy
- ▶ CCU admission : CK 800
- ▶ Discharge Dx : ?subendocardial infarct
- ▶ CK at discharge >800



Physical Examination

- No rash
- Weak proximal and distal muscles
- Chest clear



Laboratory

- ▶ Haemoglobin 110
- ▶ ESR 54
- ▶ CPK 1459
- ▶ ANA 1:320
- ▶ ENA positive
- ▶ EMG inflammatory myopathy
- ▶ CT chest ground glass lower lobes

Treatment of PM/DM

PHARMACOLOGIC

➤ Corticosteroids

- PREDNISONE 40-80 mg/day
 - divided dose initially, reduce only with remission

➤ Anti-Inflammatory/Immunosuppressive

➤ METHOTREXATE

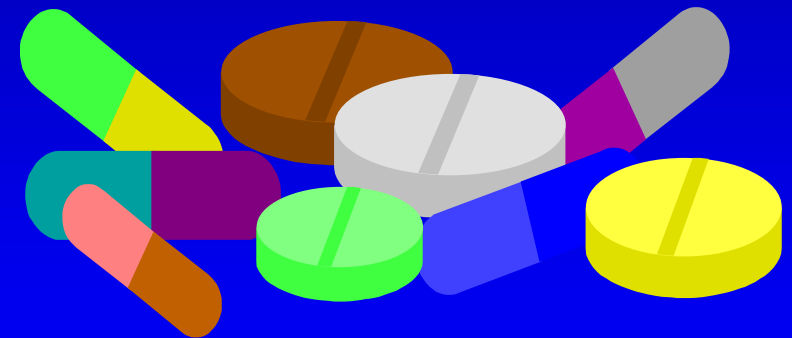
- PO/IM/IV
- Up to 50 mg/week

➤ IMURAN

- PO
- 2 mg/kg/day

➤ CYCLOPHOSPHAMIDE / CSA / PLASMAPHERESIS

➤ HCQ



NON-PHARMACOLOGIC



Immunopathology

	PM	DM
Cell	CD8+	CD4 +/-B
Damage	Cell med	Humoral
Target	Cell	Vessel
Mediator	MHC Class I	C5-9 MAC