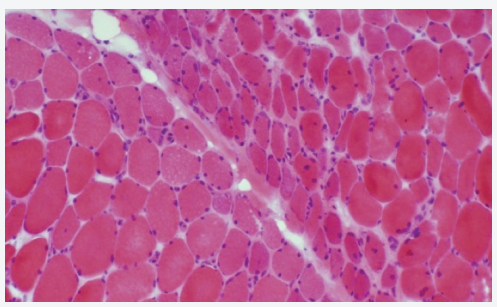
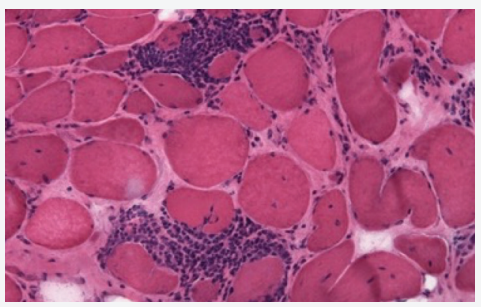
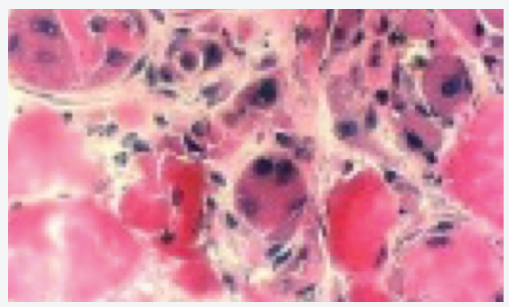
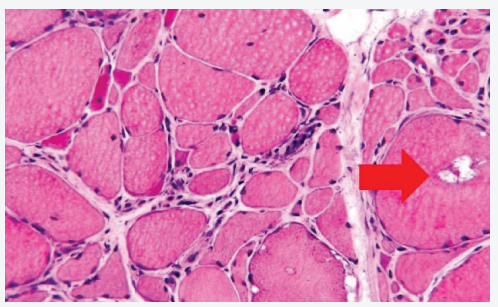
MANAGE DERMATOMYOSITIS

Features of dermatomyositis in comparison with other idiopathic inflammatory myopathies

	Dermatomyositis (DM)	Polymyositis (PM)	Necrotising autoimmune myopathy (NAM)	Sporadic-inclusion body myositis (sIBM)
Onset and disease course	• Acute or insidiously progressive, symmetrical, proximal **muscle weakness** and/or a characteristic **skin rash**[2] • Usually painless[2]	• Progressive onset of symmetrical, proximal muscle weakness[2] • Commonly painful[2]	• Subacute onset, progressive symmetrical, proximal muscle weakness[1,2] • Weakness develops more rapidly than DM and PM, and is more severe[2,3]	• Progresses over several years[2] • Unique versus other IIMs as it affects both the proximal and distal musculature in a symmetrical/asymmetrical pattern[2]
Age at onset	• Children (Juvenile dermatomyositis)[3] • Adults[3]	• Adults (usually)[2]	• Adults[2]	• Adults > 40 years old[2]
Symptoms, specific features	• +/- dysphagia, specific **skin and organ manifestation**[2,3] • Rash may precede onset of muscular weakness[1,2] • Involvement of pulmonary system e.g. interstitial lung disease[1,2,3] • Malignancy in adults[2,3] • Heliotrope rash: reddish purple rash on or around the eyelids[2] • V-sign: Rash on anterior chest[2] • Gottron's sign[1], Gottron's papules[2] • Shawl sign: rash on neck, back and shoulders[2] • Inverse Gottron's papules on the volar aspect of hands, dilated capillary loops at the nail beds with periungual telangiectasias and thickened, cracked skin on the dorsal and ventral surfaces of the hands (mechanic's hands)[2]	• **No rash**[2] • Muscular and extramuscular organ involvement similar to DM[2]	• **No rash**[1,2] • Myalgias, dysphagia may occur[2] • Creatinine kinase usually higher than in other IIMs[1,2] • Extramuscular manifestations include congestive heart failure[2,3]	• **No skin changes**[3] • Weakness in flexor forearm muscles Quadriceps weakness leads to falls/ tripping[2] • Dysphagia very common[2] • Mild facial weakness common[2]
Muscle pathology	• **B cells** and **CD4+ T cells** in perimysial and perivascular areas[2] • **Perifascicular muscle fibre atrophy**[2] • Membranolytic attack complex **(MAC) deposition** in microvasculature[2]	• Invasion of endomysial **cytotoxic CD8+ T cells** in muscle fibres[2] • **Macrophages** invade non-necrotic muscle fibres[2]	• **Macrophage-mediated** immune response[2] • **Necrotic myofibres** surrounded by sparse inflammatory infiltrate (predominantly lymphocytes)[2] • **MAC deposition** in microvasculature[2]	• Similar to PM, invasion of muscle fibres by **cytotoxic CD8+ T cells** and **macrophages**[2] • **Rimmed vacuoles** characteristic of degenerative changes[2] • **Amyloid deposits**[2]
Treatment response	• Usually **responsive** to **immunotherapies**[2,3]	• Usually **responsive** to **immunotherapies**[2,3]	• Usually **responsive** to **immunotherapies**[2,3]	• Usually **refractory** to **immunotherapies**[2]
Histological section (images courtesy of Prof. Patrick Cherin)				

1. Goyal NA. Continuum (MinneapMinn) 2019; 25:1564-85.
2. Malik A et al. Front Neurol 2016; 7:64.
3. Schmidt J. J Neuromuscul Dis 2018; 5:109-29.