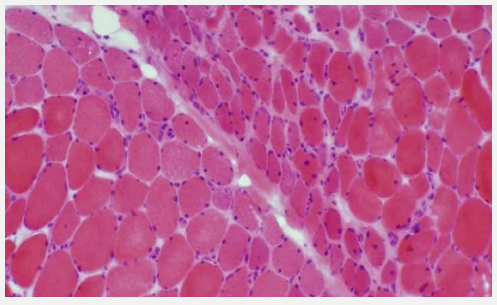
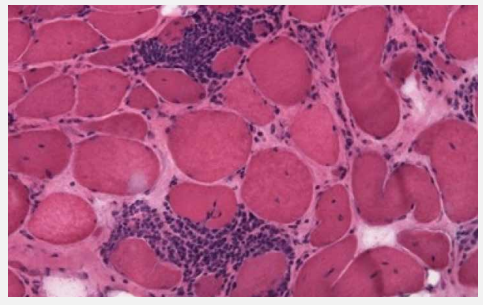
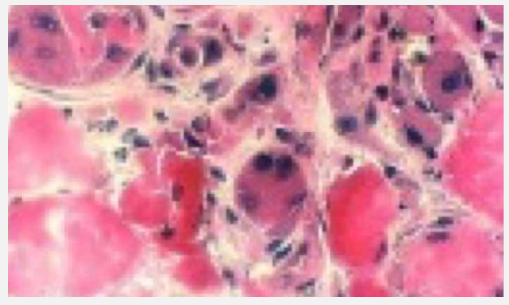
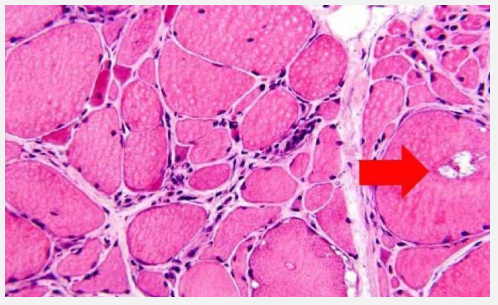
MANAGE DERMATOMYOSITIS

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[1] • Usually painless[1]	• Progressive onset of symmetrical, proximal muscle weakness[1] • Commonly painful[1]	• Subacute onset, progressive symmetrical, proximal muscle weakness[1,2] • Weakness develops more rapidly than DM and PM, and is more severe[1,3]	• Progresses over several years[1] • Unique versus other IIMs as it affects both the proximal and distal musculature in a symmetrical/asymmetrical pattern[1]
Age at onset	• Children (Juvenile dermatomyositis)[3] • Adults[3]	• Adults (usually)[1]	• Adults[1]	• Adults > 40 years old
Symptoms, specific features	• +/- dysphagia, specific skin and organ manifestation[1,3] • Rash may precede onset of muscular weakness[1,2] • Involvement of pulmonary system e.g. interstitial lung disease[1,2,3] • Malignancy in adults[1,3]	• No rash[1] • Muscular and extramuscular organ involvement similar to DM[1]	• No rash[1,2] • Myalgias, dysphagia may occur[1] • Creatinine kinase usually higher than in other IIMs[1,2] • Extramuscular manifestations include congestive heart failure[1,3]	• No skin changes[3] • Weakness in flexor forearm muscles Quadriceps weakness leads to falls/ tripping[1] • Dysphagia very common[1] • Mild facial weakness common[1]
Muscle pathology	• B cells and CD4+ T cells in perimysial and perivascular areas[1] • Perifascicular muscle fibre atrophy[1] • Membranolytic attack complex (MAC) deposition in microvasculature[1]	• Invasion of endomysial cytotoxic CD8+ T cells in muscle fibres[1] • Macrophages invade non-necrotic muscle fibres[1]	• Macrophage-mediated immune response[2] • Necrotic myofibres surrounded by sparse inflammatory infiltrate (predominantly lymphocytes)[1] • MAC deposition in microvasculature[1]	• Similar to PM, invasion of muscle fibres by cytotoxic CD8+ T cells and macrophages[1] • Rimmed vacuoles characteristic of degenerative changes[1] • Amyloid deposits[1]
Treatment response	• Usually responsive to immunotherapies[1,3]	• Usually responsive to immunotherapies[1,3]	• Usually responsive to immunotherapies[1,3]	• Usually refractory to immunotherapies[1]
Histological section (images courtesy of Prof. Patrick Cherin)				

1. Malik A et al. Front Neurol 2016; 7:64.
2. Goyal NA. Continuum (MinneapMinn) 2019; 25:1564-85.
3. Oddis CV & Aggarwal R. Nat Rev Rheumatol 2018; 14:279-89.