



POLYMYOSITIS

DERMATOMYOSITIS

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Definition



PM and DM are autoimmune disorders that cause nonsuppurative muscle inflammation

Epidemiology



- **incidence of : 1 in 100,000**
- **Age:PM is predominantly a disease of adults. DM affects both children and adults**
- **Sex:women more often than men**

Classification

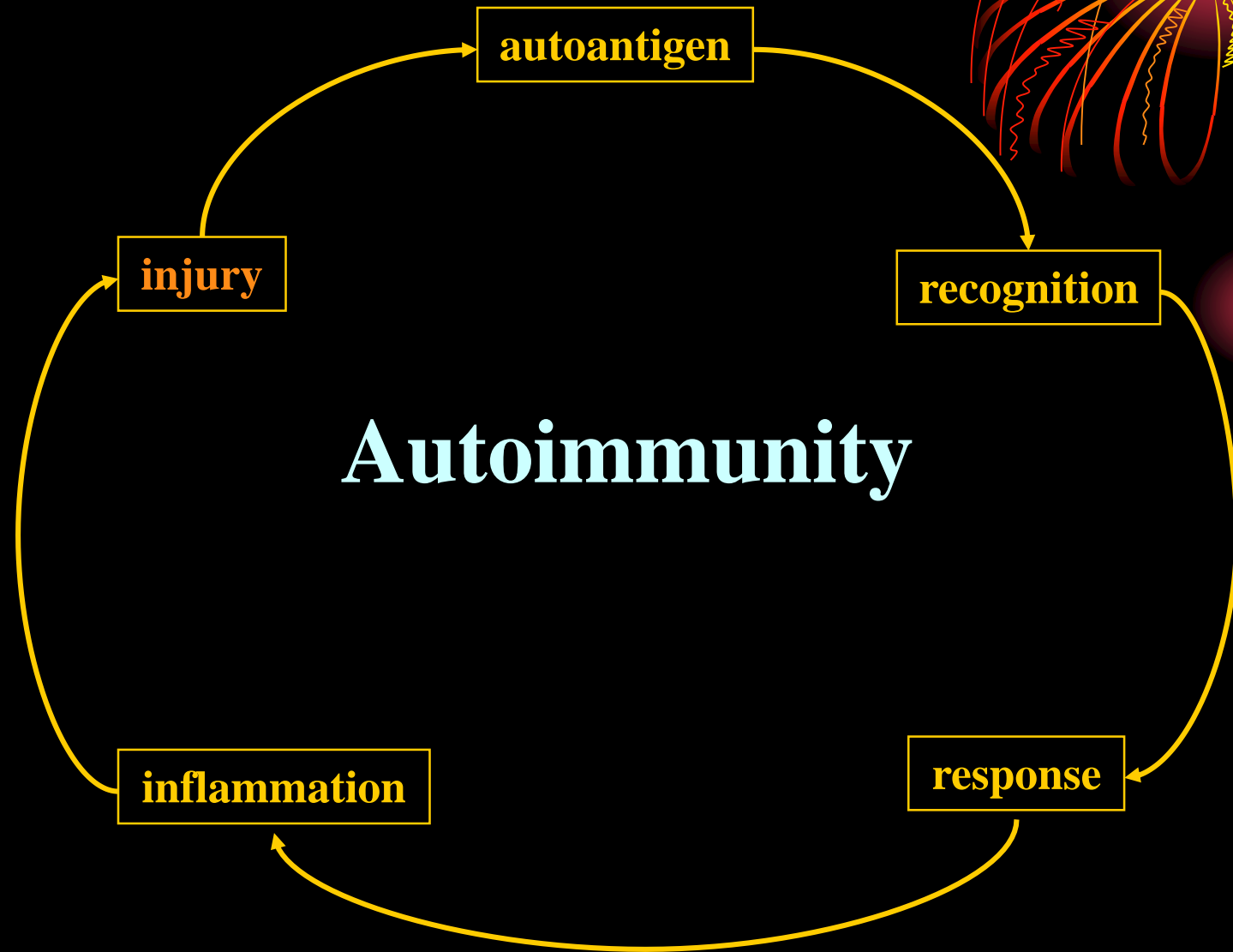


- **Adult PM**
- **Adult DM**
- **PM/DM associated with malignancy**
- **Childhood DM**
- **PM/DM associated with other CTD**
- **IBM(Inclusion body myositis)**

pathphysiology



- **autoimmune**
- **B cell-mediated: Autoantibodies**
- **T cell-mediated myocytotoxicity**
- **Tissue injury**



Autoimmunity

injury

autoantigen

recognition

response

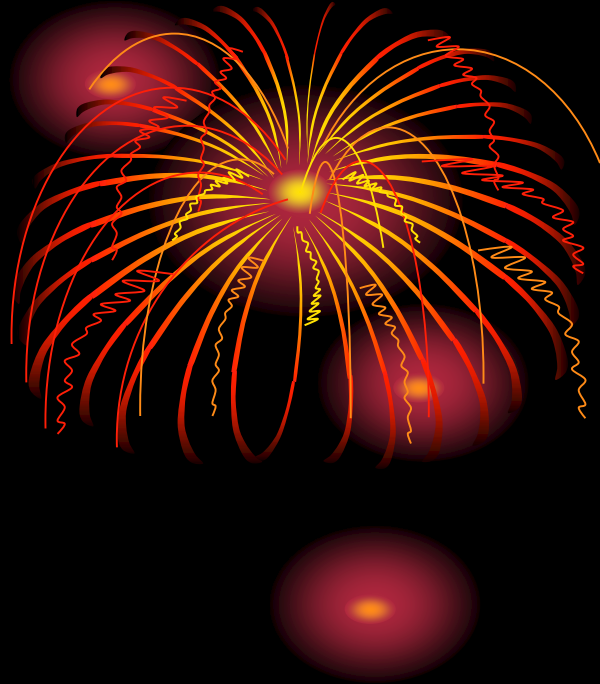
inflammation

Autoantibodies



- **ANA**
- **Antisynthetase(anti-Jo-1)**
- **Anti-SRP**
- **Anti-Mi-2**
- **Anti-RNP**

Clinical



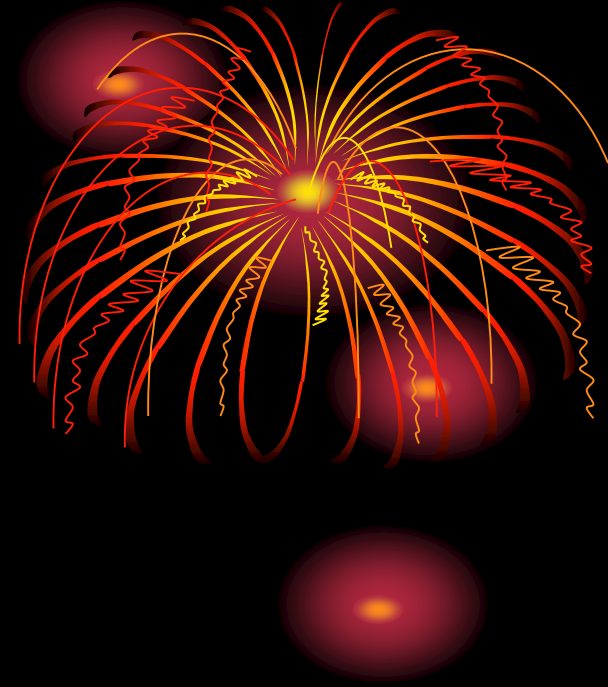
Weakness

Proximal weakness: the most common site of weakness in a myopathic disorder

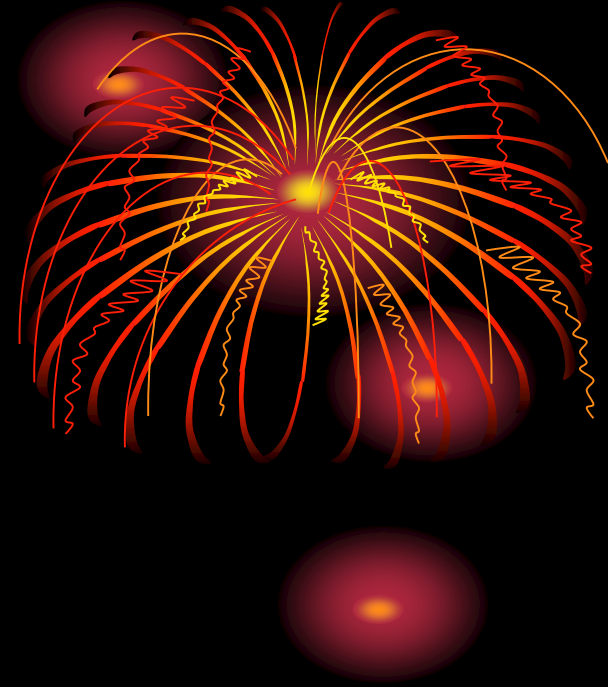
- Lower extremities
 - difficulty climbing stairs
 - arising from a low chair or toilet,
 - getting up from a squatted position.
 - Abnormal gait(waddling gait)
- Upper extremities are involved,
 - trouble lifting objects over their head
 - brushing their hair.



Myalgia

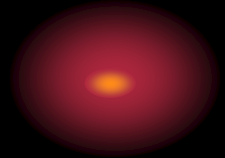


Skin



Skin

- Heliotrop rash

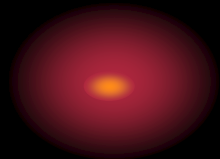
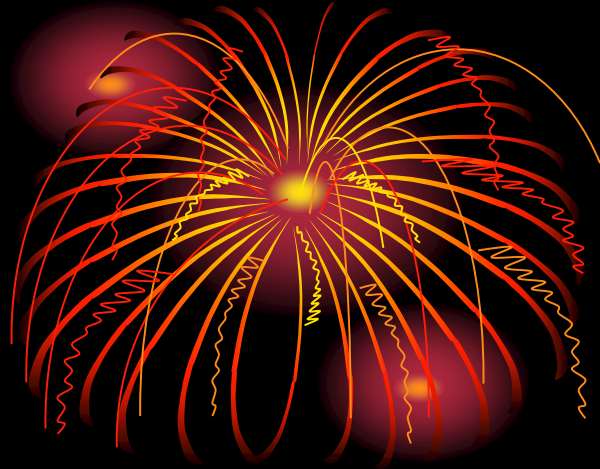


Dermatomyositis: heliotrope rash



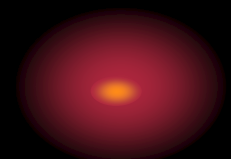
Dermatomyositis: rash, chest





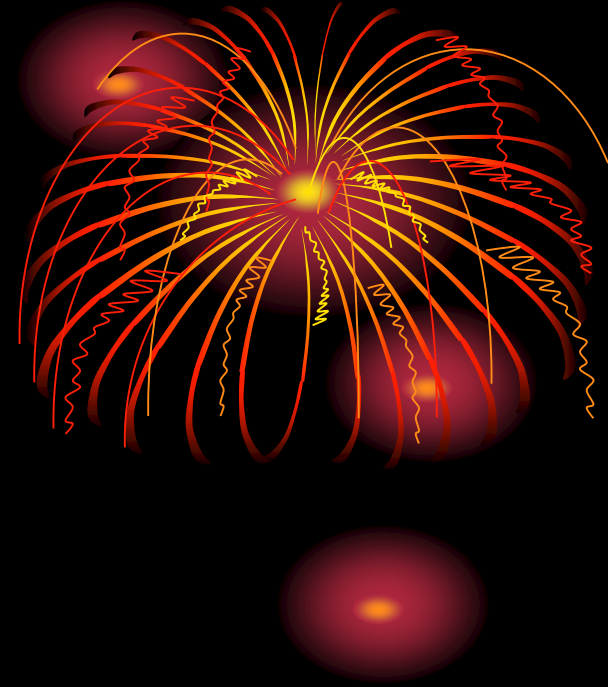
Skin

- Gottron's papules



Skin

- Shawl-sign rash



Skin

- Mechanic's hands



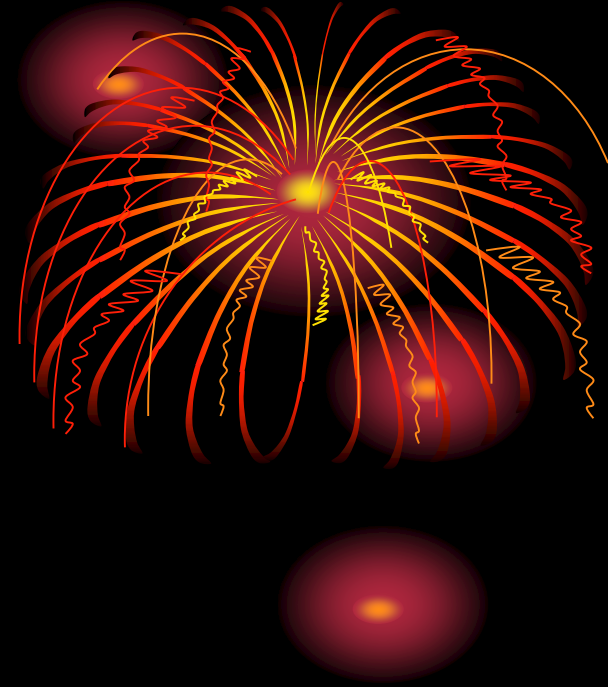
Skin

- Subcutaneous calcification



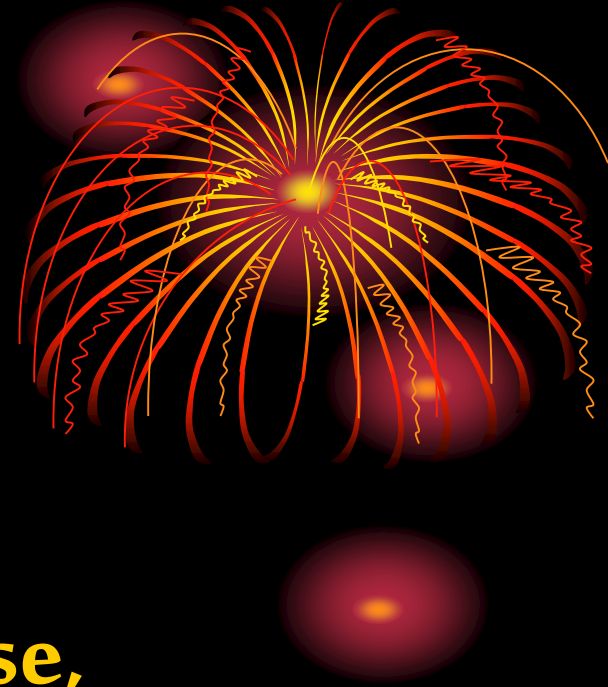
Extramuscular Manifestations

- *Systemic symptoms*
- *Arthritis*
- *gastrointestinal: Dysphagia*
- *Cardiac*
- *Pulmonary dysfunction*
- *Malignancies: 15%*



Lab.test

- **CBC , ESR**
- **Muscle enzyme:CPK,Aldolase,
SGOT,LDH,SGPT**
- **Autoantibodies**



EMG -NCV



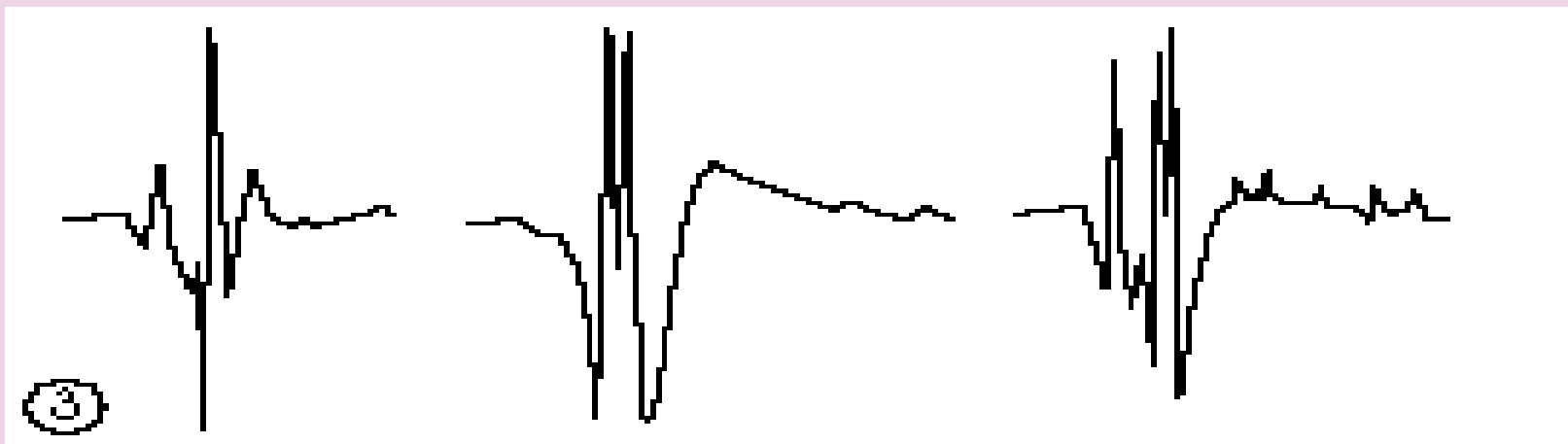
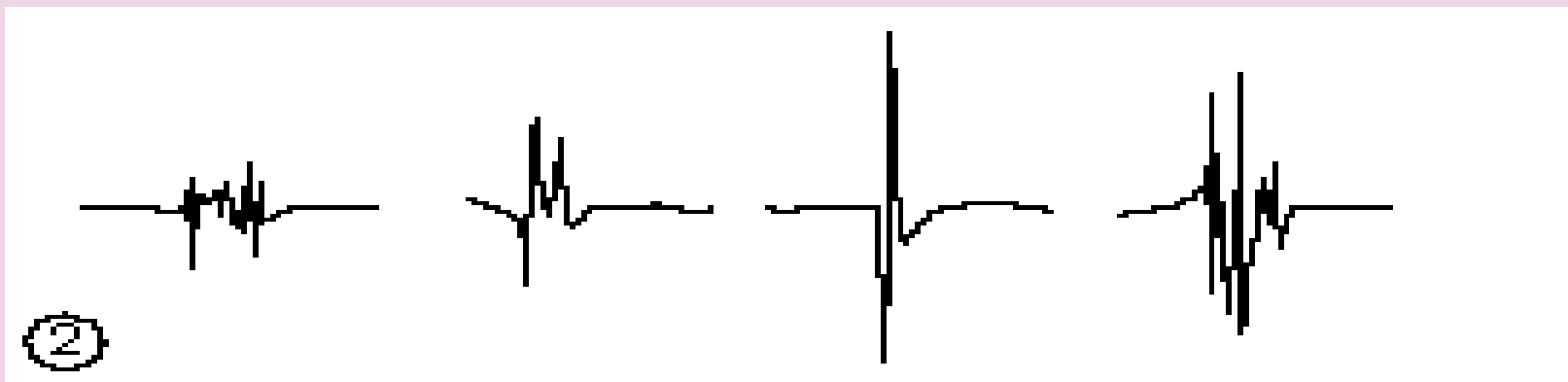
- *myopathic potentials characterized*

by:

short-duration,

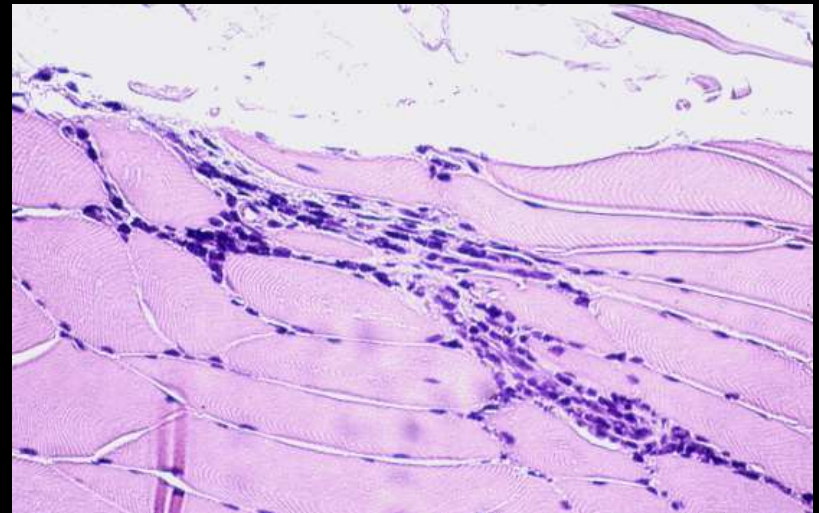
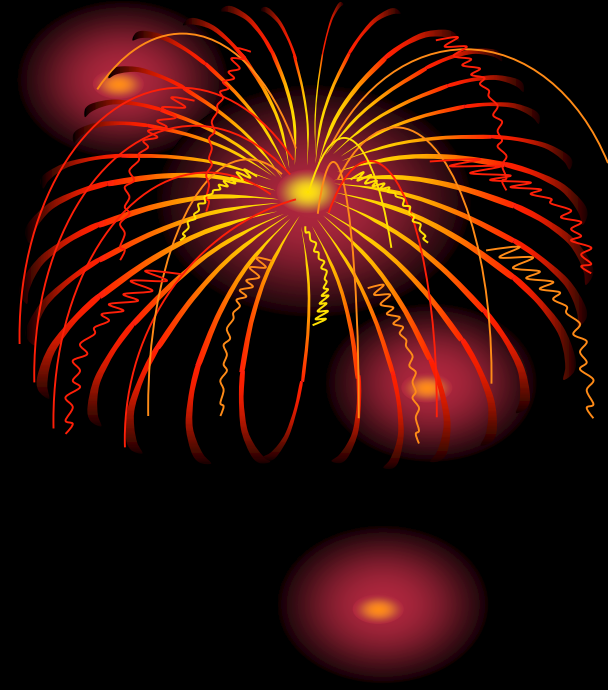
low-amplitude

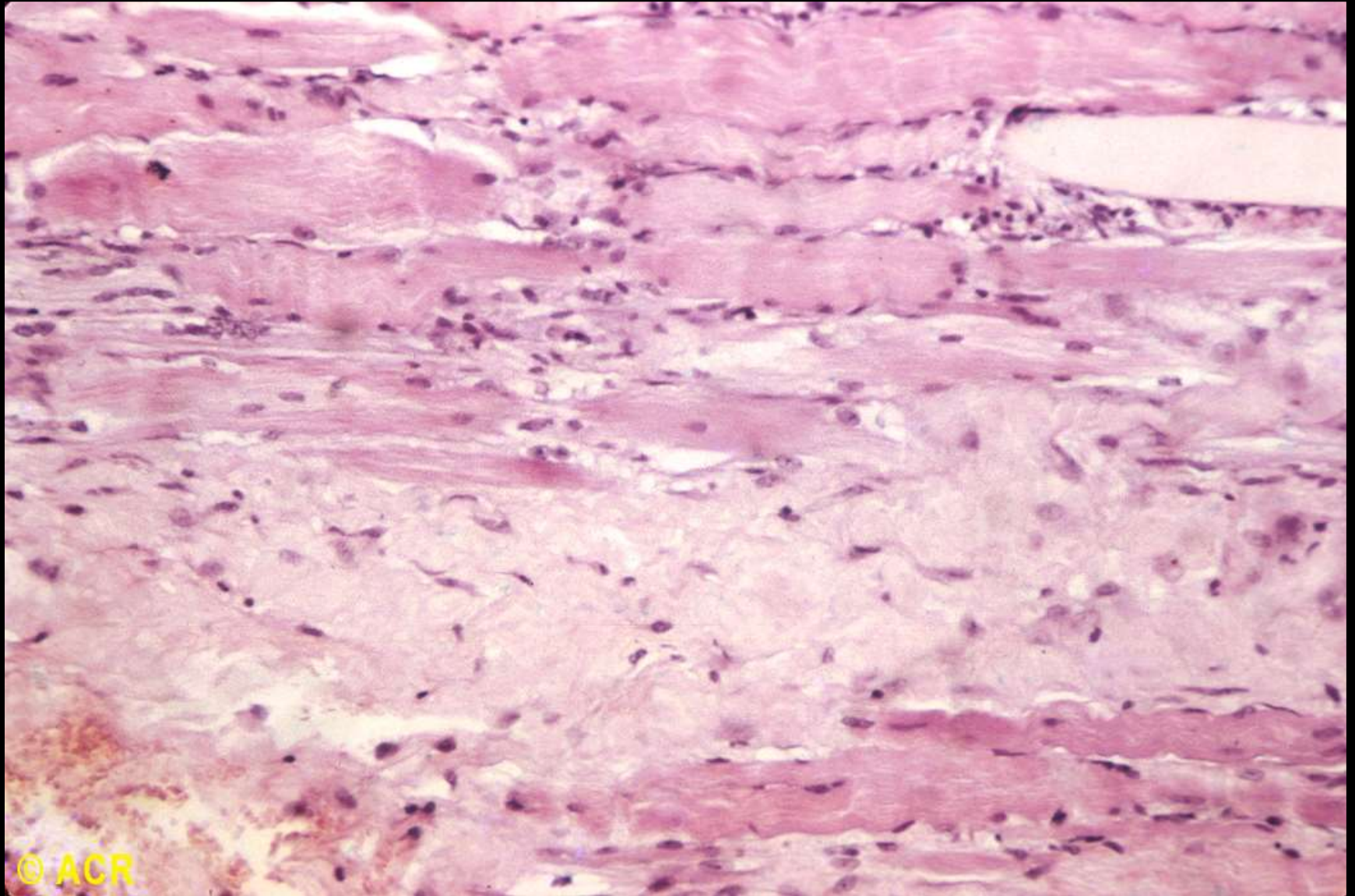
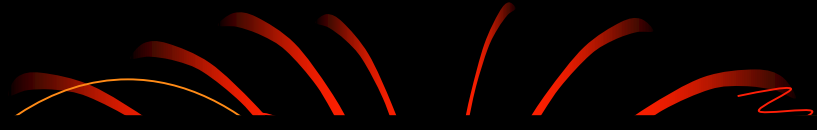
polyphasic units



Pathology

- **endomysial inflammation**
- **muscle fibers undergo necrosis, degeneration, and phagocytosis**

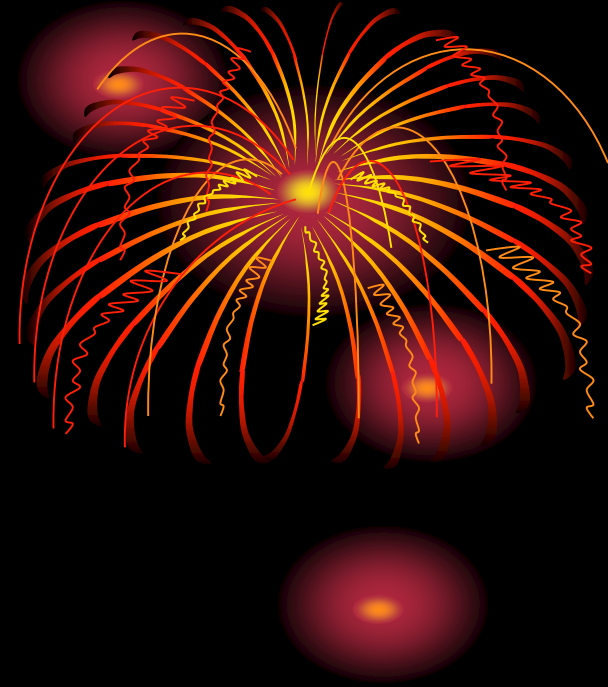




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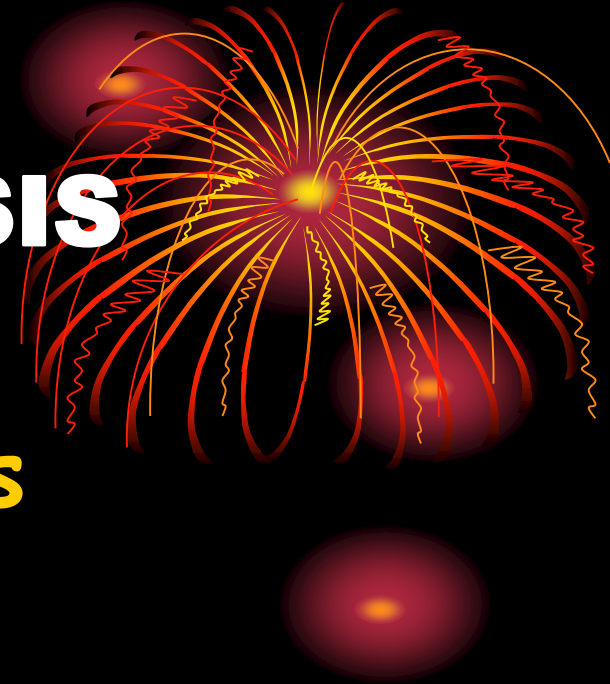
Diagnosis

- **Muscle weakness**
- **Rash**
- **Muscle enzyme**
- **EMG**
- **Histology**



DIFFERENTIAL DIAGNOSIS

- Neuromuscular diseases
 - Muscular dystrophies
 - Neuromuscular junction Di.
- Endocrine disorders
 - Hypothyroidism
 - Hyperthyroidism
 - Acromegaly
 - Cushing's di.



DIFFERENTIAL DIAGNOSIS



- Drugs and toxins
- Metabolic disorders
 - storage di.
 - Nutritional di.
 - Electrolyte disorders
- Miscellaneous
 - Other rheumatic disorders
 - Malignancy
 - Organ failure

CLINICAL FEATURES OF INCLUSION BODY MYOSITIS

1. Insidious yet progressive proximal and distal muscle atrophy and weakness
2. Affects predominantly elderly male population
3. Rare or no association with malignancy or other connective tissue diseases
4. Creatine kinase normal or only minimally elevated (usually <5–6 times normal)
5. Mixed myopathic and neuropathic electromyographic features
6. Resistance to corticosteroids and immunosuppressive drugs

Treatment

- **Corticosteroids**
- **Immunosuppressive agents**

