Idiopathic Inflammatory Myopathies; Clinical Manifestation

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IUMS

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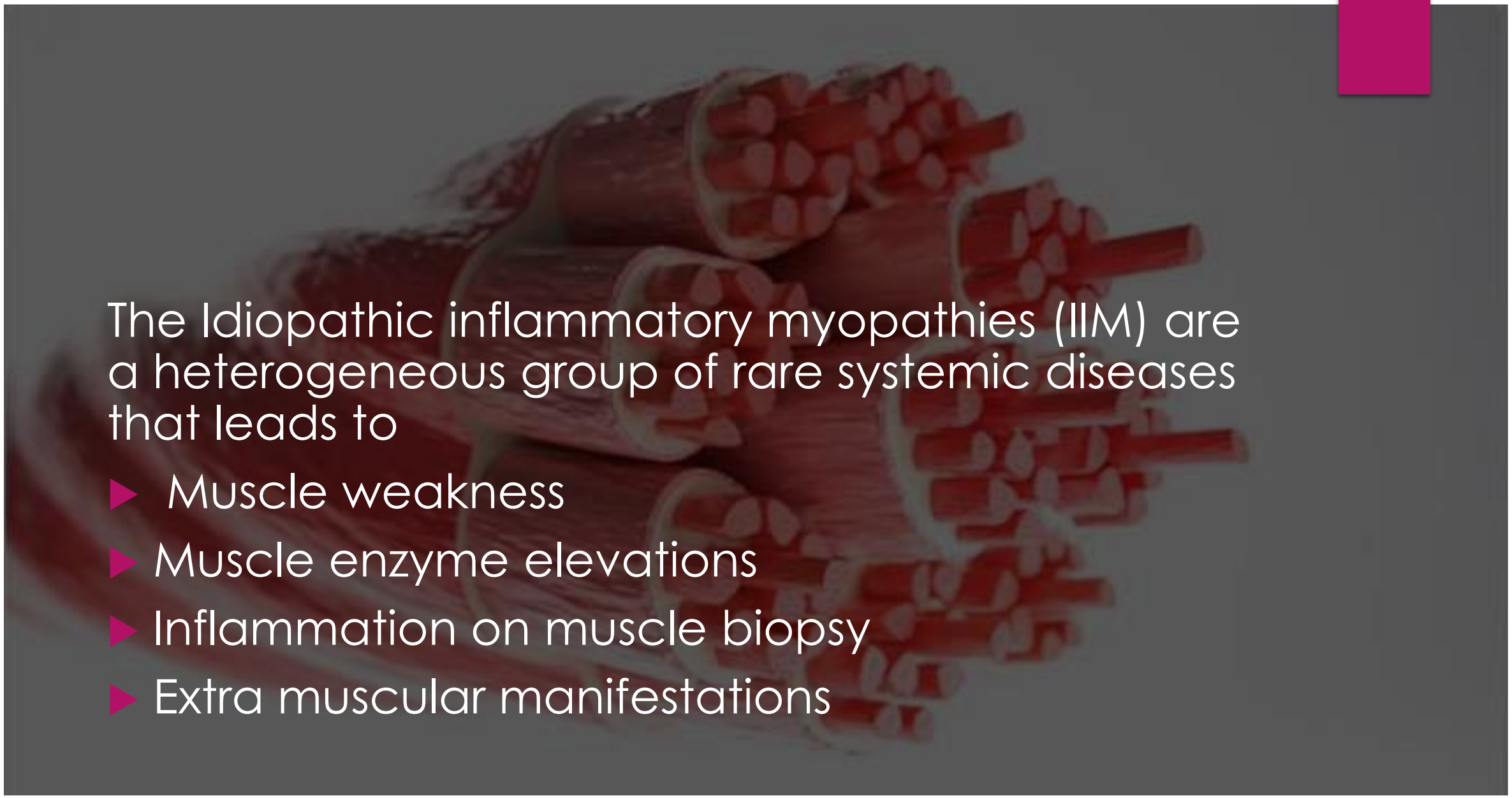


The Idiopathic inflammatory myopathies (IIM) are a heterogeneous group of rare systemic diseases that leads to

- Muscle weakness
- Muscle enzyme elevations
- Inflammation on muscle biopsy
- Extra muscular manifestations

Classification of IIM

- Inclusion body myosositis (IBM)
- Immune mediated necrotizing myopathies (IMNM)
- Dermatomyositis (DM)
- Anti synthetase syndrome (ASyS)

Secondary IIM

Polymyositis (PM)

Classification of IIM

- Inclusion body myosositis (IBM)
- Immune mediated necrotizing myopathies (IMNM),
- Dermatomyositis (DM)
- Anti synthetase syndrome (ASyS)

First Classification of IIM

- Dermatomyositis (DM)
- Polymyositis(PM)
- Inclusion body myositis (IBM)

Muscular Manifestations

Muscular Manifestations

- **Bilateral, symmetric, proximal limb weakness**
- **Weakness of axial muscles**
- **Pharyngeal muscle weakness**

In all forms of inflammatory myopathy, pharyngeal and neck-flexor muscles are often involved, causing dysphagia or difficulty in holding up the head (head drop).

Muscular Manifestation

- Myalgia and muscle tenderness may occur in a small number of patients, usually early in the disease, and particularly in DM associated with connective tissue disorders

- In advanced and rarely in acute cases, respiratory muscles may also be affected.

Muscular Manifestation

- Severe weakness, if untreated, is almost always associated with muscle wasting.

Muscular Manifestation

Progressive and symmetric muscle weakness except for IBM, which can have an asymmetric pattern.

Difficulty with everyday tasks requiring the use of proximal muscles, such as getting up from a chair, climbing steps, stepping onto a curb, lifting objects, or combing hair

Muscular Manifestations

Fine-motor movements that depend on the strength of distal muscles, such as buttoning a shirt, sewing, knitting, or writing, are affected only late in the course of PM and DM, but fairly early in IBM

Muscular Manifestation

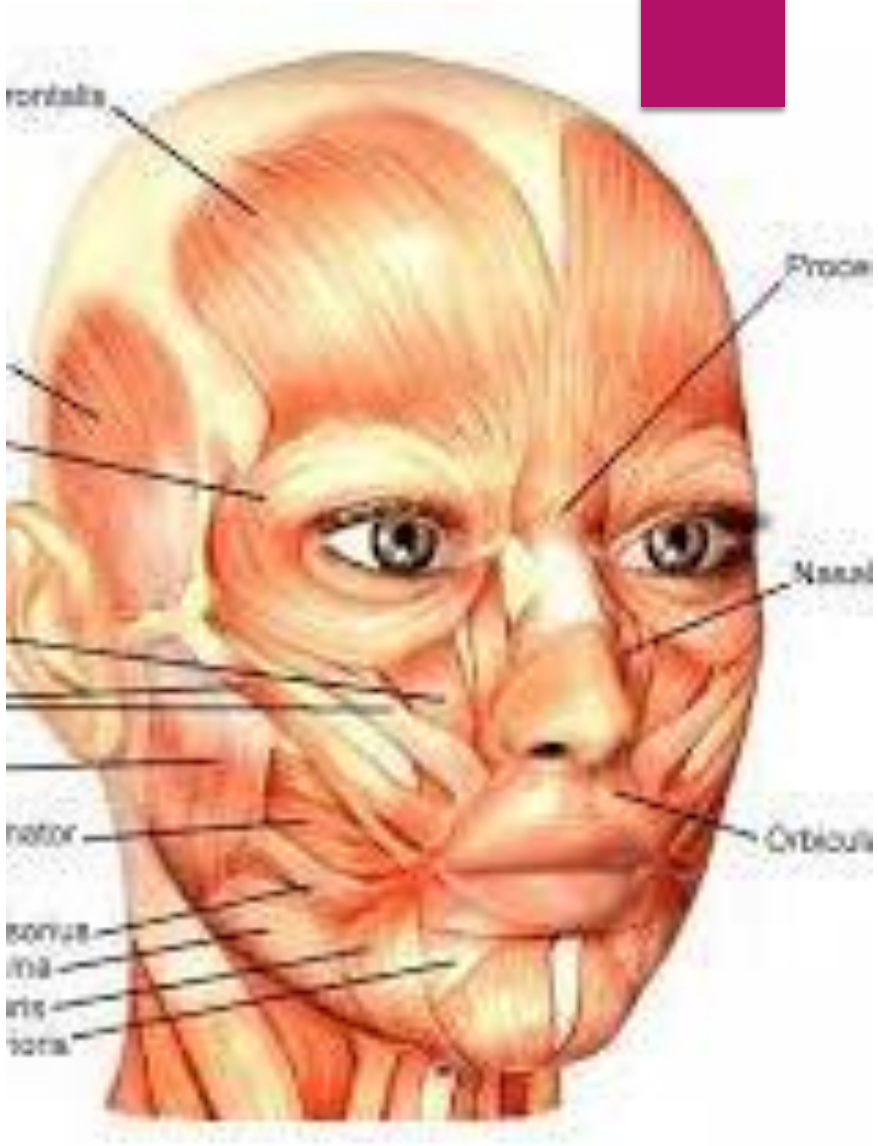
- Falling is common in IBM because of early involvement of the quadriceps muscle

Ocular muscles are spared, even in advanced, untreated cases

If these muscles are affected, the diagnosis of inflammatory myopathy should be questioned

Muscular Manifestation

- Facial muscles are unaffected in PM and DM, but mild facial muscle weakness is common in patients with IBM.



Extramuscular Manifestations

Systemic Symptoms

- Fever
- Malaise
- Weight loss

- **Especially when inflammatory myopathy is associated with a connective tissue disorder**

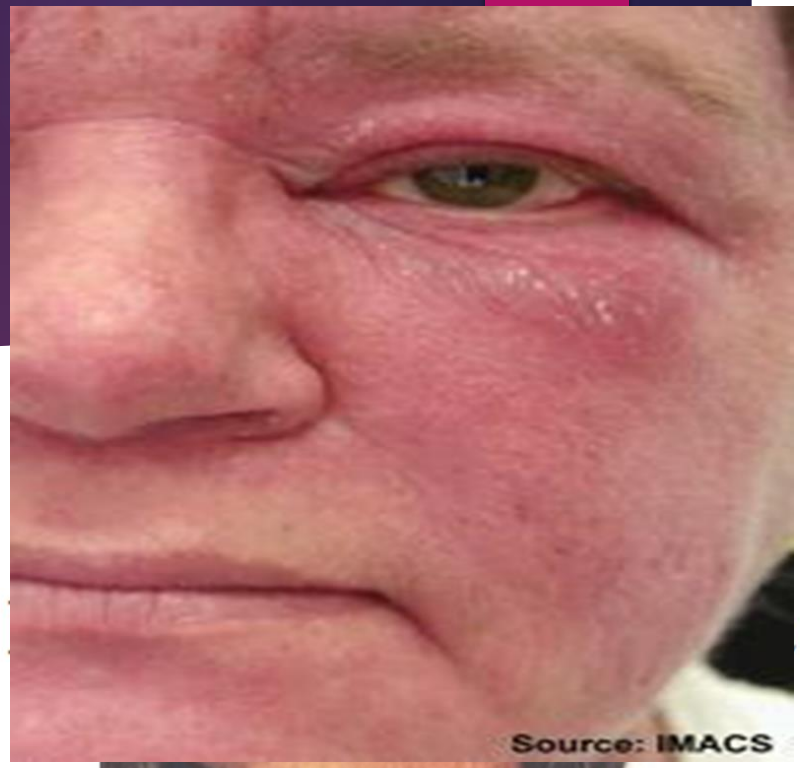
Skin Manifestation

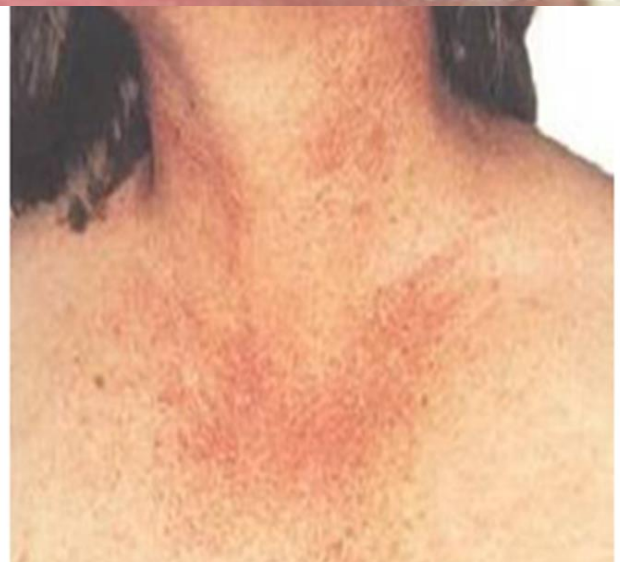
Skin Manifestation

- The skin lesions are frequent in IIM(>20%) and maybe observed in DM and ASyS
- Skin changes are key for the diagnosis of DM
- Skin changes can precede the muscular weakness 3-6 months

Skin Manifestations

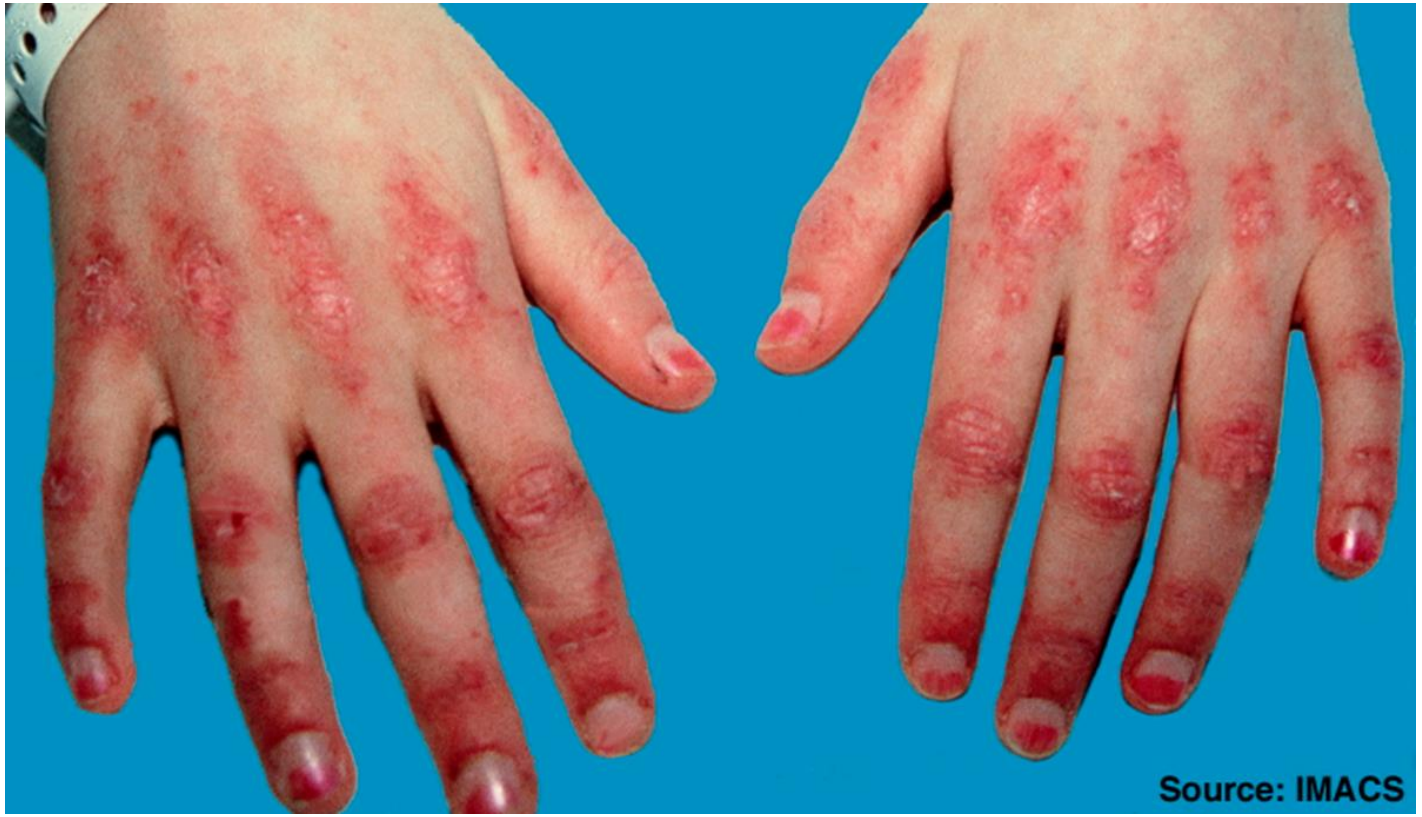
The sites of predilection
Upper eyelids (Heliotrope rash)
Malar areas
Bridge of nose
Nasolabial folds
"V" area of anterior chest (V sign)
Dorsum MCPs & PIPs
Periungual areas





Gottron's Papules

Scaly erythematous papules over the extensor side of finger joints.

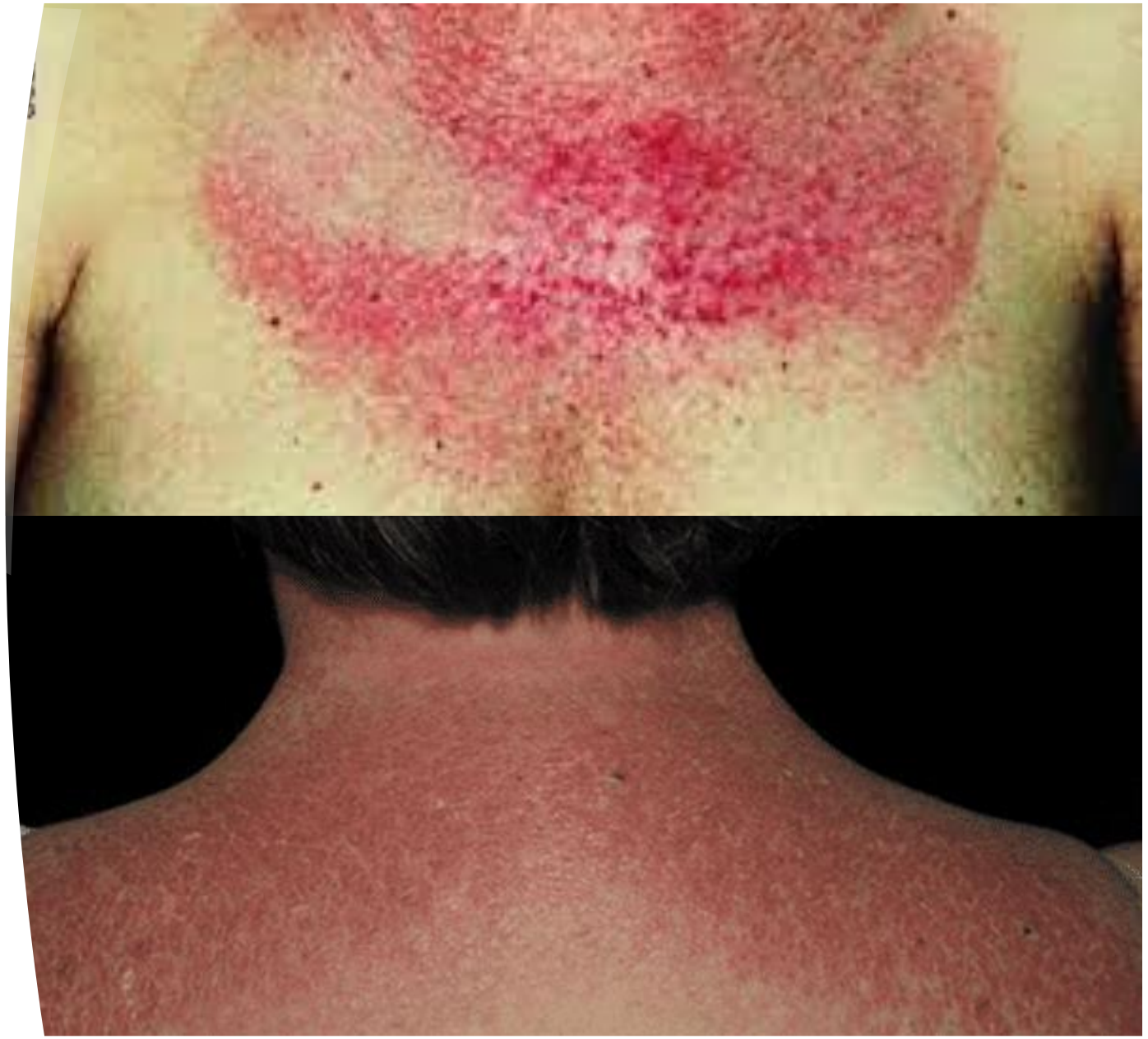


Heliotrope Rash

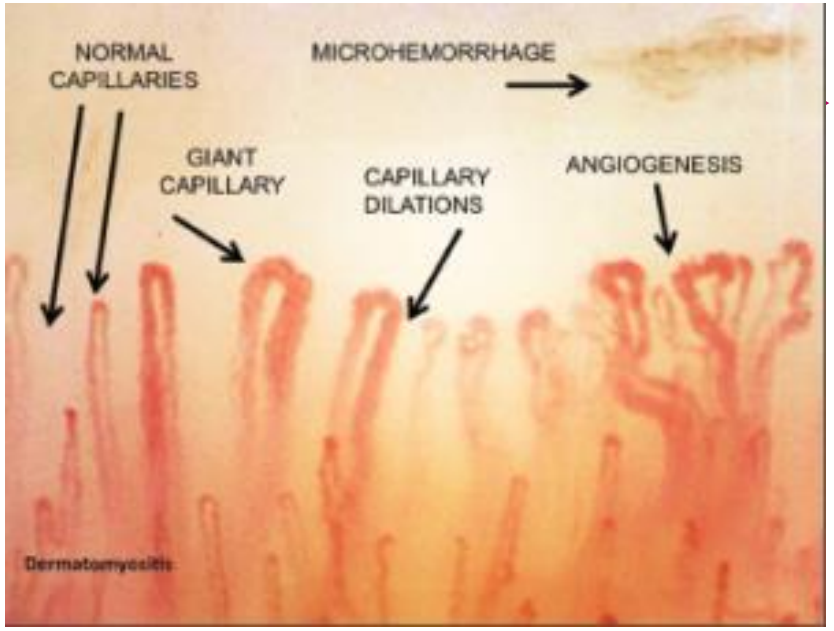
- lilac discoloration or a violaceous to dusky erythematous rash
- With or without edema in a symmetric distribution involving the periorbital skin and usually the upper eyelid.

Erythema

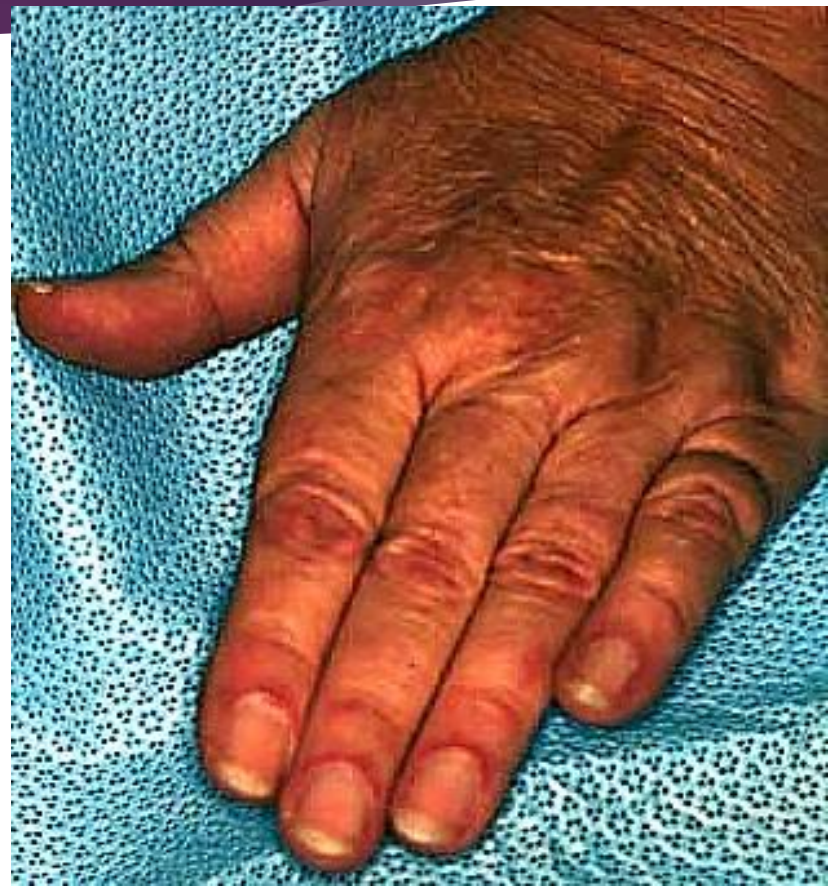
- Macular violaceous erythema with symmetric distribution
- **V sign**
- **Shawl sign**



Periungual Erythema



Nail fold capilleroscopy of periungual erythema shows dilated capillary loops.

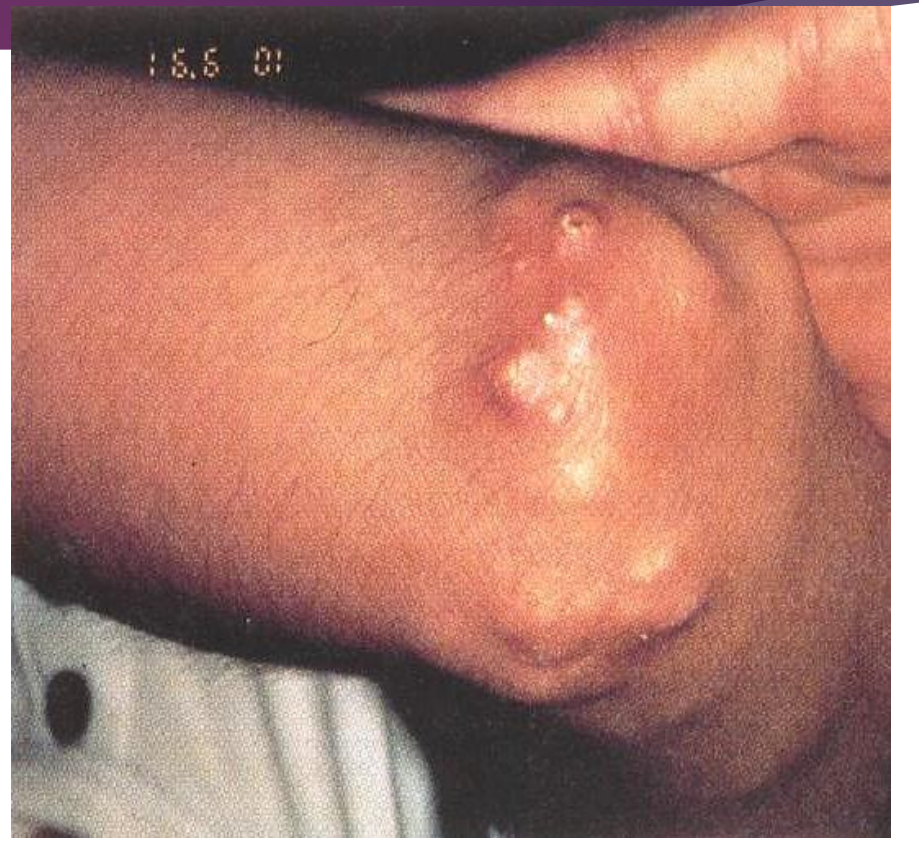


Calcinosis

Late problem

Calcinosis: firm papules or nodules which may centrally ulcerate releasing a chalky material,

Sites of repeated microtrauma



Visceral Involvement

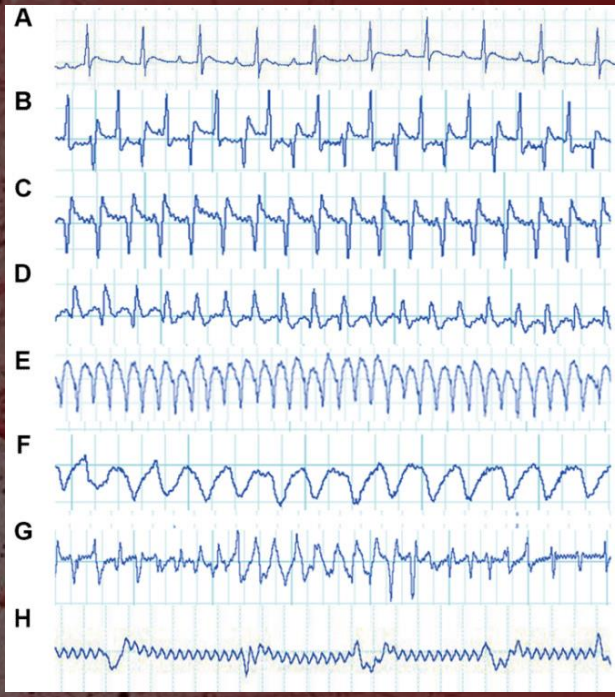
Visceral Involvement

- Interstitial Lung Disease
- Carditis (Arrhythmia, CHF, Pericarditis)
- Peripheral Vasculature (Raynaud's phenomenon)
- Gastrointestinal involvement (Dysphagia, Vasculitis)

Pulmonary Dysfunction

- Interstitial lung disease
- Anti-MDA5 DM and ASyS are the most frequent myositis associated with ILD (respectively 90% and 80%).
- Diapragmatic dysfunction due to weakness of the thoracic muscles,
- Aspiration pneumonia

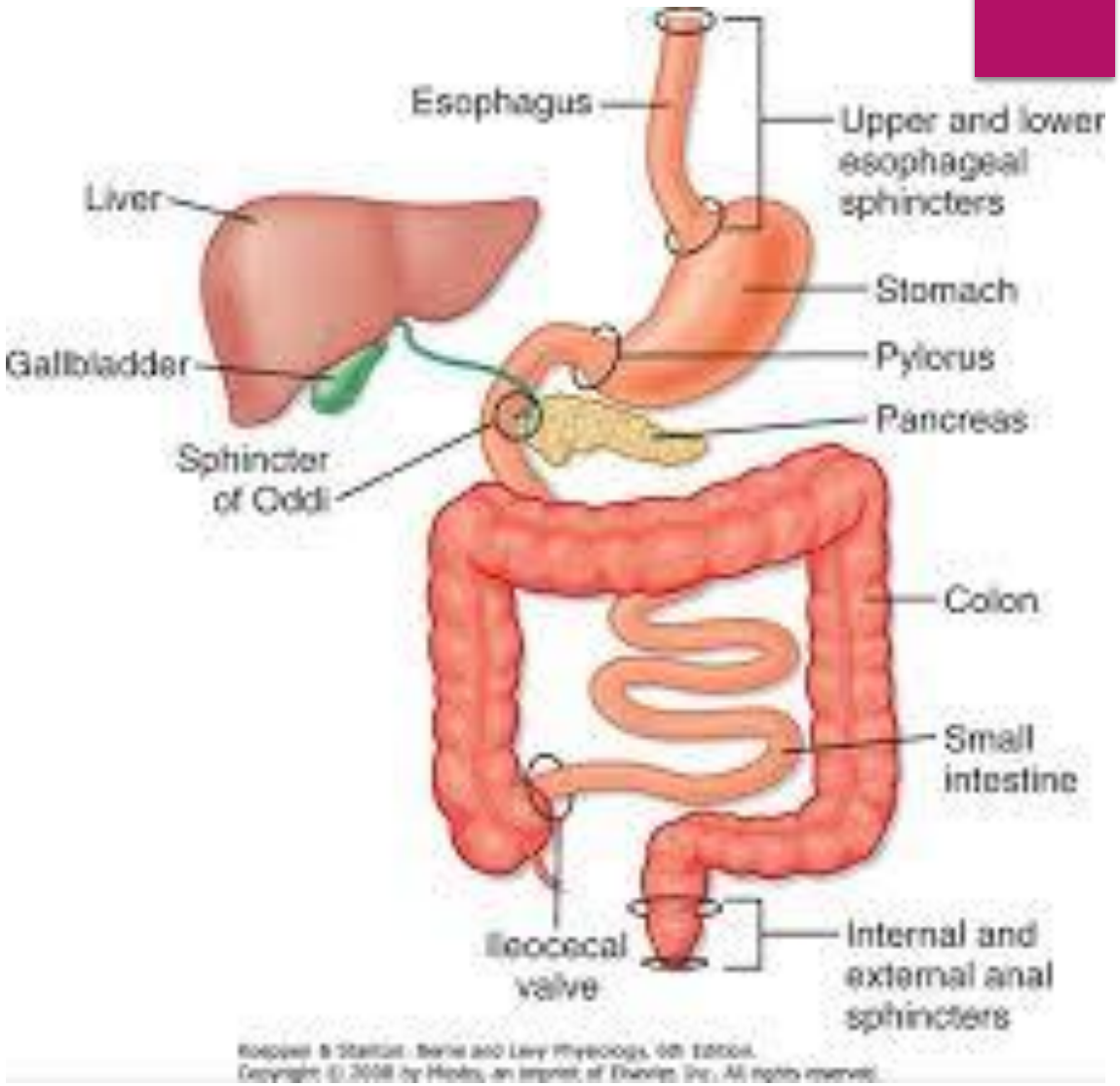
Cardiac Disturbances



- Atrioventricular conduction defects
- Tachyarrhythmias
- Dilated cardiomyopathy
- Low ejection fraction, congestive heart failure

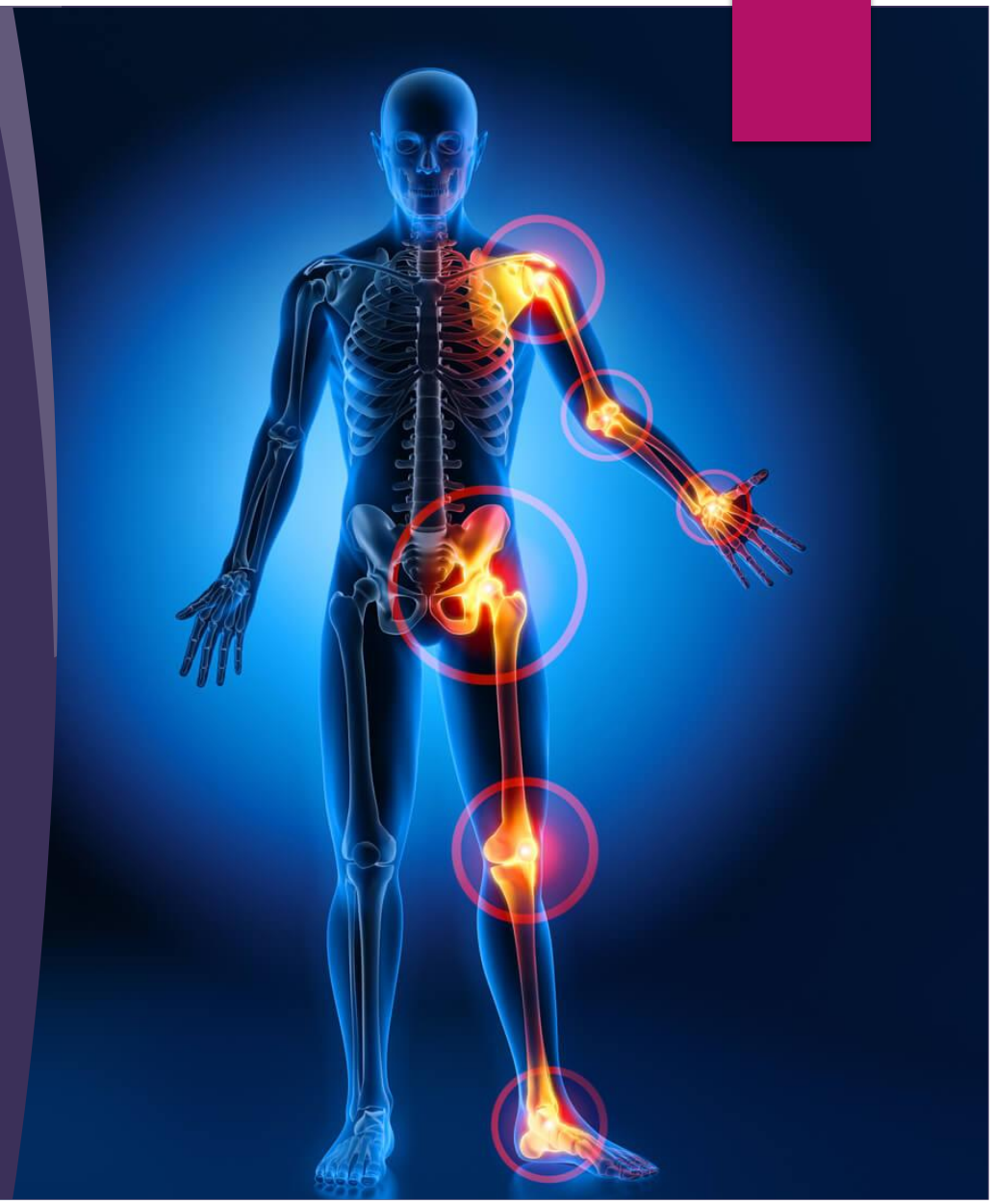
GI Symptoms

- *Dysphagia and gastrointestinal symptoms*, due to involvement of oropharyngeal striated muscles and upper esophagus, especially in DM and IBM



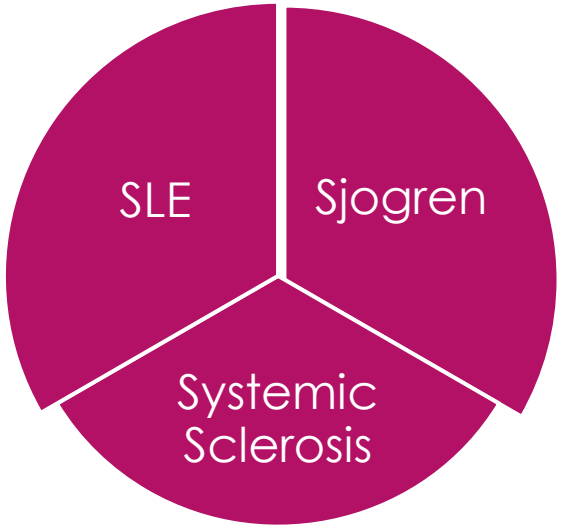
Articular Involvement

- Joint involvement is frequent and is mainly observed in the ASyS group (up to 90%) and in DM with anti MDA5+ antibody
- Arthralgia and arthritis occur usually at disease onset (83%)



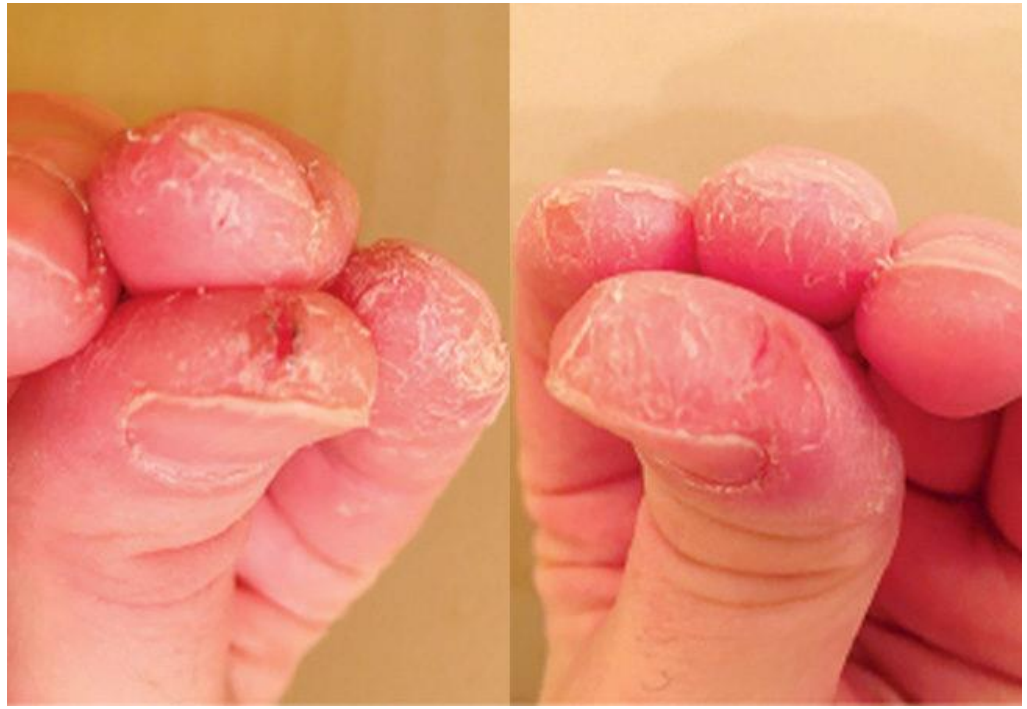
Overlap Syndrome

- Inflamatory myopathy in association with connective tissue diseases most notably systemic sclerosis, Sjogrens syndrome, and SLE



Antisynthetase Syndrome

- Is the constellation of Myositis, raynaud's phenomenon, arthritis, and ILD.
- It presents with mechanic's hands.
- It is characterized by the presence of antibodies to aminoacyl transfer ribonucleic acid synthetases most commonly Anti Jo1



Necrotizing Autoimmune Myopathy

- A sub acute, progressive proximal muscle weakness without a rash.
- Weakness generally develops more rapidly than PM and is markedly severe.
- There may be associated myalgias and dysphagia.
- CK is usually higher than seen with other IIM.
- NAM is thought to be immune mediated with a trigger such as drugs or associated with malignancy

Inclusion Body Myositis

- Duration of illness longer than 6 months
- Age at onset older than 30 years, more in male
- Asymmetric weakness of proximal and distal muscles of the upper and lower extremities
- In contrast to PM and DM, mild facial weakness is common
- Frequent falling
- Serum CK level less than 12 times normal


Thank
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