MYOSITIS

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"Myositis means muscle inflammation, and can be caused by infection, injury, certain medicines, exercise, chronic disease, or autoimmune disease"

Infammatory Myopathies……?

The inflammatory myopathies are a group of diseases that involve chronic muscle inflammation.

Sabacute or slowly progressive, symmetrical weakness primarily affecting the proximal limb and trunk muscles

Introduction

- Inflammatory Myopathies are sporadic disorders representing the largest group of acquired and potentially treatable causes of skeletal muscle weakness.
- Annual incidence ~ 1 in 100,000
- Women > Men , Polymyositis (PM) and Dermatomyositis (DM)
- Men > Women , Inclusion Body Myositis (IBM)

Introduction

- Age at onset for Inflammatory myopathies is different.
- PM > 18 years.
- DM both Juvenile population and Adulthood.
- IBM affects persons aged >50 years.

Classification

- There is no internationally accepted classification system for Inflammatory myopathies.
- For discussion purpose IM's can be classified as :-
  - Polymyositis.
  - Dermatomyositis and Juvenile Dermatomyositis.
  - Inclusion Body Myositis.
  - Myositis associated with Collegen Vascular disorder.
  - Myositis associated with malignancy

Polymyositis

- A persistent inflammatory muscle disease that causes weakness of the skeletal muscles, which control movement.
- Medically, polymyositis is classified as a chronic inflammatory myopathy.

Clinical Features

Polymyositis

- A rare, subacute inflammatory myopathy affecting adults and rarely children.
- Progressive, symmetric proximal muscle weakness with sparing of ocular and facial muscles.

  trunk, particularly hips, thighs, shoulders, upper arms and neck.

  As muscle weakness progresses, difficult to climb stairs, rise from a seated position, lift objects or reach overhead
- Pharyngeal and Neck Flexors, also involved causing dysphagia and Head drop.

Clinical Features

Polymyositis

- Associated with Myalgia and Muscle tenderness in some cases
- In Advanced and Acute cases Respiratory muscles may be affected.
- Severe weakness if untreated leads to muscle wasting.
- Sensations and DTR’s remain intact.

- Polymyositis is often associated with other conditions that may cause further complications of their own, or in combination with polymyositis symptoms. Associated conditions include:
- Raynaud's phenomenon.
- Other connective tissue diseases.
- Cardiovascular disease.
- Lung disease.

Clinical Features

Dermatomyositis

- Subacute inflammatory myopathy with muscle weakness similar to polymyositis with distinctive rash more often preceding the muscle weakness, that occur in children and adults.
- Systemic disorder most frequently affects the skin and muscles but may also affect the joints; the esophagus; the lungs; and, less commonly, the heart.

- The muscle weakness may appear suddenly or develop slowly over weeks or months. may have difficulty raising arms over head, rising from a sitting position, and climbing stairs.
- The rash may appear over the face, knuckles, neck, shoulders, upper chest, and back.

Dermatomyositis – skin findings

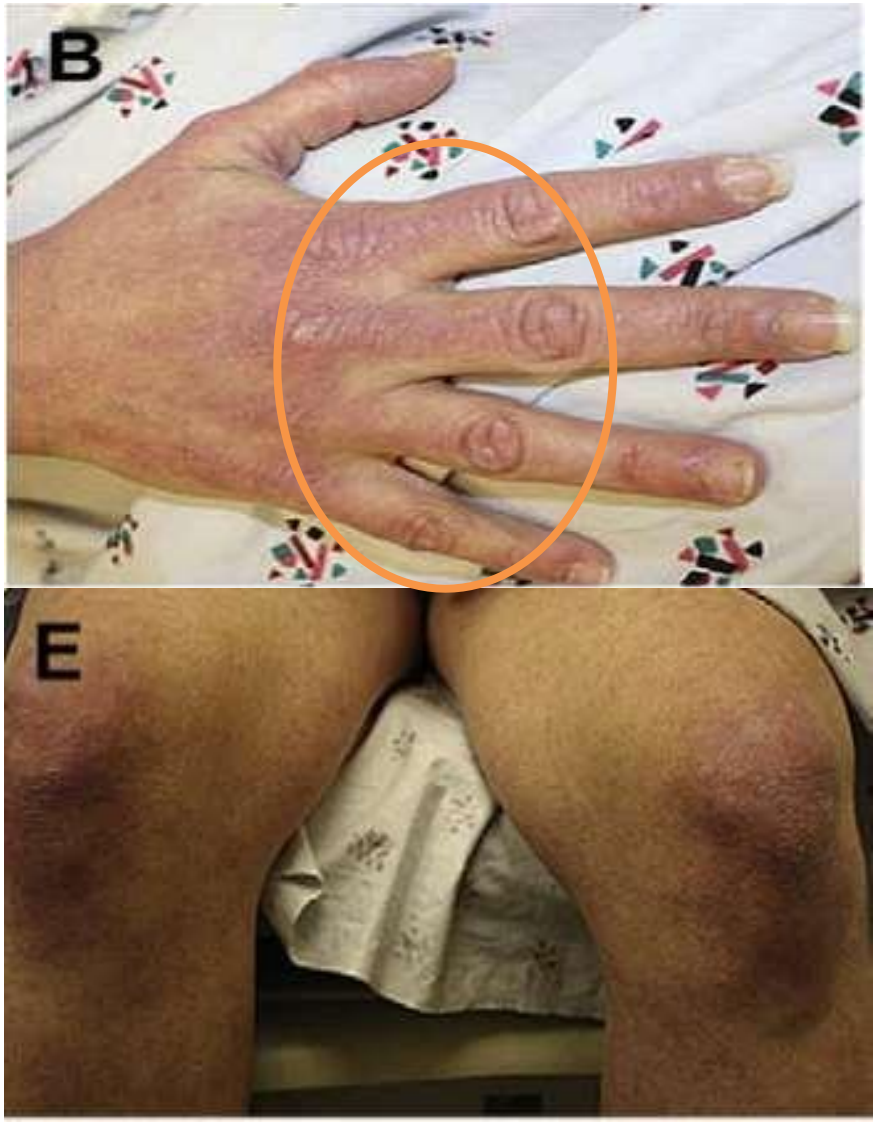
- Gottron's sign
- Heliotrope rash
- Shawl sign & V sign
- Mechanic's hands

Clinical Features

DM : Dermatologic Manifestations

Gottron Papules

- Violaceous flat topped papules and plaques over dorsal aspect of interphalengeal and MCP joints
- Pathognomic of DM, seen in > 80% patients with DM

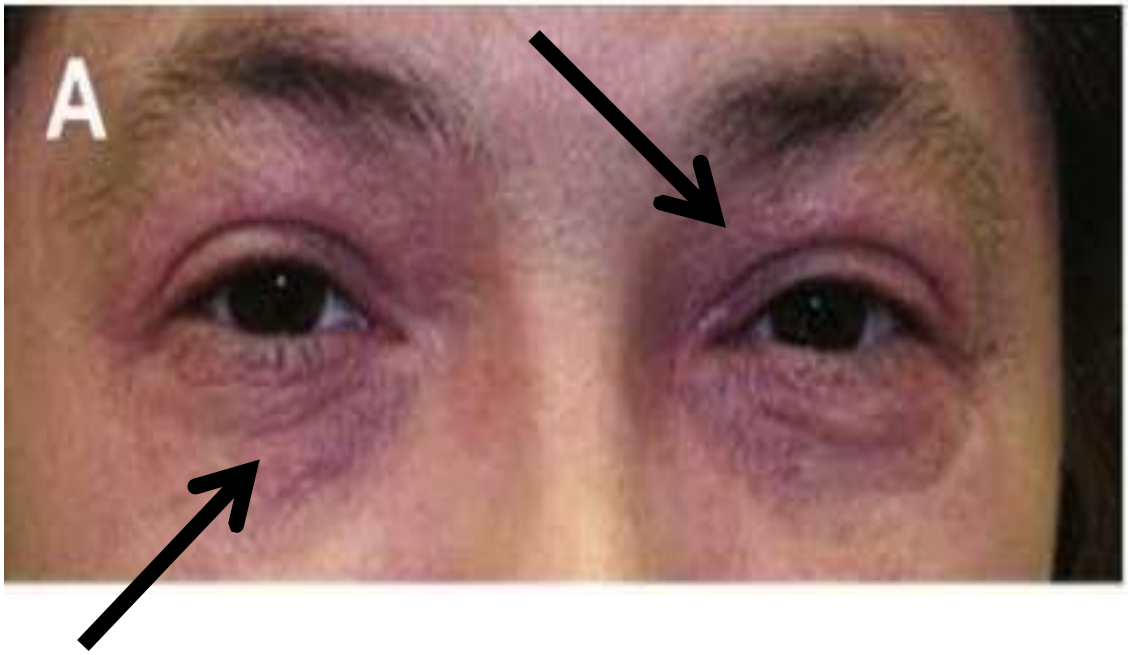


Clinical Features

DM : Dermatologic Manifestations

Heliotrope rash

- Periorbital violaceous erythema with or without edema of eyelids and periorbital tissue.
- Highly characteristic of DM.

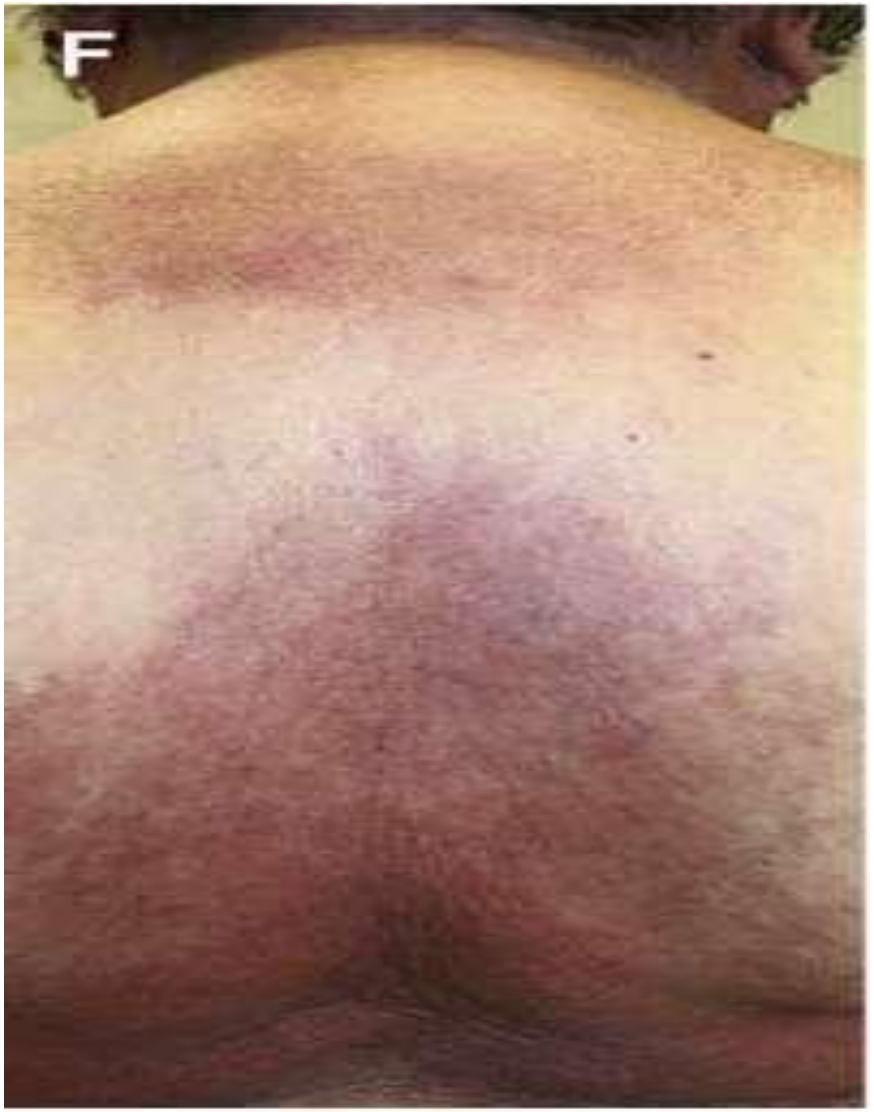


Clinical Features

DM : Dermatologic Manifestations

Shawl Sign

- Macular violaceous erythema over Nape,back and shoulders.
- May show photosensitivity.

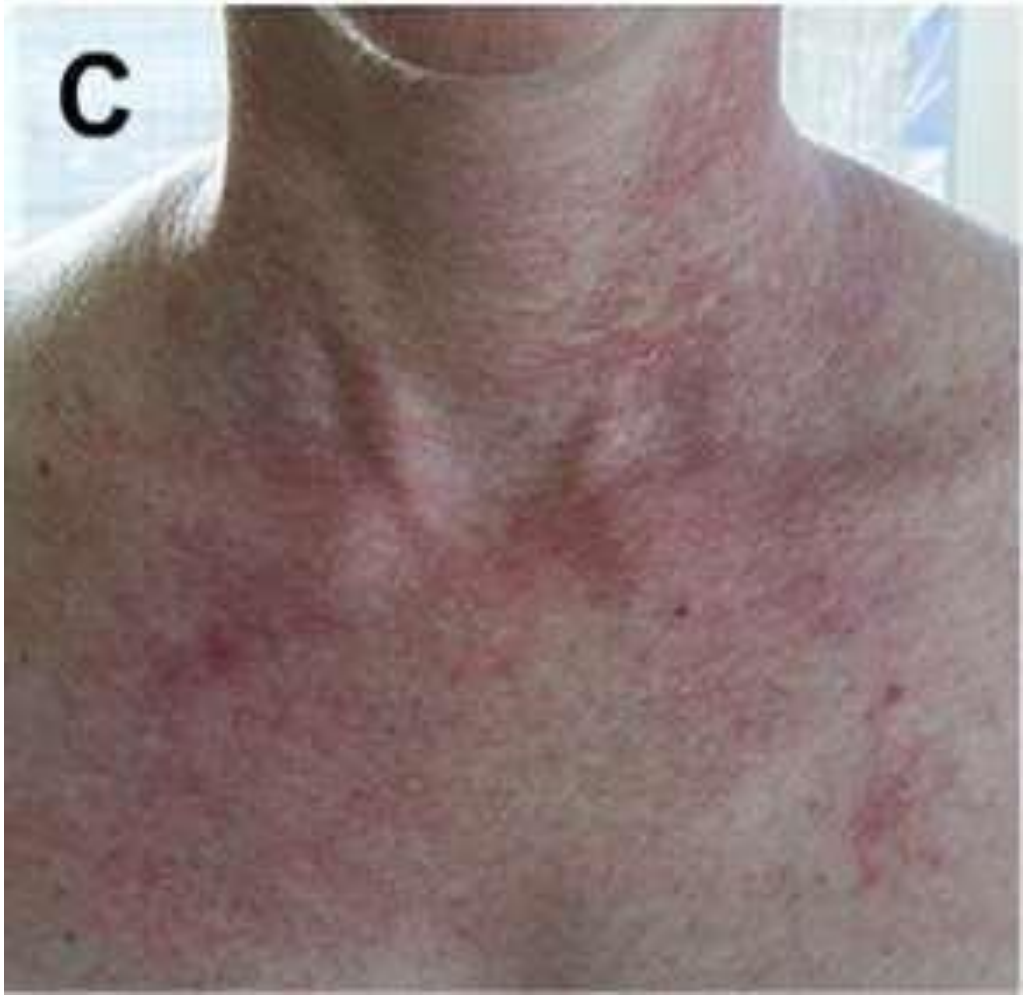


Clinical Features

DM : Dermatologic Manifestations

V- sign

- Macular violaceous erythema over V-shaped region of the neck and upper chest.
- Sometimes rash is pruritic.

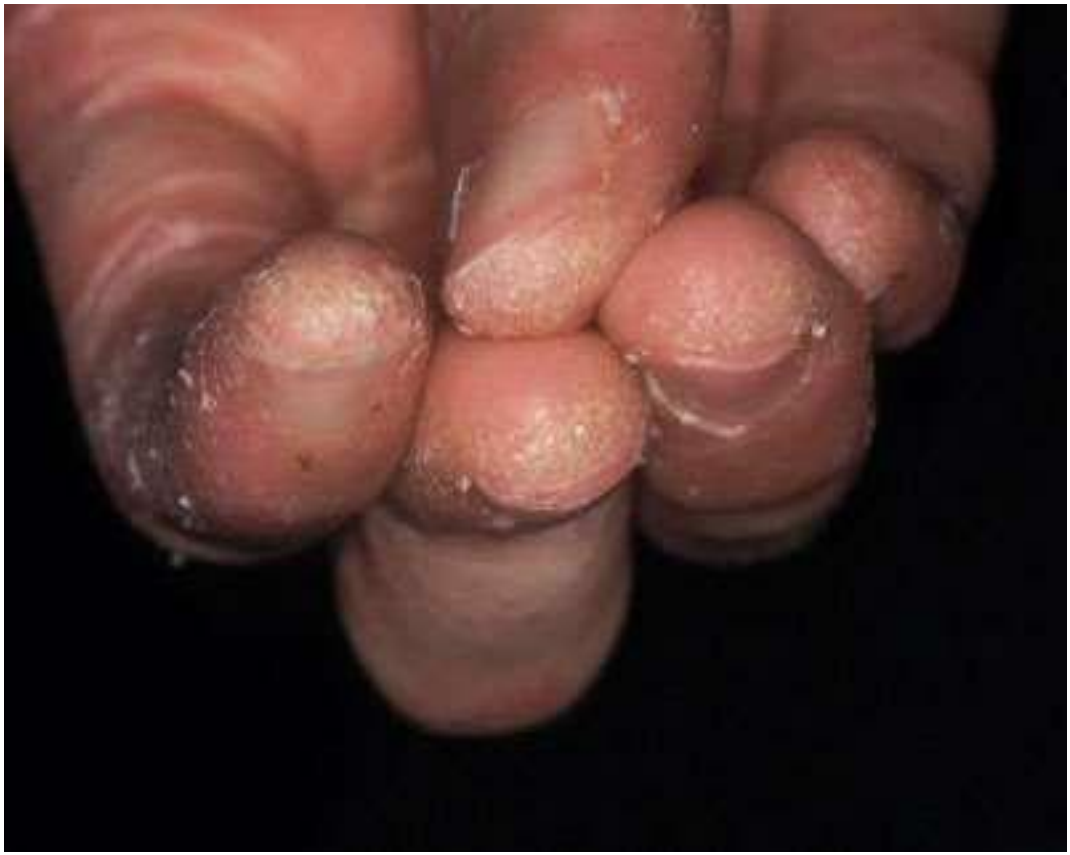


Clinical Features

DM : Dermatologic Manifestations

Mechanic's Hand

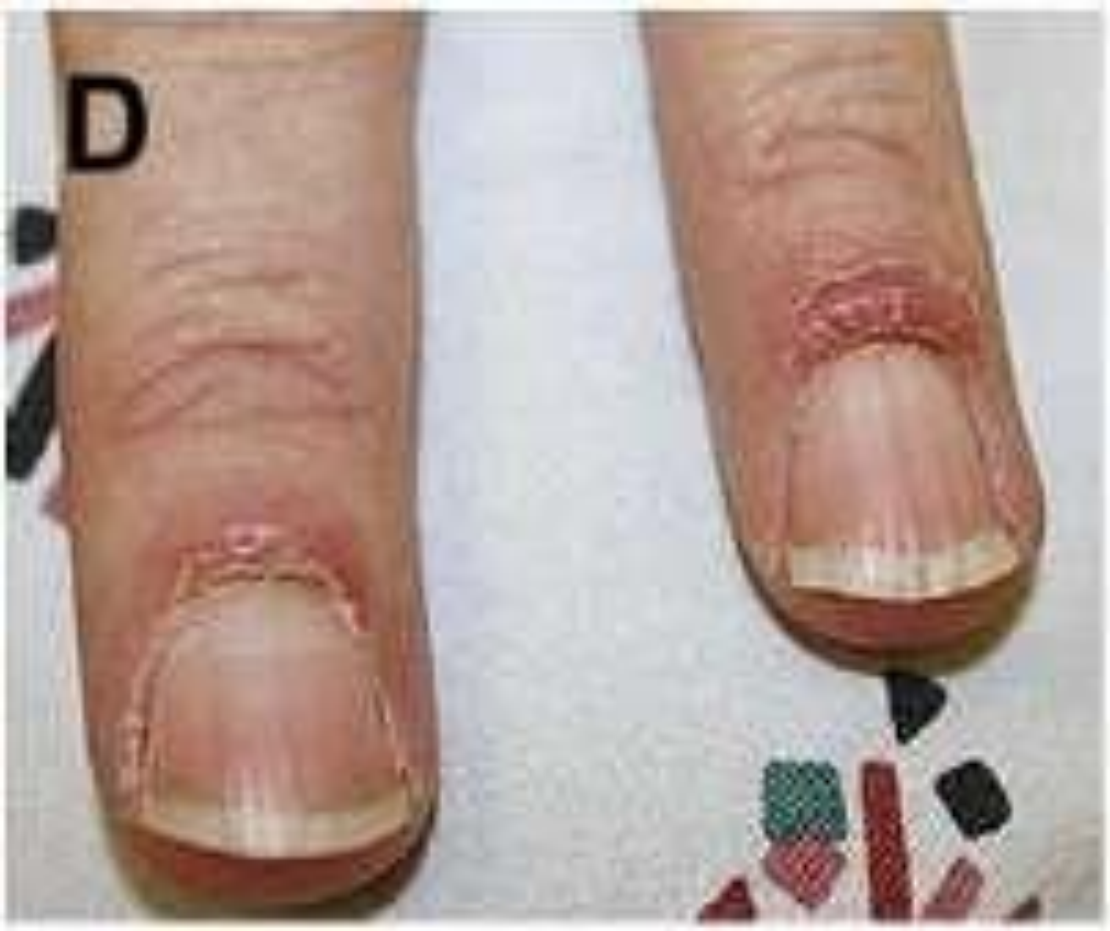
- Also considered characteristic.
- hyperkeratosis, scaling, and horizontal fissuring of the palms and fingers bilaterally.
- Can be a manifestation of the antisynthetase syndrome.



Clinical Features
DM : Dermatologic Manifestations

Nailfold Talengectasia

- Occur in 30 to 60 % early in disease.



Inclusion Body Myositis

- Symmetric or asymmetric weakness.
- Insidious onset.
- After 50 years, Male to female ratio of 3:1.
- Proximal and/or distal muscle involvement.
- Classically forearm flexors, finger flexors and Quadriceps.
- Facial and Pharyngeal muscles are more commonly involved than PM/DM.
- presence of typical inclusion bodies on muscle biopsy

- **Symptoms of Inclusion body myositis**
- Muscle weakness.
- Painless
- Heart and lungs are not affected in IBM.

Diagnosis

- In 1975, Bohan and Peter used the following criteria for the diagnosis and classification of PM and DM.

The Bohan and Peter classification criteria
1. Symmetric proximal muscle weakness.
2. Elevation of skeletal muscle enzyme levels.
3. Abnormal EMG results - Polyphasic, short, small motor unit potentials; fibrillation; positive sharp waves; insertional irritability; and bizarre, high-frequency, repetitive discharges.
4. Muscle biopsy abnormalities -Degeneration/regeneration, perifascicular atrophy, necrosis, phagocytosis, fiber size variation, and mononuclear inflammatory infiltrate
5. Typical skin rash of DM

Drawbacks of Bohan and Peter criteria

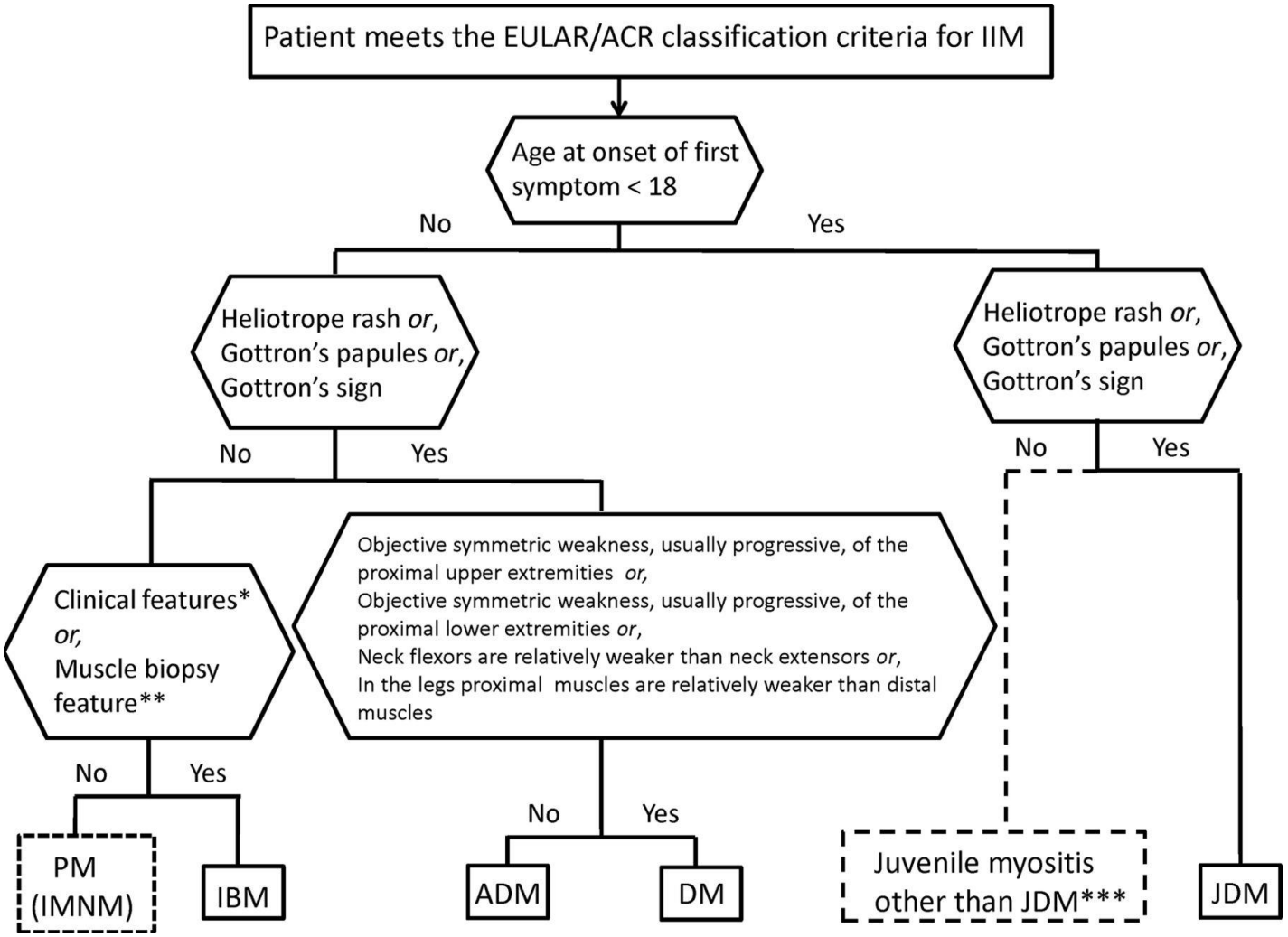
- Case series and data developed from a single institution and based on clinical observations.
- Only skin features were used to differentiate DM from PM.
- IBM was not recognized as a separate entity.

New Diagnostic Criteria

- New diagnostic criteria considers :
  1. Muscle Weakness.
  2. Creatine Kinase
  3. Electromyographic Findings.
  4. Muscle biopsy.
  5. Rash / Calcinosis.

  (EULAR/ACR) classification criteria for idiopathic inflammatory myopathies (IIMs), collectively known as myositis, and the major subgroups of IIM have been developed

Classification tree for subgroups of idiopathic inflammatory myopathies (IIMs).



Matteo Bottai et al. RMD Open 2017;3:e000507

Muscle Weakness

- In PM and DM there is progressive, symmetric proximal muscle weakness with sparing of ocular and facial muscles.
- In IBM : Symmetric or asymmetric weakness, involving the proximal and/or distal muscles classically forearm flexors, finger flexors and quadriceps.

Muscle Enzymes

- In PM, Creatine Kinase is always elevated upto 5 – 50 times the ULN.
- In DM, CK is elevated upto 50 times the ULN, in 90% of patients. While in others CK levels may be normal.
- In IBM, CK levels are elevated upto 10 times of UNL or may be Normal.

EMG in Myopathies

- In acute myopathies EMG must be conducted about 3 weeks from the onset of symptoms to ensure good sensitivity.
- EMG of the muscle is done in two situations : at rest and at voluntary activity.
- The Resting values also called spontaneous activity (not seen in normal muscles) are of three types.
  1. Fibrillations and Positive waves
  2. High frequency discharges
  3. Myotonic Discharges

- **PM/DM** **NEUROPATHIC DISORDER**
- **EMG**

  -Poly phasic action potentials -Poly-phasic action potentials

  -long duration -short duration

  -low amplitude -large amplitude

- **NCV**

  Normal Abnormal

- MUSCLE BIOPSY:
  - biopsy a clinically weak muscle, contralateral to an abnormal muscle (by EDT), MRI directed.

Perivascular and endomysial inflammation

CD8+ T cells in PM,

CD8+, CD4+ T and B cells in DM

b. b. Muscle fiber necrosis and regeneration

Diagnostic criteria for IM

	Polymyositis			
Criterion	**Definite**	**Probable**	**Dermatomyositis**	**Inclusion Body Myositis**
Myopathic muscle weakness	Yes	Yes	Yes	Yes; slow onset, early involvement of distal muscles, frequent falls
EMG findings	Myopathic	Myopathic	Myopathic	Myopathic with mixed potentials
Muscle enzymes	Elevated (up to fiftyfold)	Elevated (up to fiftyfold)	Elevated (up to fiftyfold) or normal	Elevated (up to tenfold) or normal
Muscle biopsy findings	"Primary" inflammation with the CD8/MHC-I complex and **no vacuoles**	Ubiquitous MHC-I expression but **minimal inflammation** and **no vacuoles**	**Perifascicular, perimysial,** or **perivascular** infiltrates, **perifascicular atrophy**	Primary inflammation with CD8/MHC-I complex; **vacuolated fibres** with **-amyloid deposits**; **cytochrome oxygenase–negative** fibers; signs of chronic myopathy
Rash or calcinosis	Absent	Absent	Present	Absent

Other test

- ELEVATED ESR , CRP:- 50%
- POSITIVE ANA:- 50-80%

- AUTOANTIBODIES:-
  - anti- RNP (MCTD)
  - anti-PM/Scl (OVERLAP)

Treatment

- Goals
- To eliminate inflammation.
- To restore muscle performance.
- To prevent chronic muscle disease.
- To prevent other organ system damage
- To regain quality of life.

Treatment

1. STEROIDS
2. IMMUNOSUPPRESSIVE AGENTS: methotrexate, azathioprine, etc
3. IMMUNOMODULATORY AGENTS: IVIG, Plasmapheresis
4. REHABILITATION

Prognosis

- Older studies (before the availability of steroids) revealed a 50% mortality from complications.
- Current estimates of mortality, excluding patients with malignancy, is less than 10% at 5 years after initial diagnosis.
- DM has a favorable prognosis among all, IBM has least favorable prognosis.

Poor prognostic factors

- Older age
- Malignancy
- Delayed steroid treatment
- Dysphagia with aspiration
- ILD

- Inflammatory myopathies should be considered in all patients with proximal muscle weakness.
- Diagnosis is made by Clinical findings, raised CPK, typical EMG and muscle biopsy findings.
- Treatment is by immunosuppression.
- Prognosis is generally good for DM and worst for IBM.