

FEATURES | INVESTIGATIONS | TREATMENT

Polymyositis & Dermatomyositis

Introduction

- Autoimmune inflammatory conditions
- Affect Skeletal & Smooth muscles
- **Skeletal Muscles** - Proximal skeletal muscle involvement - more common
- **Smooth Muscles** - Cardiac and Gut smooth muscles
- Characteristic skin changes in dermatomyositis

Introduction

Polymyositis & Dermatomyositis

- **Paraneoplastic** phenomenon
 - Lung, Pancreatic, Ovarian, or Bowel malignancy
 - Screen these patients
- **Isolated Disease** *or* **Associated** with other **Autoimmune** conditions

Introduction

Polymyositis & Dermatomyositis

- **Rare** conditions
- **Incidence** _ 2 – 10 cases / million / year
- **Age of Onset** _ 40 – 60 year age

Clinical Features

Clinical Features

(Muscles)

- Insidious onset, Progressive, Symmetrical, Proximal Muscle Weakness
- Lower limbs affected more than the upper limbs
- **Respiratory or Pharyngeal** muscle - Dysphagia, Dysphonia, or Respiratory weakness



**Ventilatory failure or
Aspiration**

Polymyositis
&
Dermatomyositis

Clinical Features

(Extra- Muscular)

- Fever, Fatigue & Weight Loss
- Arthralgia
- Raynaud's Phenomenon
- Interstitial Lung Disease
 - 30% Cases
 - Associated with Anti-synthetase (Anti-Jo1 Ab)
- Myocardial Involvement - Myocarditis & Arrhythmias

Clinical Features

(Skin)

- **Gottron's papules**
 - Scaly, Erythematous or Violaceous, Psoriasiform plaques
 - Extensor surfaces of metacarpophalangeal, PIP, and DIP joints
 - Pathognomonic *if present with raised CK and muscle weakness*
- **Heliotrope** rash - Lilac-purple rash on eyelids with periorbital oedema
- **Nailfold Erythema** - Due to dilated capillary loops
- **Shawl sign** - Macular rash over the upper back, chest, and shoulders

Differential Diagnosis

Differential Diagnosis

Polymyositis & Dermatomyositis

- i. Carcinomatous myopathy
- ii. Inclusion-body myositis
- iii. Muscular dystrophy
- iv. Polymyalgia rheumatica (PMR)
- v. Endocrine/metabolic myopathy (*e.g., steroids*)
- vi. Rhabdomyolysis
- vii. Certain infections (*e.g., HIV*)
- viii. Drugs - *Penicillamine, Colchicine, Statins, or Chloroquine*

Investigations

Investigations

Polymyositis & Dermatomyositis

- **Muscle Enzymes**
 - Creatinine Kinase – Raised & used for disease activity monitoring
 - Aldolase, LDH, ALT, AST
- **Electromyography** – Characteristic Fibrillation potentials
- **Muscle Biopsy**
 - Confirmatory Test
 - Histopatholgy - *Fibre necrosis, regeneration and inflammatory cell infiltrate*
- **MRI**
 - Identify abnormal areas for muscle biopsy
 - Appearance - Muscle oedema in acute myositis

Investigations

- **Autoantibodies**

- Anti-Mi2 Abs
- Anti-synthetase (Anti-Jo1 Ab) – Acute disease onset, & associated with ILD

- **Malignancy Screening**

- Complete examination
- Chest X-ray
- Serum and Urine Protein Electrophoresis
- CT of chest/abdomen/pelvis
- Prostate-specific antigen in male patients
- Mammography in Females

Treatment

Treatment

- **Corticosteroids**

- Prednisolone – 1 mg/kg daily
- Pulsed Methylprednisolone – 1 g IV for 3 days in Respiratory/Pharyngeal weakness

- **Immunosuppressives** – Used in resistant to steroid cases

- Methotrexate, Mycophenolate mofetil (MMF)
- Azathioprine & ciclosporin
- Rituximab with Steroids

Treatment

Polymyositis & Dermatomyositis

- **IV Immunoglobulins (IVIG)**
- **Skin Disease**
 - Hydroxychloroquine
 - Mepacrine (*aka* Quinacrine)
 - Topical Tacrolimus



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