▶ idiopathic inflammatory myopathies :

- Rare
- More common : Polymyositis > dermatomyositis
- Association with other immune disease
- F > M
- 4X Blacks > caucasians
- age polymyositis → 50 - 60
  - dermato → 2 peaks
    - 5 - 15 (juvenile)
    - 45 - 65
- Inclusion body myositis age → > 50

** Association of IIM
- → Autoimmune CT diseases
- → Malignancy (Dermato > Poly)
  - ovary
  - Breast
  - Melanoma
  - colon

(OBMC)

** Clincal
- → constitutional
  - fatigue
  - fever (unknown origin)
  - weight loss
- → Skin (imp. in dermato)
  - Gottron's papules (phalangeal joints)
  - V. sign (عالصدر)
  - shawl sign (عالضهر)
  - heliotrope rash (حوالين عيونه)
  - malar
  - periungal telangiectasia
  - Mechanical hands → edematous, dry
    - زي الايدي مجرحه
  - Nail fold cap. changes
    - (nail bed) (بتوسع الاوعية كثير عال)
  - Holster sign → thigh ← edema, Redness, photosensitivity

* SLE → rash w/o (nasolabial fold, forehead)

Dermatomyosites → rash with naso + forehead.

SLE → interphalangeal rash.

Dermato → gottron on joints only.

- **Soft tissue** — Calcinosis
  - Chronic
  - Childhood
  - Dermato
  - * seen in limited scleroderma
- **Muscle** — weakness
  - insidious (3-6 months)
  - symmetrical
  - proximal muscle
  - → No facial muscle weakness
  - early morning stiff. / w/o pain / ما بقدر يمشط شعره / صعوبة انه يقوم من الكرسي
  - # fibromyalgia with pain
- **joints**
  - Arthralgia → Mild
  - Arthritis
  - Rheumatoid - like (symmetric)
  - Deforming arthropathy of poly
    - → Minimal bone erosive
    - (rheumatoid a lot of erosive)
- **lung**
  - RS muscle weakness
  - interstitial lung disease + fibrosis
- **Heart**
  - Arrhythmias
  - myocarditis
  - Pericardial effusions
- **GI** — upper dysphagia (swallowing problems)
  - ↳ Aspiration → pneumonia

** Diagnosis

- ✓ muscle enzymes → CK, AST, ALT, LDH, Aldolase
- ✓ EMG
- ✓ Muscle biopsy
- ✓ Autoantibodies → Jo-1, Mi-2, SRP
- ✓ MRI

↑CK → more inflammation

↑ Jo-1 → more "

⊗ ESR/CRP → not collerate with disease activity

Myositis - specific antibodies :-

- → Anti - synthetase : Jo-1
- → SRP
- → Mi2

[1] Anti - Synthetase syndrome

Poly & Derma have abs to tRNA synthetase Jo-1.
→ Respond well to treatment

- ✓ interstical lung disease
- ✓ Mechanical hands
- ✓ Arthritis
- ✓ Raynaud
- ✓ Fever

[2] Anti - SRP → resistant to treatm.

- ✓ polymyositis
- ✓ cardiac

[3] Anti - Mi2 → good prognosis

Derma with U sign or shawl sign

Pathogenesis

** Poly:- Cell mediated

cellular immune attack (T-cells) Attack muscle fibers

=> endomysial area abundant C8 + T-cell

(around muscle fibers)

** Derma:- Humoral

B-cell & CD4+ T Helper cell infiltrate in perifascicular area.

(Atropy of fibers)

(perimysial)

** Inclusion body myositis

- Age > 50
- M > F (2-3 folds)
- Distal weakness
- Microscope:- vacuoles rimmed by basophilic material
  - ⊕ eosinophilic cytoplasm => Trichrome stain
  - ⊕ nuclear inclusions

- Radiology :-

MRI (STIR technique)

inflammation => white bright.

** Overlap syndromes

- Inflammatory myopathies ± CT diseases
- Derma + systemic sclerosis
  - sclerotic thickening of dermis
  - contractures
  - eso hypo-motility
  - microangiopathy
  - Ca^{2+} deposits

Derm + systemic S
have
↳ Anti-poly/scl ab

-Treatment => ① corticosteriods
② with azathioprine immunosuppressive agents, if
- ↳ fail to respond to high dose
- persistant disease despite treat
- inability tapper the steriod w/o reccrance
- sever steriod side effects

** 18-24 month duration of treatment

** Cancer screening >50
- CXR
- CT
- Mammogram
- Testicular exam
- fecal occult blood
- scopy

** Sjogren syndrom

- inflammatory autoimmune affect exocrine gland
↓
lymphocytic infiltrate
↓
↓ exo secretions
↓
xerophthalmia ← mucosal dryness → xerostomia
↓
glands enlargment

- Autoab
  - Anti - Ro/SS-A
  - Anti - La/SS-B

* Non erosive polyarthritis & Raynaud phenomenon
* extra - glandular
  - vasculitis
  - peri - neuropathy
  - glomerulonephritis

** ↑ risk of lymphoma

** Treatment
- sugar - free flavored lozenges
- ⊗ dry food / smoking / drugs anti cholinergic
- oral hygiene
- hydroxychloroquine for joint pain
- pilocarpine
- artificial tears