Dermatomyositis

Introduction

-Dermatomyositis is a rare autoimmune disease , characterized by inflammation of the skeletal muscles .

-Has 2 peaks (5-15 /45-65) years .

-Female-male varies between 1.5-2.4:1

- **Rash** more **often preceding the muscle weakness**.
- Other subdivisions include :

1- DM sin dermatitis (rash is transient /subtle or absent)

2- Amyopathic DM (Absent muscle weakness/ rash present) >>?? Lung disease/ malignancy

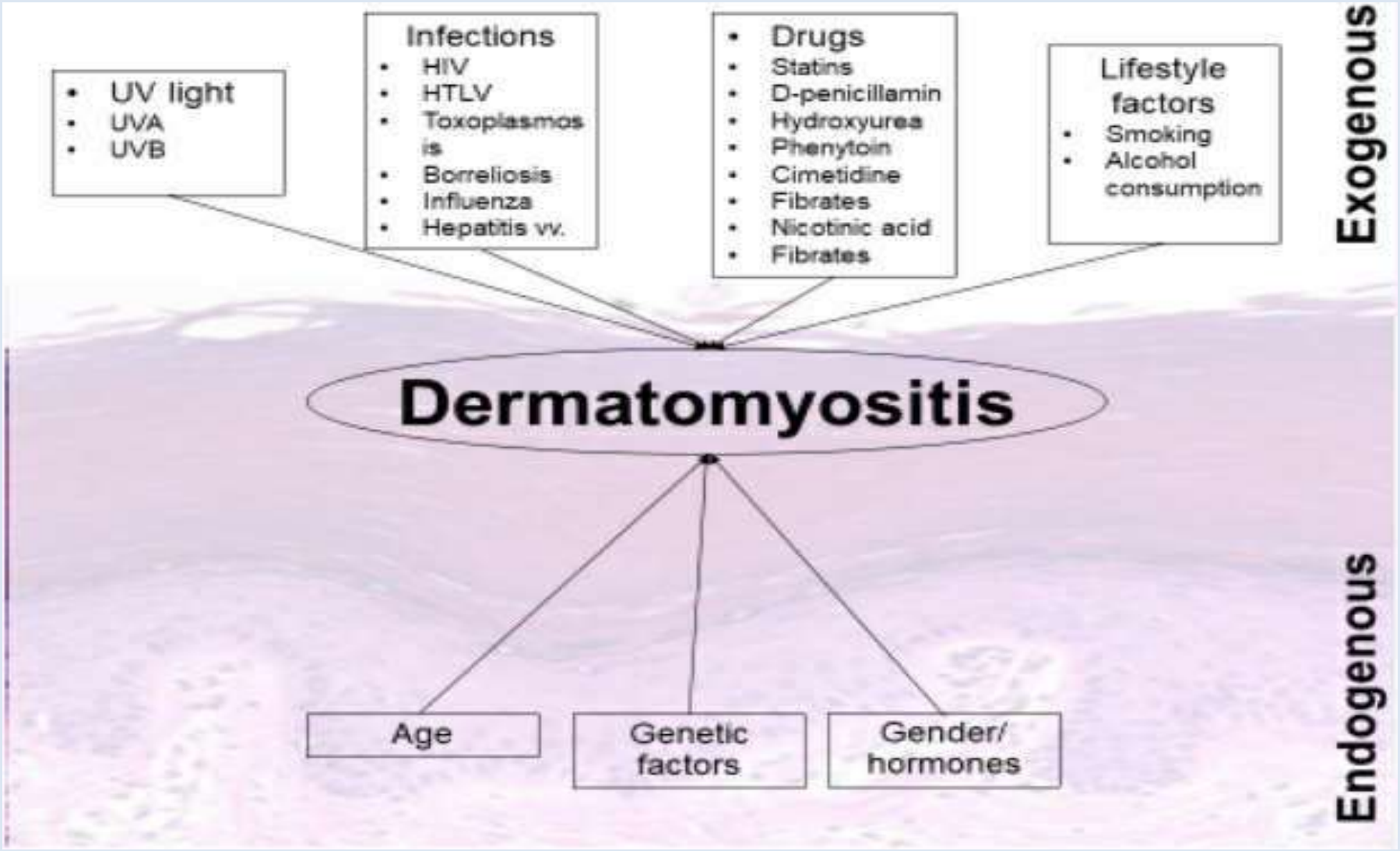
3-Hypomyopathic DM (high CK without weakness , possible disease activity on MRI or biopsy)

Diagnostic criteria:(Bohan and Peter)

Item	Description
1	Symmetrical weakness of limb-girdle muscles and anterior neck flexors
2	Muscle biopsy evidence typical of myositis
3	Elevation of serum skeletal muscle enzymes, particularly CK
4	Typical EMG features of myositis
5	Typical DM rash, including heliotrope and Gottron's papules

For the diagnosis of PM:		For the diagnosis of DM:	
Definite	All of items 1–4	Definite	Item 5 plus 3 of items 1–4
Probable	3 of items 1–4	Probable	Item 5 plus 2 of items 1–4
Possible	2 of items 1–4	Possible	Item 5 plus 1 of items 1–4

Pathogenesis:

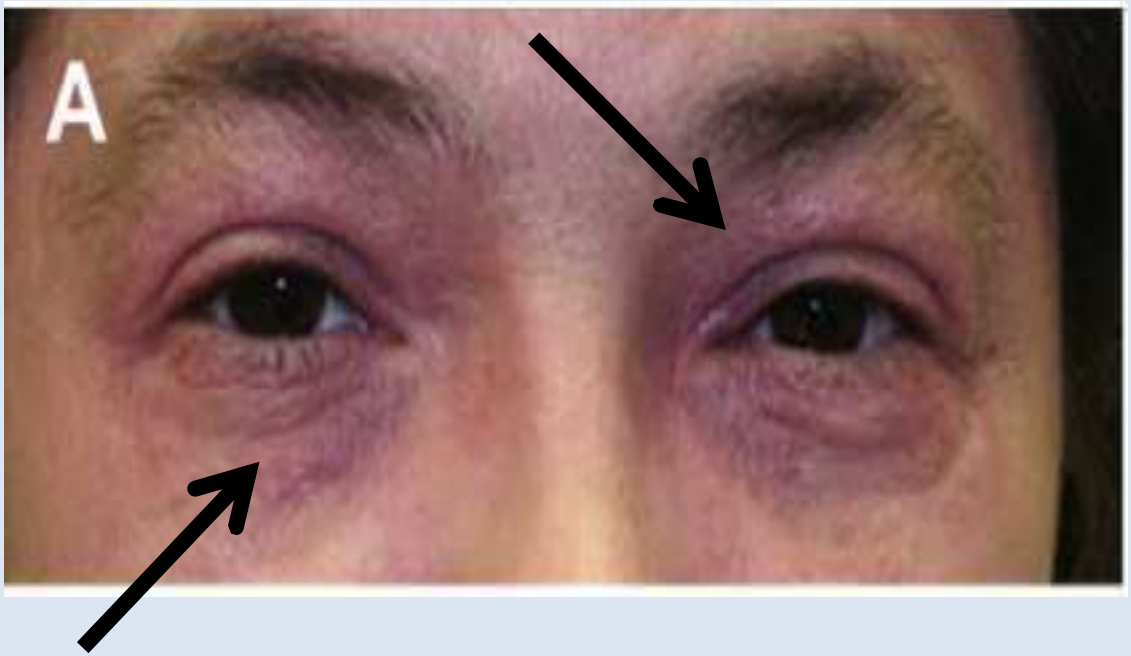


Clinical Features

1- Dermatologic Manifestations

Heliotrope rash

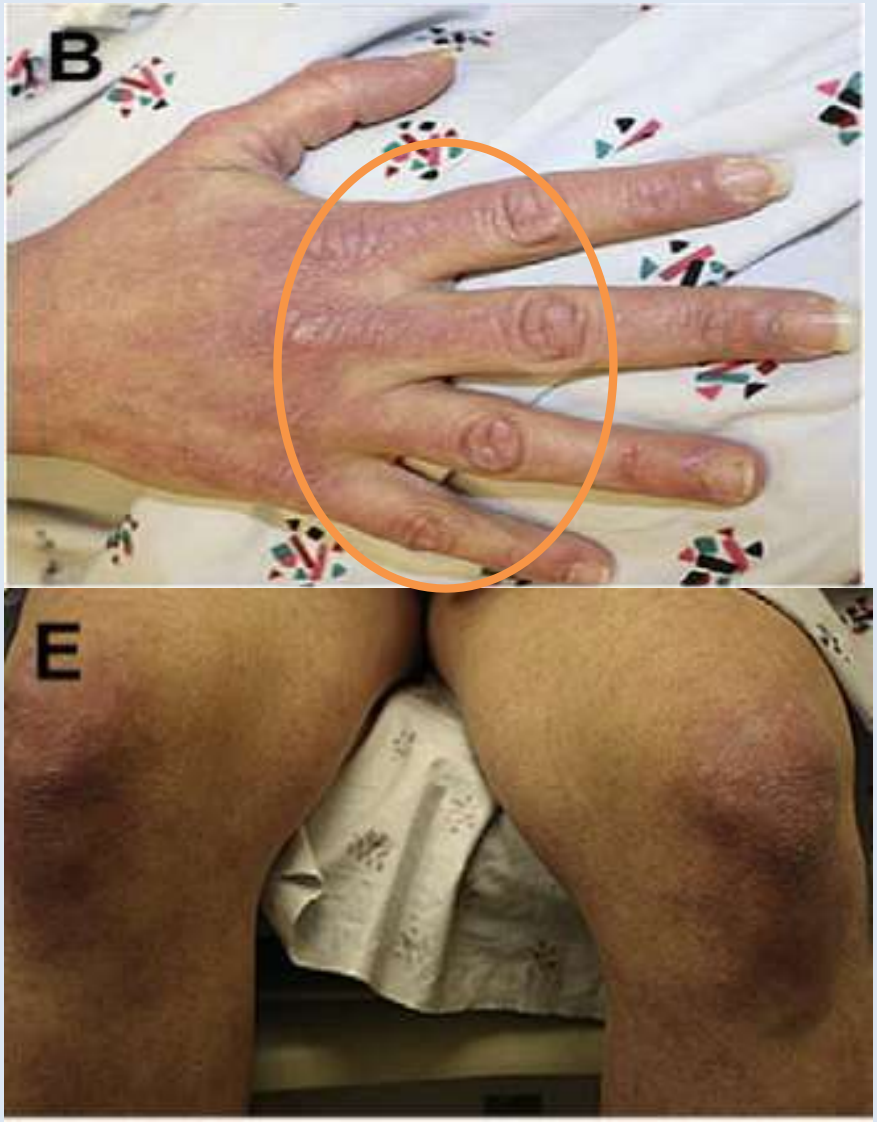
- Periorbital violaceous erythema with or without edema of eyelids and periorbital tissue.
- Highly characteristic of DM.



Clinical Features

Gottron Papules

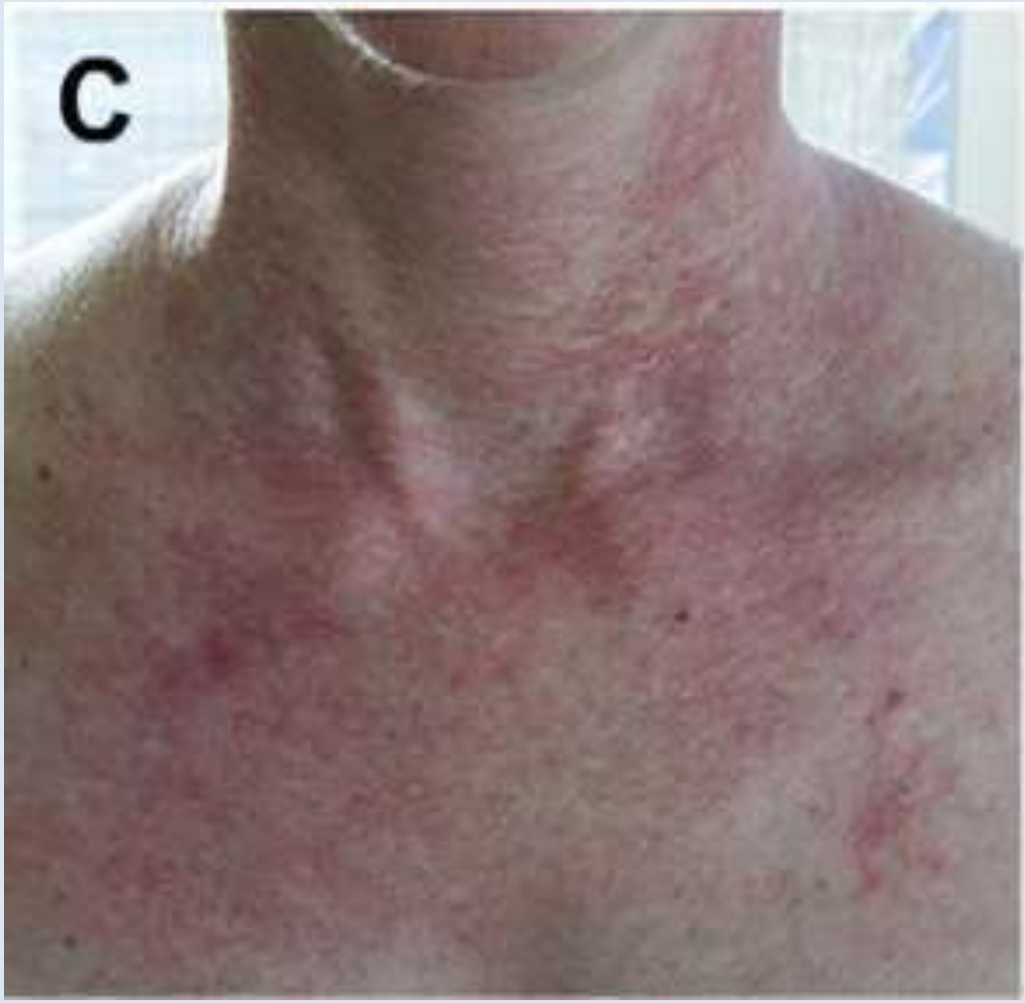
- Violaceous flat topped papules and plaques over dorsal aspect of interphalengeal and MCP joints
- Pathognomic of DM, seen in > 80% patients with DM



Clinical Features

V. <u>sign</u>

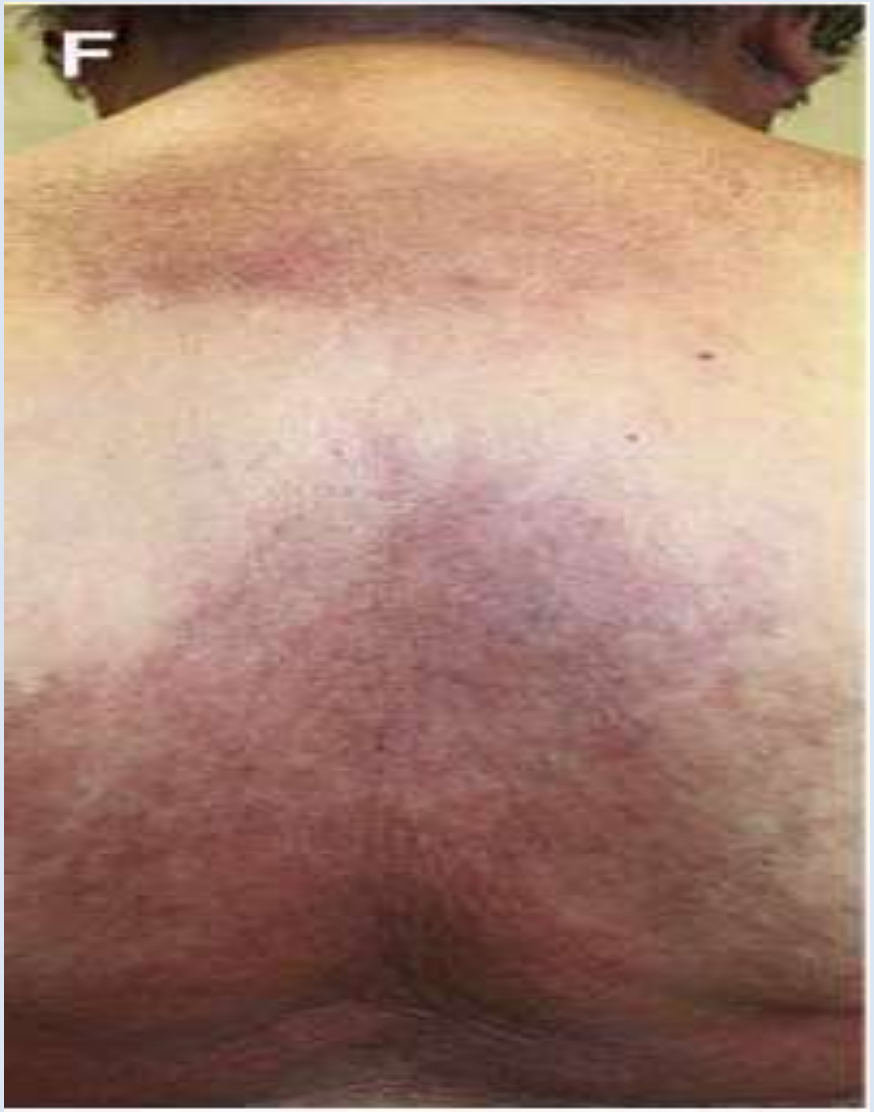
- Macular violaceous erythema over V-shaped region of the neck and upper chest.
- Sometimes rash is pruritic.



Clinical Features

Shawl Sign

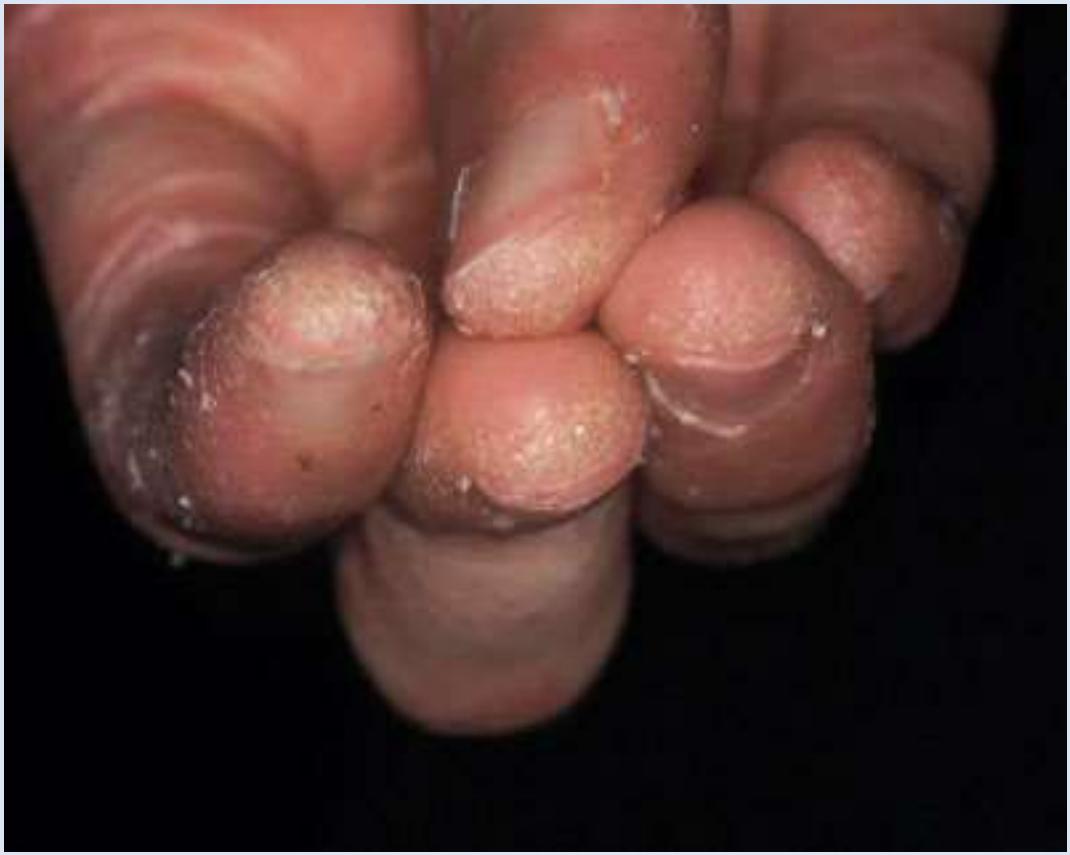
- Macular violaceous erythema over Nape,back and shoulders.
- May show photosensitivity.



Clinical Features

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

Nailfold Talengectasia

- Occur in 30 to 60 % early in disease.

2- Muscle weakness / arthritis

-Usually proximal , bilateral , symmetrical over periods of weeks or months .

-Myalgia present in 25% of cases .

- Arthitis is common in myositis /CTD overlap syndromes .

3- Respiratory involvement:

- Interstitial lung disease may present in up to 65% of cases at diagnosis .
- 70% of anti JO1 associated with ILD .
- Right sided hear failur due to ILD is common cause of death .

4- Malignancy :

-Increase risk with DM even 5 years after the myositis.

-Risk factors: old age , male, severe skin / muscle disease , high inflammatory markers , negative antibodies , low albumin

Investigations:

1- CK , LDH , AST, ALT, Ferritin , aldolase , ESR , CRP
2- Antibodies :
3- EMG : small, short ,low amplitude polyphasic motor potentials with bizzare high frequency repetitive discharges
4- Muscle biopsy:
5- Muscle MRI
6- Cancer screen
7- PFT , CT chest
8- ECG / ECHO / barium swallow

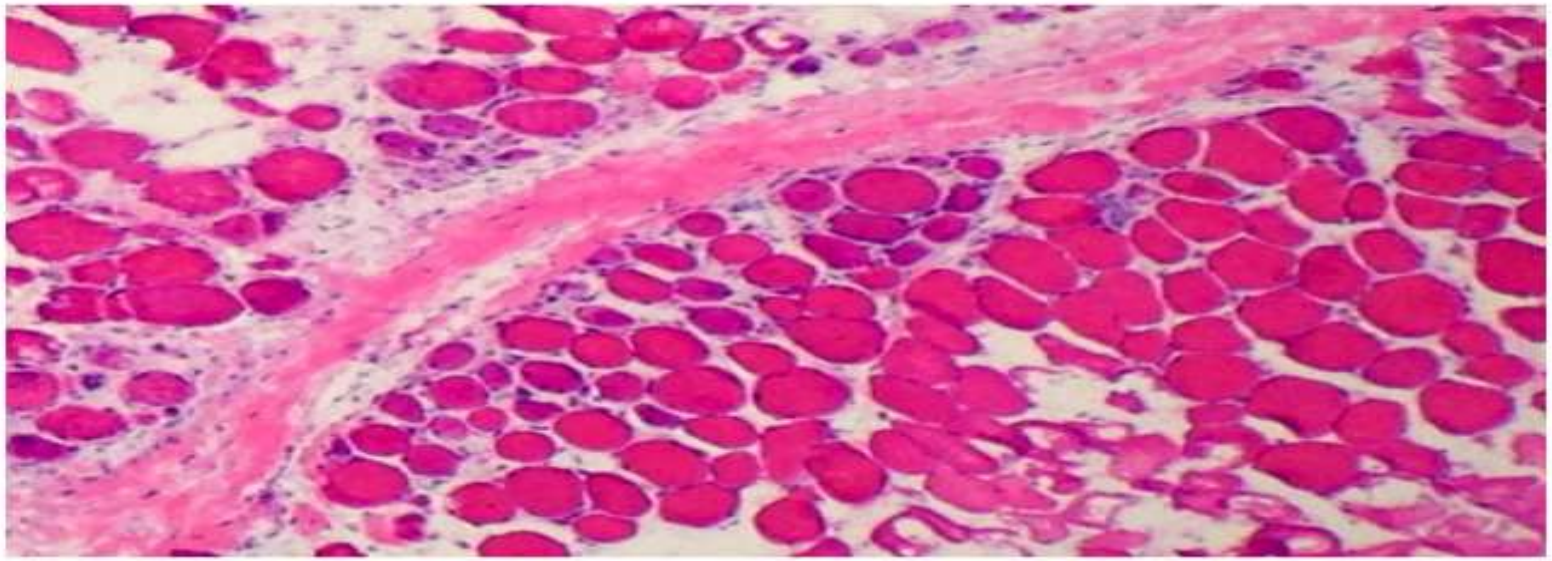
Antibodies :

- -Anti Mi2 (10-20%) , cutanuous , mild muscle disease , lower mortality rate
- -Anti -155/140 >cancer associated myositis
- -Anti NXP2> systemic features / ILD
- Anti SAE > cutanuous precedes muscle disease
- Anti MDA5 >Amyopathic / progressive ILD

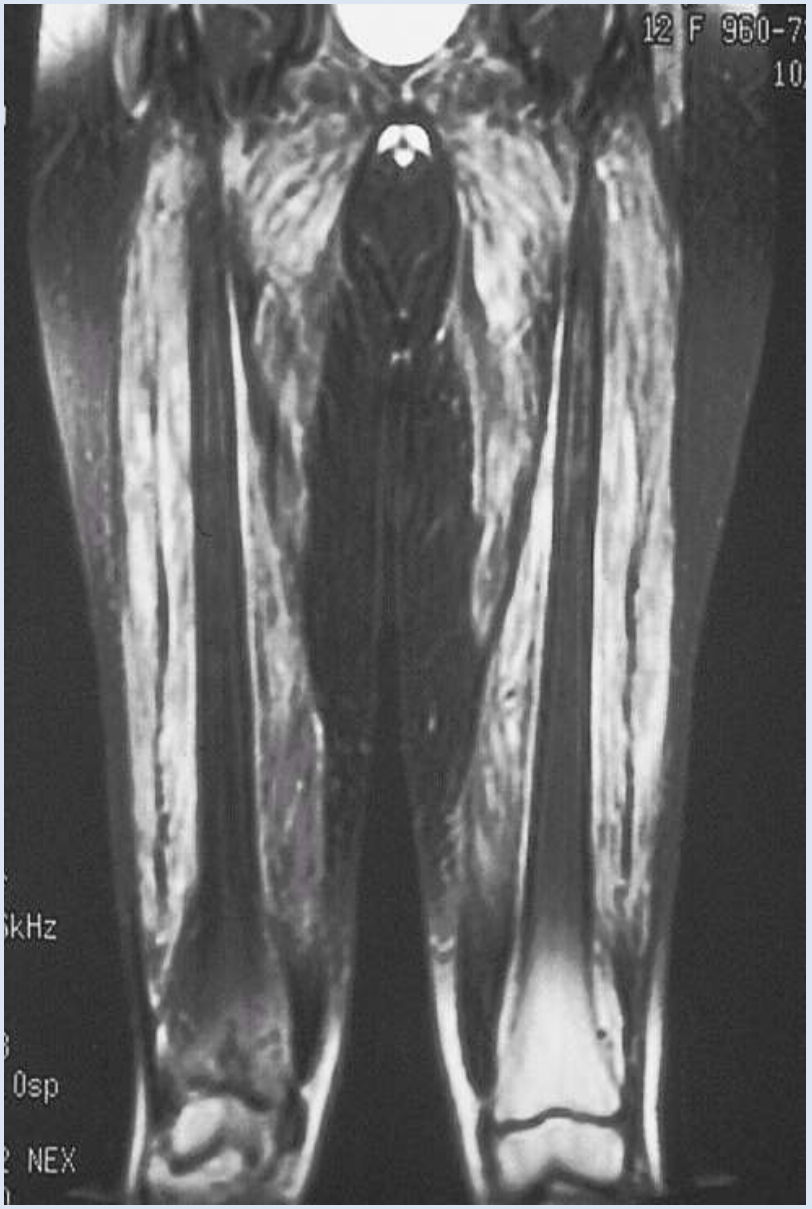
Muscle Biopsy:

-Perifascicular atrophy is the hallmark of **dermatomyositis**

-Muscle may have altered muscle fiber sizes, but there is less of a tendency to hypertrophy muscle fibers (more common in dystrophy)
May be increased internal nuclei and basophilic myofibers.
- CD4 Positive inflammatory cells



STIR imaging show fibrosis or diffuse or patchy signal symmetric increase in the proximal muscles and intramuscular fascia indicative of muscle oedema due to inflammation.



Malignancy work up :

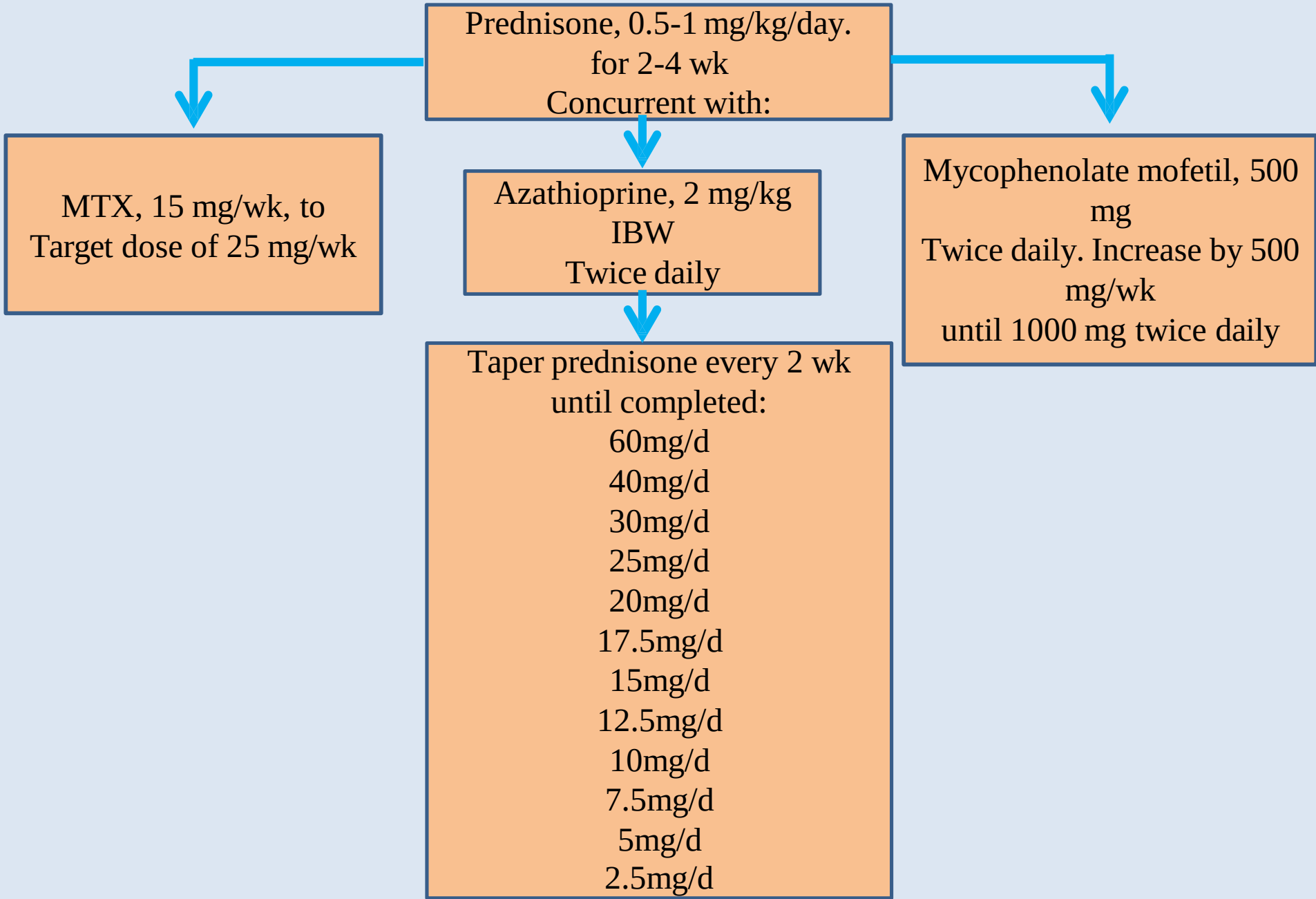
- Chest , Abdomen and Pelvic CT scans
- Mammogram, Breast and Pelvic examination in women
- Colonoscopy in patients – Age >50 years or in those with GI symptoms.

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.

Initial Treatment Approach



Severe and Refractory IM

- Myopathy refractory to conventional therapy or with severe organ-threatening manifestations like ILD, severe dysphagia, notable weight loss, severe rash, or weakness> WE may consider IVIG .

THANK YOU