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Dermatomyositis

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Introduction

Dermatomyositis (DM) is a rare inflammatory myopathy characterized by muscle weakness and distinctive skin findings. First described by Bohan and Peter in 1975, the criteria for diagnosing DM include

i. Progressive proximal symmetrical muscle weakness.
ii. Elevated muscle enzymes
iii. Abnormal EMG
iv. Abnormal finding on muscle biopsy
v. Compatible cutaneous disease

DM is associated with malignancy and can affect various organs, such as the oesophagus and lungs, with calcinosis being a complication observed in children.

The exact cause of DM is unknown, but genetic, immunologic, infectious, and environmental factors may play a role. Certain HLA types and immunologic abnormalities, including circulating autoantibodies, are common in DM. Infections and drugs, such as hydroxyurea and statins, have been implicated as triggers. DM can occur at any age but is twice as common in women.

Clinical presentation of DM often involves skin manifestations, including heliotrope rash and Gottron papules, as well as muscle weakness. Other systemic features can include joint swelling, Raynaud's phenomenon, and pulmonary involvement.

Diagnosis of DM involves laboratory tests, serological tests, and imaging studies, such as MRI and CT scans. Treatment typically includes steroids, immunosuppressive agents like methotrexate and azathioprine, and other supportive measures.

The prognosis of DM varies, with some cases spontaneously remitting and others requiring long-term treatment. Those with associated malignancy, cardiac or pulmonary involvement, or elderly patients have a poorer prognosis.

Pathophysiology:

Bohan and Peter (1975) suggested 5 subtypes of myositis

- Dermatomyositis
- Polymyositis
- Myositis with malignancy
- Childhood dermatomyositis/polymyositis
- Myositis overlapping with another collagen vascular disorder.

There is another subset of patients with a disease that only affects the skin (Amyopathic dermatomyositis (ADM) or dermatomyositis – sine myositis.

Etiology

Unknown – however genetic, immunologic, infections and environmental factors have been implicated. A link to certain HLA types (Examples) DR3, DR5, and DR7 may exist. Immunologic abnormalities are common - have circulating autoantibodies.

ANA and antibodies to cytoplasmic antigens (ie, anti-transfer RNA synthetases)

Infectious agents have been suggested as triggers

i. Viruses – Coxackie virus, parvovirus, HTLV – 1, HIV
ii. Toxoplasma species
iii. Borrella species

Drug-induced DM have been reported

1. Hydroxyurea
2. Statins
3. Penicillamine
4. TNF inhibitors
5. Quinidine
6. Interferon

Epidermology - 9.63 per million – can occur at any age, twice as common in women.

Clinical presentation:

Skin disease is one of the initial manifestations in 40% of muscle diseases may occur concurrently, may precede skin disease or may follow the skin disease.

Clinical findings:

1. Heliotrope rash over periorbital skin.
2. Gottron papules over bony prominence metacarpophalangeal joints (MCP), the proximal interphalangeal joints (PIP), and the distal interphalangeal joints (DIP)
3. Also over Elbow / Knees
4. Calcinosis in juvenile dermatomyositis
5. Others- not pathognomonic features.
   a. Malar Erythema
   b. Poikiloderma
   c. Violaceous erythema on the extensor surface
   d. Periungal and cuticular changes
   e. Eruption on photo-exposed surfaces
   f. Pruritus
   g. Scaly Scalp with Alopecia
   h. Small joint swelling in a few patients of DM.
6. Muscle disease – Proximal symmetrical muscle weakness.

Mild to moderate to severe. Sometimes Quadriparesis.

Other systemic features:

Joint swelling, Raynaud's phenomenon.

Patients with pulmonary disease crackles from interstitial fibrosis.

Patients with associated malignancy may have relevant findings.

Lab studies

Abnormal muscle enzymes – Creatine kinase also AST LDH may be abnormal.

Serological Tests

76 in > 30% cases

Positive ANA myositis-specific antibodies (MSA's)

Anti-M1 2 antibodies

Anti – J0 1, anti MDA5, Anti TIF1Y, anti – NXP2 – Anti SAE.

Other – anti-SRP, anti-PM Scl and anti-Ku

Imaging MRI
USG
EMG
Barium Swallow
CT chest

Other Muscle Biopsy
PFT
Cancer antigen Ca 125, Ca-19-9
ECG

Treatment:

Steroids

Steroid-sparing immunosuppressive or cytotoxic agent

a. Methotrexate
b. Azathioprine
c. Cyclophosphamide
d. Cyclosporine
e. Mycophenolate mofetil
f. Leflunomide
g. Tacrolimus

Other – Sun protective measure

Hydroxychloroquine

Prognosis: Disease may spontaneously remit in 20% of cases 5% fulminant course. May require long-term treatment. Those with associated malignancy, cardiac or pulmonary involvement or elderly have a poorer prognosis.

Figure 1: *Heliotrope rash in a woman with dermatomyositis*

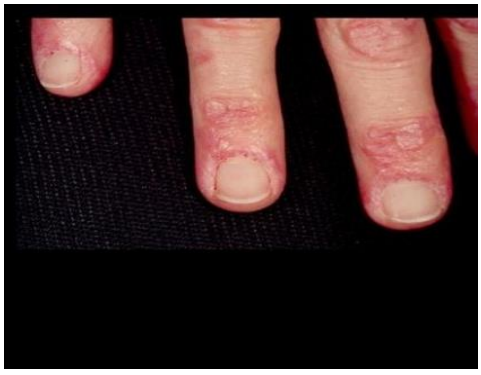

Figure 2: *Gottron papules and nail fold telangiectasia are present in this patient with dermatomyositis.*

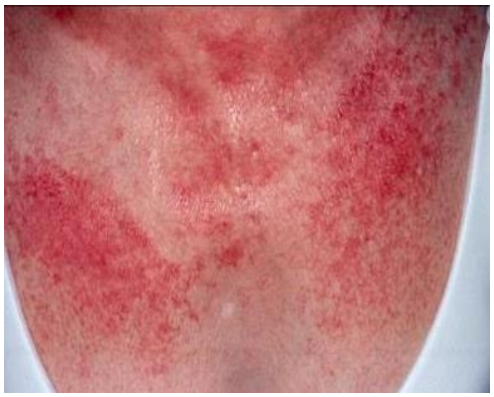

Figure 3: *Diffuse alopecia with scaly scalp dermatosis is common in patients with dermatomyositis*

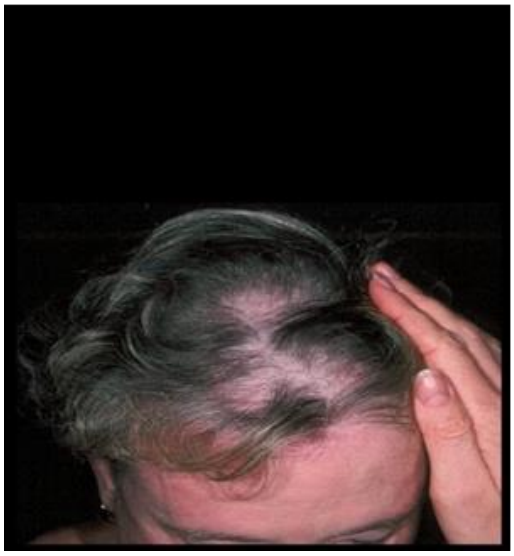

Figure 4: *Dermatomyositis is often associated with a poikiloderma in a photodistribution*

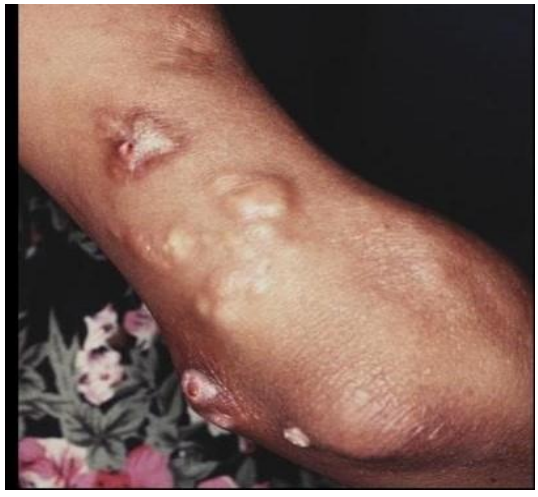

Figure 5: *Calcinosis caused by dermatomyositis in childhood can be observed in patients who had active dermatomyositis 15 years before the time of this photograph.*

Case Report

A 20-year-old female, married 6 months back reported to GKNM Hospital on 7.1.2006 with complaints of fever for 2-3 months. Fever moderate grade, on and off and increased since 1 ½ months.

Apparently well 3 months ago, noted to have significant hair loss and pigmentation over the hand Consulted a local Dermatologist. Thereafter developed fever, and pigmentation of their face, limbs, and chest. Complained of weakness, progressively worsened bed-bound difficulty in getting up, and standing without help.

At admissions conscious pallor + Mild icterus, significant hair loss (Alopecia) HR – 120/min. BP – 100/70mmHg, hyperpigmentation over face, upper limb, and neck.

CVS – Tachycardia.

Chest Basal Crepitations.

CNS – HMF speech, cranial nerve normal. Proximal muscle weakness in all 4 limbs 2-3/5 DTJ sluggish. Planter ↓ ↓ sensory retained.

Provision Diagnosis: Dermatomyositis

Investigations:

CBC - HB 5.8g%, TC 11000, N 83, L 33, platelet 203000, ESR 60mm.

RBS, RFT & Electrolytes are normal. CPK 4005 U/L, ANA positive. SGPT 151 U/L, SGOT 107 U/L other LFT normal. HIV, HBsAg – Non-reactive. Chest X-Ray Haziness lower zone. Ultrasound abdomen – Hepatomegaly. Skin Biopsy – Nil definitive.

Treated as a case of inflammatory myopathy (Dermatomyositis) with? Overlap features with IV pulse steroids, hydroxychloroquine, Azathioprine and other supportives. Improved significantly. Afebrile. Rash reduced. Could walk with help.

Review after a week showed significant improvement. Afebrile, ambulant independently. CPK 355 U/L, TSH 10.410. During follow up maintained improvement and continued on Omnacortil 10mg, hydroxychloroquine 200mg, Azathioprine 50mg BD & thyroxin 50mcg.

During review remained stable. No muscle weakness or skin lesions subsided.

Had intercurrent illnesses, required admission. Improved.

Developed fever, cough, weight loss – chest x-ray suggestive of pulmonary TB. Treated with ATT 6 months improved.

Desired to have children hence Azathioprine was stopped, continued on Omnacortil, hydroxychloroquine, thyroxine and supplements.

Uneventful pregnancies (2) normal delivery. Underwent sterilization after that. Restarted Azathioprine 50mg OD along with others. Remained well.

In 2019 in view of a stable picture suggestive of remission and normal markers. (CPK, SGOT & SGPT) decided to withdraw Azathioprine and continued on Omnacortil 5mg, hydroxychloroquine 200mg, Thyroxine 50mcg.

In 2021 noted to have proximal muscle weakness- CPK 468, SGOT 1218, SGPT 371 hence Azathioprine restarted. Improved thereafter and remained well. On Omnacortil 5mg hydroxychloroquine 200mg and supportives. Last review on 2022 the patient remained well.

Conclusion:

Dermatomyositis is a complex inflammatory myopathy with characteristic skin

findings and systemic manifestations. While its exact cause remains unknown, advancements in understanding its pathophysiology, diagnosis, and treatment have improved patient outcomes. Early recognition of symptoms, prompt diagnosis, and appropriate management are crucial in improving the quality of life for individuals affected by this rare disease. Further research is needed to elucidate the underlying mechanisms of DM and to develop more effective treatments.

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