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Review Article

Dermatomyositis

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Abstract Dermatomyositis (DM) is an idiopathic inflammatory disease affecting primarily the muscles. It is accompanied by many cutaneous changes some of which are pathognomonic for DM. Clinically it has a wide range of manifestation. In certain patients it is associated with some internal malignancy. The present review focuses on the diverse cutaneous signs seen in DM.

Key words
Dermatomyositis, Gottron papules, helitrope erythema

Definition

Dermatomyositis (DM) is a disorder mainly of skin, muscle and blood vessels in which characteristic erythematous and edematous changes in skin are associated with muscle weakness and inflammation.[1]

Epidemiology

There has been an increase in the incidence of DM worldwide. Estimated incidence in the US is estimated to be 5.5 per million. In children under 16 years of age, it is 1.9 per million. DM can affect any age group. Two peak ages of onset exist. The peak age of onset in adults is 50 years and in children it is 5-10 years. Median age of onset is 6.8 years. Males are affected twice as often as females. Mean age of onset is later in men than in women.[1,2,3]

Etiology

The cause of dermatomyositis is unknown. However, a few factors have been implicated in the etiology. There seems to be a genetic predisposition linked to certain HLA types e.g. DR3, DR5, DR7. Abnormal T-cell activity may be involved in pathogenesis of both skin and muscle disease. Autoantibodies to nuclear and cytoplasmic antigens may be present. Infectious agents such as *Toxoplasma* and *Borrelia* species and viruses such as coxsackie virus, parvovirus, echovirus, HTLV-1 (human T-cell lymphotropic virus) and HIV may have a role. Cases of drug-induced disease have been reported with hydroxyurea (in patients with CML or essential thrombocytosis), penicillamine, statin drugs, quinidine and phenylbutazone. Dermatomyositis may be initiated or exacerbated by silicone breast implants or collagen injections.[2,3]

Classification

Dermatomyositis has been classified in different ways. A practical approach is to divide it into adult-onset and juvenile-onset. Each group is further divided into subgroups.

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Adult onset

- Classic dermatomyositis alone
- Classic dermatomyositis with malignancy
- Classic dermatomyositis of an overlap connective tissue disorder
- Amyopathic dermatomyositis

Juvenile onset

- Classic dermatomyositis
- Amyopathic dermatomyositis
- Hypomyopathic dermatomyositis[7]

Clinical features

Skin disease is one of the initial manifestations. In 40% patients, skin disease may be the sole manifestation at onset. Presenting symptoms may be fever and malaise.[2,3]

Skin disease

In skin disease, there is history of an eruption on exposed surfaces and rash is often pruritic. Intense pruritus may disturb sleep pattern. There may be history of diffuse hair loss.[2,3]

Pathognomonic manifestations
These are Gottron's papules and Gottron's sign.

Characteristic manifestations
These are heliotrope rash, shawl sign/v-sign, mechanic's hand and periungual telangiectasias.

Less common manifestations
These are facial swelling, malignancy, erythroderma, cutaneous vasculitis and panniculitis.

Rare manifestations
These are follicular hyperkeratosis, papular mucinosis, hypertrichosis, malignant erythema, urticarial vasculitis, partial lipodystrophy, zebra-like striping and vulvar or scrotal involvement.

Compatible manifestations
These are poikiloderma atrophicans vasculare and calcinosis cutis.[4,5,6]

Gottron papules
These are slightly elevated violaceous, erythematous papules. Common sites are metacarpo-phalangeal joints, proximal interphalangeal joints and distal interphalangeal joints (**Figure 1**). These may be seen on elbows, knees or feet.[4,5,6] A slight scale, occasionally a thick psoriasiform scale may be present. These are seen in 70% patients of dermatomyositis. Lesions may resemble lesions of lupus erythematosus, psoriasis or lichen planus.[2,3]

Heliotrope rash
Heliotrope rash is named after the tendency of certain plants to grow towards sun. It is a violaceous to dusky erythematous rash with or without edema in a symmetrical distribution involving eyelids, periorbital skin**,** upper cheeks**,** forehead and temples (**Figure 2**). Sometimes only a mild discoloration is seen along eyelid margin. Its presence is highly suggestive of dermatomyositis. Affected areas are typically more sensitive to sun exposure. Rash occurs early in the course of disease in 30-60% patients.[2,3]

Mechanic's hands
These are seen in patients with anti-synthetase antibodies. There is hyperkeratosis, fissuring and linear

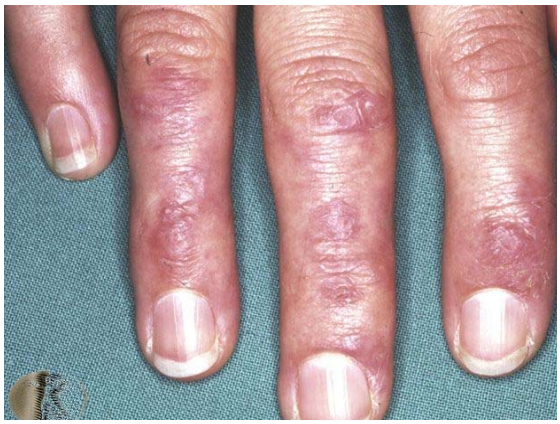

Figure 1 Gottron's papules [4].

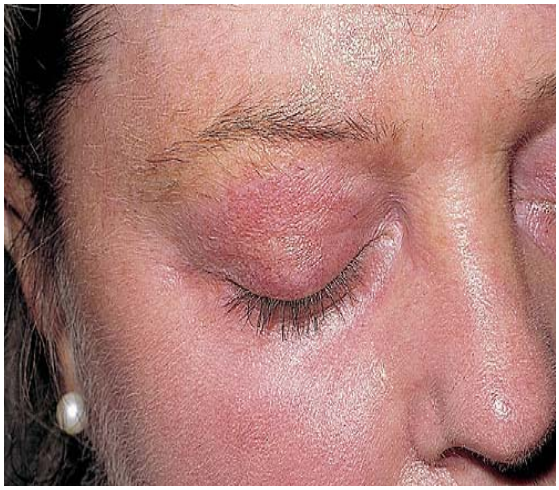

Figure 2 Heliotrope erythema with periorbital edema [4].

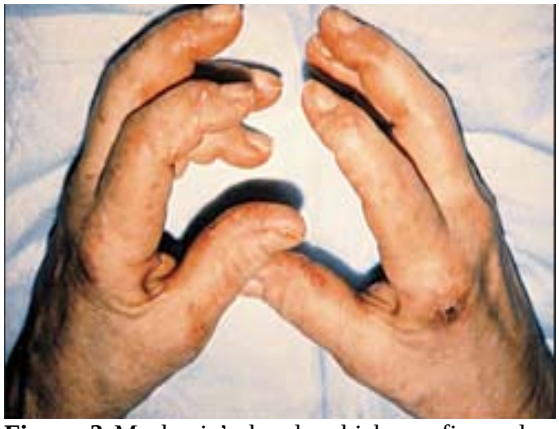

Figure 3 Mechanic's hands which are fissured, scaly, hyperkeratotic and hyperpigmented, suggestive of manual labour [4].

hyperpigmentation of radial and palmar surfaces of fingers (**Figure 3**).[1]

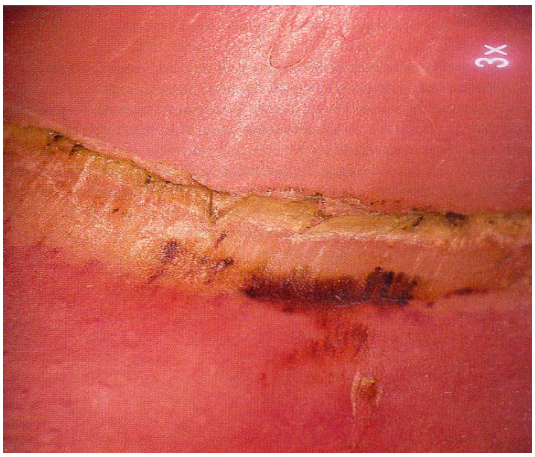

Figure 4 Dilated nailfold capillaries and hypertrophic ragged cuticle [4].

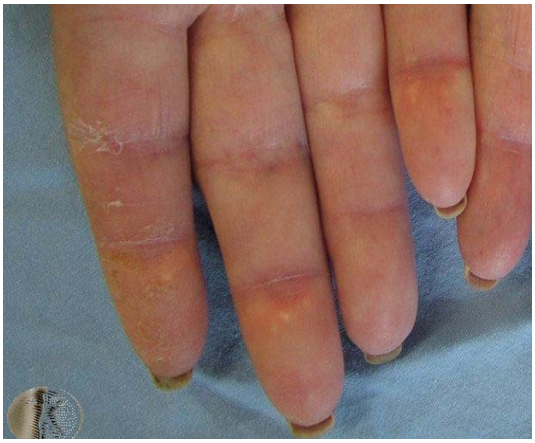

Figure 5 Calcinosis cutis on palms [4].

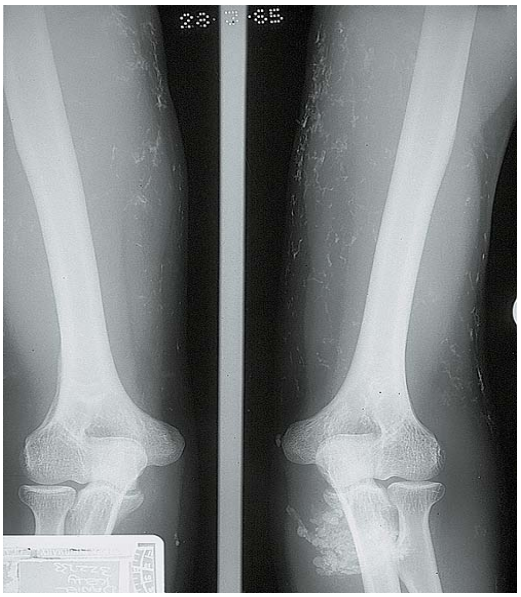

Figure 6 Radiograph showing subcutaneous calcification. This is a particular feature of childhood dermatomyositis [4].

Shawl sign

It is erythematous, poikilodermatous macules distributed in a "shawl" pattern over the shoulders, arms and upper back.[4,5,6]

V sign

It is erythematous, poikilodermatous macules distributed in a V-shaped distribution over the anterior neck and chest. [4,5,6]

Poikiloderma atrophicans vasculare (poikilodermatomyositis)

This is circumscribed violaceous erythema with associated telangiectasia, hypopigmentation and superficial atrophy. It is most commonly found over posterior shoulders, back, buttocks, V-shaped area of the anterior neck and chest and is often a late finding.[4,5,6]

Scalp involvement

Scalp involvement is relatively common. There is an erythematous to violaceous psoriasiform dermatitis. Non-scarring alopecia may occur in some patients.[2,3,10]

Nailfold changes

There are periungual telangiectasias. These are apparent clinically or are visible only on capillary microscopy (**Figure 4**). There is characteristic cuticular change with hypertrophy of cuticle and small hemorrhagic infarcts.[2,3]

Calcinosis cutis

Calcinosis cutis occurs in 30-70% cases of juvenile dermatomyositis and in 10% of adult cases. There are firm yellow or white nodules under the skin and are present on buttocks, elbows, knees or traumatized areas (**Figure 5**). It is associated with increased disease activity and duration.[4,5,6]

Muscle disease

Muscle disease may occur concurrently, may precede or may follow skin disease by weeks to years. Muscle involvement is manifested by progressive, proximal, symmetrical muscle weakness affecting shoulder and pelvic girdles mainly.

Patients complain of fatigue of muscles or weakness while climbing stairs. There is also difficulty in rising from sitting position, combing hair and reaching for items above the shoulders. Muscle tenderness is a variable finding. Testing of muscle strength is part of assessment of these patients. Extensor muscles are more affected than flexors. Distal strength is almost always maintained.[2,3]

Other systemic features

Systemic features of dermatomyositis include arthralgia, arthritis, dyspnea, dysphagia, dysphonia, arrhythmias, abdominal pain and visual changes.[4,5,6]

Joint involvement

Joint swelling occurs in some patients. Small joints of hands are most frequently involved. Arthritis associated with dermatomyositis is not erosive or deforming.[2,3]

Juvenile dermatomyositis

Its clinical presentation is usually different from presentation of adult type. Skin lesions are similar. Common findings are low-grade fever, increased risk of gastrointestinal manifestations and symmetric arthritis of large and small joints. There is an increased

incidence of calcinosis cutis in juvenile patients (**Figure 6**). Asymptomatic cardiac conduction delays and right bundle branch block is found in 50% patients. There is no association between juvenile dermatomyositis and malignancy.[11,12]

Histopathology

Skin

There is hydropic degeneration of basal cells of epidermis, edema of upper dermis, vascular dilatation and perivascular inflammatory infiltrate composed mainly of CD4+ T cells and HLA-DR-expressing macrophages. B lymphocytes are absent. Histiocytes, plasma cells and eosinophils are present. PAS-positive fibrinoid deposits are seen at dermo-epidermal junction and around dermal capillaries. There is accumulation of mucopolysaccharides in dermis (evident with use of special stains). Hyperkeratosis, acanthosis, and mild papillomatosis are seen in Gottron's papules. In later stages there is thickening of collagen of dermis, epidermis becomes atrophic, there is flattening of rete ridges and increased pigment in basal layer (**Figures 7** and **8**).[1]

Muscles

In the early stages, there is loss of transverse striation of muscle fibres, hyalinization of sarcoplasm and increase in sarcolemmal nuclei. In late stages, fibres fragment and show granular and vacuolar degeneration (**Figure 9**). Later, muscle becomes atrophied and sclerosed.[1]

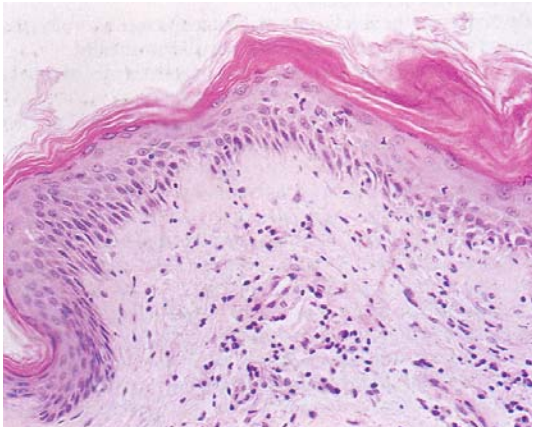

Figure 7 Focal, mild basal cell hydropic degeneration and a chronic inflammatory cell infiltrate is present in the dermis [4].

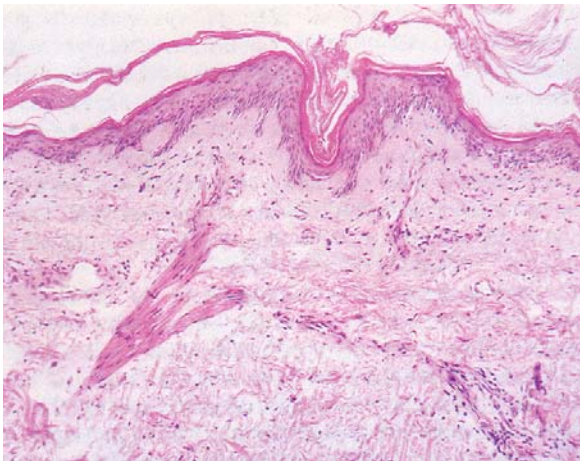

Figure 8 Hyperkeratosis and epidermal atrophy [4].

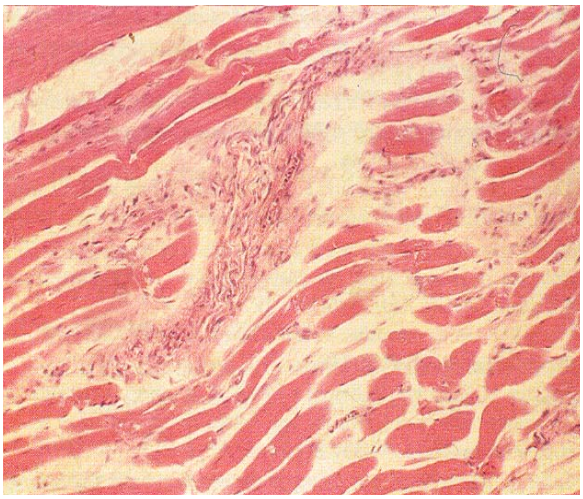

Figure 9 showing degeneration of muscle bundles with edema and inflammatory cells [4].

Relationship to malignancy

Primary tumour occurs in lungs, breast, female genital tract, stomach, rectum, kidney or testis. Dermatomyositis precedes the neoplasm in 40% cases, both may occur together in 26% cases and neoplasm may occur first in 34% cases.[1,8]

Complications of dermatomyositis

Complications of dermatomyositis include contracted limbs, aspiration pneumonia, type 2 respiratory failure, diffuse interstitial pneumonitis, pulmonary fibrosis, pericarditis, cardiac failure, cardiomyopathy, cardiac conduction defects, vasculitis, large bowel infarction (secondary to vasculopathy), muscle atrophy, muscle calcification, malnutrition, acute kidney failure, malignancy, Raynaud's phenomenon, hypertrichosis and sclerosis of skin. Ocular complications include nystagmus, cotton-wool spots, optic atrophy and conjunctival edema.[4,5,6]

Amyopathic dermatomyositis

Amyopathic patients have pathognomonic skin changes, without clinical or laboratory evidence of muscle involvement. It is reported in 2-11% patients. Lethargy, pruritus, fatigue, photosensitivity and arthralgia are the usual presenting features.[4,5,6]

Overlap syndrome

A number of patients with dermatomyositis also meet the criteria for one of the connective tissue disorders. These disorders include rheumatoid arthritis, scleroderma, systemic lupus erythematosus and polyarteritis nodosa. 11-40 % pts have a concomitant diagnosis of a connective tissue disorder. Overlap syndrome is common in females. Male to female ratio is 9:1.[4,5,6]

Table 1 Diagnostic criteria for dermatomyositis [4,5,6].

1. Skin lesions
   - i. Heliotrope rash
   - ii. Gottron's sign
   - iii. Erythema on the extensor surface of extremity joints, slight raised red-purple erythema over elbows or knees
2. Proximal muscle weakness
3. Elevated serum creatine kinase or aldolase level
4. Muscle pain on grasping or spontaneous pain
5. Myogenic changes on electromyography
6. Positive anti-Jo-1 antibody test
7. Nondestructive arthritis or arthralgia
8. Systemic inflammatory signs such as temperature more than 37°C [98.6°F] at axilla, elevated serum C-reactive protein level or ESR of more than 20 mm per hour.
9. Pathological findings compatible with inflammatory myositis.

Patients presenting with at least 1 finding from the above three and 4 findings from features 2-9 are said to have dermatomyositis.

Differential diagnosis

Differential diagnosis of dermatomyositis includes HIV infection (at onset of immunodeficiency), lichen planus, Polymorphous light eruption, seborrheic dermatitis, systemic lupus erythematosus, psoriasis, contact dermatitis, atopic dermatitis, trichinosis (caused by periorbital swelling and edema), alcohol and drugs that can cause myositis (penicillamine, NSAIDS, hydroxyurea, pravastatin, clofibrate).[1,4,5,6]

Diagnosis

The diagnostic criteria for dermatomyositis is shown in **Table 1**. Patients presenting with at least 1 out of 4 cutaneous findings

and 4 findings from extracutaneous features 2-9 are said to have dermatomyositis.

Evaluation of dermatomyositis

Evaluation of dermatomyositis is done on the basis of history and physical examination, chest radiograph, laboratory tests, urine examination especially 24 hour urinary creatine level, skin and muscle biopsy, MRI and electromyography.[4,5,6]

Laboratory manifestations of dermatomyositis

1. Muscle enzyme elevation (CPK, serum aldolase, LDH, ALT, AST, carbonic anhydrase isoenzyme II)

2. Autoantibodies

i. ANA levels elevated in 60 to 80% of patients with classic dermatomyositis
ii. Anti-Jo-1 most common antisynthetase found; 20% of patients with dermatomyositis may have positive result
iii. Anti-EJ may be more associated with typical skin lesions
iv. SRP occurring in 5% patients
v. Mi-2 antibodies (a nuclear protein complex): occurring in 15 to 20% of patients with classic dermatomyositis, associated with a more treatment-responsive form, shawl sign and prominent cuticular changes
vi. *Anti-PM-Scl antibodies* associated with overlap of scleroderma and dermatomyositis.
vii. *Anti-Ku antibodies* associated with overlap of scleroderma or SLE with dermatomyositis.

3. ESR

It is elevated in approximately 50% patients (does not correlate well with disease activity).

4. Rheumatoid factor

It is seen in 20% patients, mostly in those with overlap syndrome.

5. von Willebrand factor

It is reported to correlate with juvenile dermatomyositis.

6. EMG

There is myopathic pattern, 10% are false-negative.

7. Magnetic resonance imaging

It is useful for assessing the presence of an inflammatory myopathy in patients without weakness. It is also useful in differentiating steroid myopathy from continued inflammation and may serve as a guide in selecting a muscle biopsy site.[2,3,4,5,6]

Treatment

Treatment can be divided into general measures and medical treatment.

General measures

Bed rest is valuable for patients with severe muscle inflammation. A program of physical therapy is useful to help prevent contractures. Sun avoidance and use of a broad-spectrum sunscreen is recommended.

Table 1 Different systemic medications used in dermatomyositis [4,5,6,9].

Treatment modality	*Dosage*	*Comments*
Oral corticosteroids	0.5 to 1.5 mg/kg daily until serum CK normalizes, then slowly taper over 12 months	Consider adjunctive therapy if no improvement in objective muscle strength after three months of therapy
Methotrexate	Oral: 7.5 to 10 mg/week, increased by 2.5 mg/week to total of 25 mg/week Intravenous: 10 mg/week, increased by 2.5 mg/week to total of 0.5 to 0.8 mg/kg	First-line adjuvant therapy in patients unresponsive to steroids
Azathioprine	2 - 3 mg/kg/day tapered to 1 mg/kg/day once steroid is tapered to 15 mg/day	Screen patients for thiopurine methyltransferase deficiency before therapy
Cyclophosphamide	Oral: 1 -3 mg/kg/day Intravenous: 2 -4 mg/kg/day, in conjunction with prednisone	In refractory cases only
Hydroxychloroquine	200 mg twice daily in adults; 2-5 mg/kg/day in children	
Intravenous immunoglobulin	2 g per kg in divided doses once per month for 3 months	

Diet

Diet should be well-balanced. Patients with severe muscle inflammation may need extra protein to balance their loss. Patients of dysphagia may require a special diet depending upon the severity of esophageal dysfunction.[13]

Medical treatment

Table 2 shows different drugs used in the management of dermatomyositis. Antihistamines are given for control of severe pruritus and pain relievers to treat associated pain.

Dermatomyositis treatment options still under investigation

Plasmapheresis

It is also called plasma exchange in which damaging antibodies are removed from blood.

Radiation therapy

Irradiation of the lymph nodes is done to suppress immune system.

Fludarabine

It prevents the development and growth of malignant cells.

Tacrolimus

This transplant rejection drug may work to inhibit immune system.

Monoclonal antibodies

These are man-made antibodies designed to target and destroy specific types of cells.

Prognosis

The disease may spontaneously remit in 20% of patients. 5% of patients have fulminant progressive course with eventual death. Many patients require long-term therapy.[2,3]

Poor prognostic indicators

Poor prognostic indicators are recalcitrant disease, delay in diagnosis, old age, malignancy, fever, asthenia, anorexia, pulmonary interstitial fibrosis, dysphagia and leukocytosis. In juvenile dermatomyositis, late onset of treatment, initial treatment with a dosage of prednisolone that is too low, recalcitrant disease and pharyngeal involvement are associated with poor prognosis.[4,5,6]

Most common causes of death

Most common causes of death in patients of dermatomyositis are malignancy, cardiac or pulmonary dysfunction and infection.[4,5,6]

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