VOLUME - 12, ISSUE - 01, JANUARY - 2023 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Medicine

DERMATOMYOSITIS- A CASE REPORT AND REVIEW OF LITERATURE

Dr. Ravi Patil	Junior Resident
Dr. Ahmed Khan	Assistant Professor
Dr. Mitesh Thakkar*	Professor *Corresponding Author

ABSTRACT The idiopathic inflammatory myopathies are a heterogeneous group of disorders; excluding conditions that mimic them during the initial patient evaluation is essential. Classification of dermatomyositis (DM) for a definitive diagnosis requires a characteristic rash and other criteria, such as proximal muscle weakness and muscle enzyme level elevation. Dermatomyositis often overlaps with other connective tissue diseases. A photosensitive rash often is the initial manifestation. Cardiac and pulmonary involvement may be life-threatening. Malignancies occur in up to 25% of cases. A detailed strength examination at every visit is important for assessing treatment response. Physical therapy and occupational therapy should be started at diagnosis. Corticosteroids are the foundation of treatment. Most patients require corticosteroid-sparing medication.

KEYWORDS : Skin rash, Proximal muscle weakness, Creatinine phosphokinase, Skin biopsy

INTRODUCTION

Generalized rashes are among the most common conditions seen by primary care physicians.The initial approach to the patient presenting with a skin problem requires a detailed history of the current skin complaint and a complete skin examination.Some generalized rashes have distinctive features that allow immediate recognition, such as Tinea (flat red scaly lesion with central clearing or brownish dis colouration, Psoriasis (silvery white scale on the knees and elbows), Varicella(vesicles on erythematous papule-dewdrop on petal appearance), Pityriasis rosea (herald patch), Atopic dermatitis (lichenified skin in flexural areas), Erythema multiforme(round dusky skin lesion on palms and extensor surfaces arms and legs that evolve into target lesion), Insect bite(urticarial papule and plaques), Thrombocytopenia(pin point reddish purple spots that are easy to bruise) But these conditions, like many others, can present with similar appearances and can be mistaken for each other.Thorough history and physical examination, followed by consideration of red flags, are essential.Patients with muscle disorders are a diagnostic challenge to physicians, because of the various ways of presentation. A comprehensive approach should be followed systematically in order to reach the correct diagnosis. Weakness is a common symptom among patients including those with central or peripheral nervous systems diseases and those with muscular and/or neuromuscular diseases. Muscle weakness is not only a regular finding in rheumatologic diseases, but in inflammatory myopathies as well.Skin rash with muscle weakness predominately proximal muscle involvement indicates involvement of Derma(skin)-Myo(muscle). However, accurate diagnosis is important because treatment varies depending on the etiology, and because some rashes can be life-threatening if not treated promptly. Hence it requires multidisciplinary approach for diagnosis.

Review Of Literature

Dermatomyositis is one type of inflammatory myopathy characterized by inflammatory and degenerative changes of the muscles and skin.Incidence of Dermatomyositis is about 1 per 100,000 people per year with an estimated prevalence calculated to be about 20 cases per 100,000 people. Dermatomyositis has a bimodal distribution in the age of onset, occurring in two peaks, one at 5–14 years and the other at 45–64 years of life.The disease affects women approximately two to three times more than men.It presents with characteristic skin findings and symmetric proximal skeletal muscle weakness.Classic skin manifestations of DM include the heliotrope rash, Gottron's papules, Gottron's sign, the V-sign, and shawl sign. Additional cutaneous lesions frequently observed in DM patients include Periungual telangiectasias, cuticular overgrowth, "Mechanic's hands", Palmar papules overlying joint creases, Poikiloderma, and Calcinosis.Muscle symptoms initially presented as pains and weakness of the muscles of the trunk, upper arms, hips, and thighs (proximal muscles).

Interstitial lung disease can affect 35–40% of patients with inflammatory myopathies and is often associated with the presence of an antisynthetase antibody. Other clinical manifestations that can occur in patients with derma tomyositis include dysphagia, dysphonia, myalgias, Raynaud phenomenon, fevers, weight loss, fatigue, and non-erosive inflammatory polyarthritis. Patients with dermatomyositis have a three to eight times increased risk for developing an associated malignancy compared with the general population, and therefore all patients with dermatomyositis should be evaluated at the time of diagnosis for the presence of an associated malignancy.Clinically amyopathic dermatomyositis (CADM) is a condition in which patients have the characteristic cutaneous findings of dermatomyositis, but do not have muscle weakness.

There is ample evidence that DM is an immune-mediated disorder, given the immunohistopathology and given the response to immunosuppression.

The typical histopathologic findings of Dermatomyositis in muscle include Perifascicular atrophy, endothelial cell swelling, Vessel wall membrane attack complex (MAC) deposition, Capillary necrosis, Infarcts, Major histo compatibility complex (MHC) I upregulation, and the presence of an inflammatory infiltrate consisting of T and B lymphocytes, macrophages, and plasma cells.

There are findings of predominant perimysial and perivascular B/CD4+ T cell infiltrate and intravascular MAC deposition. Recent studies have provided evidence that 30–90% of the CD4+ cells found in dermatomyositis muscle are actually plasmacytoid dendritic cells.

The typical histopathological findings of dermatomyositis in skin include epidermal basal cell vacuolar degeneration, apoptosis of epidermal basal and suprabasal cells often with

epidermal atrophy, and increased dermal mucin deposition. A cell-poor interface dermatitis comprised of plasmacytoid dendritic cells at the dermal–epidermal junction is also characteristic. Features that are more specific to DM over cutaneous lupus include C5b-deposition in both dermal vasculature and the dermal-epidermal junction and a perivascular lymphocytic infiltrate.

For a diagnosis of dermatomyositis, patients must present with at least one of the following skin symptoms:

- Heliotrope rash – a red/purple rash with swelling on the upper eyelid
- Periungual telangiectasias – dilated capillaries at the skin folds around the fingernails, appearing as small red dots
- Gottron's sign or papules – red/purple hardened or eroding areas of skin on the upper surface of finger joints or over the elbows or knees
- Shawl sign, V-sign, and holster sign – characteristic red, sometimes raised rashes distributed in a "shawl" pattern over the shoulders, arms, and upper back, in a V-shaped pattern over the front of the neck and chest, and over the outside of the hip
- Skin biopsy demonstrating reduced capillary density, deposits of membrane attack complex on small blood vessels along the dermal-epidermal junction, and within the walls of dermal blood vessels

A diagnosis of dermatomyositis must also have at least four of the following criteria:

- Symmetrical muscle weakness in the shoulders/upper arms or hips/upper legs and trunk.
- Elevation of serum levels of skeletal muscle-associated enzymes: CK, aldolase, lactate dehydrogenase (LD or LDH), transaminases (ALT/SGPT and AST/SGOT)
- Muscle pain on grasping or spontaneous pain
- The triad of muscle-related changes on EMG:
- 1.Short, small, low-amplitude polyphasic motor unit potentials
- 2.Fibrillation potentials, even at rest
- 3.Bizarre high-frequency repetitive discharges
- Positive for any of the myositis-specific autoantibodies
- Nondestructive arthritis or arthralgias
- Signs of systemic inflammation
- Fever
- Elevated serum C-reactive protein (CRP) level, or
- Accelerated erythrocyte sedimentation rate (ESR)
- Muscle biopsy findings compatible with inflammatory myositis:

1. Perivascular inflammation with predominantly B-cells (with smaller numbers of Cd4- positive T-cells) accumulated around blood vessels
2. Perifascicular atrophy
3. Vasculitis involving endomysial and perimysial capillaries and arterioles
4. Active phagocytosis

Case Description

43 year old female presented to emergency room with unexplained rash with hyperpigmention all over body prominently on forehead, upper eyelids, neck, upper portion of chest. She also complaining of difficulty in getting up from squatting position, raising both arms or climbing stairs since last 2 months followed by difficulty in walking without support. The weakness was bilateral ,gradual in onset progressive to involve distal muscles of body. There was no history of fever, shortness of breath, dysphagia, nasal regurgitation, fasciculations.On general examination, her vitals were normal. Hyperpigmented maculopapular erythematous rash was seen on the forehead, upper eyelids, neck, upper part of chest and dorsum of hands.

The ear, nose and throat examination was normal. Neurological examination reveals normal higher mental functions and cranial nerves.Power of grade 3 at shoulder and elbow joint ,grade 4 at wrist joint, grade 3 at hip and knee joint with grade 4 at ankle joint.Tone was normal, all deep tendon reflexes were present with planters bilaterally flexors.The remainders of neurological examination was unremarkable.The respiratory, cardiovascular, abdominal and opthalmolgical examination were normal.

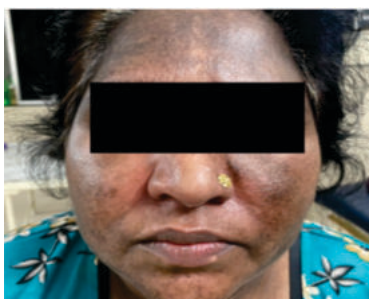
Investigation revealed
Hb-12.5 gm%, WBC-7800 /cumm, P78 L21 E1, platelets-3.4 lakhs/cumm, ESR-40 mm/1 h,CRP-27.7 mg/dl. Urine analysis, renal function tests and blood glucose were within normal limits.

The liver function test showed ALT-423 IU/L and AST 202 IU/L(reference: 0-40 IU/L).

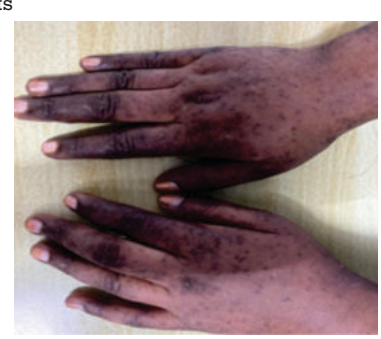
Creatinine phosphokinase(CPK)-13605 IU/L (reference: 24-190 IU/l), S. lactate dehydrogenase (LDH)-1147 IU/L (reference: 40-250 IU/L).

RA factor and Calcium were normal wiGOTTRON'S PAPULES-Voilaceous papules at knuckles at MCP and IP jointsth normal thyroid function tests. Anti nuclear antibody and other antibodies like C-ANCA and P-ANCA came negative.

Myositis profile was negative.Chest X-ray, ECG, 2D echocardiography, ultrasound abdomen and pelvis were normal.



Gottron's Papules- Voilaceous papules at knuckles at MCP and IP joints

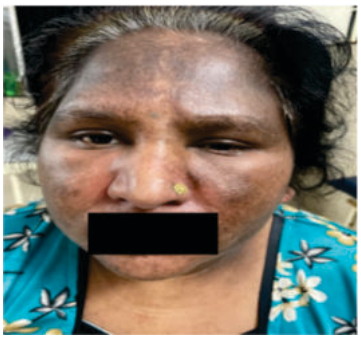


Hyperpigmented erythematous rash over cheecks

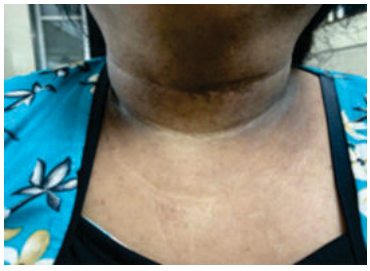
Electromyography (EMG) showed early and complete recruitment wit'V 'NECK SIGN-Erythematous rash over neckh polyphasic and low-amplitude motor unit action potential, suggestive of myopathic process affecting predominantly the **proximal and semidistal** muscles of upper and lower limbs more than face muscles.

Skin biopsy suggestive of focal basal vaculopathy and melanin pigment incontinence present in epidermis with perivascular and periadnexal inflammatory infiltrate comprising of lymphocytes 2d echo-within normal limit.

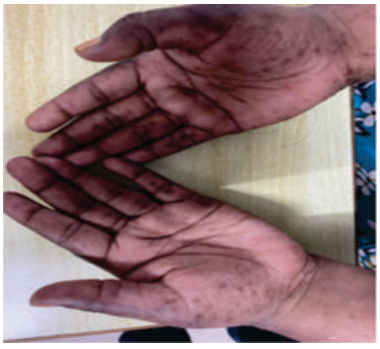
PAP smear-does not revealed any abnormality.



HELIOTROPE'S RASH-Purple discolouration on upper eyelids



V 'NECK SIGN-Erythematous rash over neck



Mechanic Hand-Dirty, horizontal lines over lateral and palmar areas of finger

SHAWL SIGN-Erythematous rash over back

CASE DISCUSSION

43 year old female with unexplained rash and proximal muscle weakness since couple of months, based on clinical, histopathological and laboratical findings, diagnosis of dermatomyotis was made.She was started on oral corticosteroid - Tab.Prednisolone 60 mg once a day and tapered gradually along with Calcium and multivitamin supplementation. She was also started on steroid sparing agent-Mycophenolate Mofetil 500 mg twice a daily for 7 days followed by 500 mg once a day for 1 month.

Her muscle power markedly improved with fall in creatinine kinase level tested at day of admission,5th day,10th day ,15th day,20th day of steroid therapy.

Day Of Steroid	0th	5th	10th	15th	20th
CPK	13605	6523	6123	4234	921

REFERENCES

1. Dermatomyositis: A case report Meenakshi Kalyan1, Shubhangi A Kanitkar1, Anu N Gaikwad1, Harsh Kumar2_https://www.jmgims.co.in/article.asp?issn=0971-9903;year=2016;volume= 21;issue=1;spage= 53;epage=55; aulast=Kalyan
2. Identifying and Managing Dermatomyositis: A Case Report and Review August 26, 2008 Philip N. Velderman, MD, Leslie A. Bruch, MD, George V. Lawry, MD The Journal of Musculoskeletal Medicine, The Journal of Musculoskeletal Medicine Vol 25 No 9, Volume 25, Issue 9 https://www.rheumatologynetwork.com/view/identifying-and-managing-dermatomyositis-case-report-and-review
3. Clinical Presentation and Evaluation of Dermatomyositis Authors: Umaima Marvi, Lorinda Chung, and David F Fiorentino https://www.ncbi. nlm.nih.gov/pmc/articles/PMC3482801/
4. The Myositis Association(TMA)