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Case Report

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A typical case of Dermatomyositis

Abstract

Dermatomyositis, a connective tissue disorder, is an idiopathic inflammatory myopathy characterized by skin manifestation. The diagnosis of dermatomyositis is based on characteristic skin rash, progressive muscle weakness, elevated serum muscle enzymes, abnormal EMG and muscle biopsy findings. Here we present an atypical case of a 68year old lady with all typical clinical findings failed to show response despite proper stepwise management.

Keywords: dermatomyositis, characteristic, Muscle weakness, skin rash

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Introduction

Dermatomyositis, classified as an idiopathic inflammatory myopathy, exhibits an incidence exceeding 4 cases per 100,000 individuals, with a prevalence ranging from 14 to 32 cases per 100,000. Typically diagnosed in individuals over 40 years old, females are affected at a rate twice that of males. Diagnosis relies on identifying key features such as characteristic skin rash, progressive muscle weakness, elevated serum muscle enzymes, abnormal electromyography (EMG), and muscle biopsy findings. Although dermatomyositis may be associated with underlying malignancy, advancing age is not a reliable predictor of the disease. Characteristic skin manifestations encompass the heliotrope rash, Gorton's papules, the V-sign, and Shawl sign. Some patients may also present with underlying Interstitial Lung Disease.

Case report

A 68-year-old lady presented to us with gradual onset progressive bilaterally symmetrical muscle weakness for past 3months. She complained of difficulty in combing her hair, getting up from squatting position and climbing stairs. 4months back, she visited a dermatologist as she experienced facial puffiness and rash over her forehead and cheek region which aggravated on exposure to sun. Subsequently similar rashes developed over her arms. There was no history of fever, dysphagia, or dyspnea. She denied of any addiction. Apart from topical ointment use as per her dermatologist, there was no drug use.[1,2]

On general physical examination, her vitals were within normal limits. There was facial puffiness along with maculopapular erythematous rash present over forehead, nose, cheek, arms, and knuckles. On Neurological examination higher mental functions were normal and cranial nerves intact. On motor examination there was power of grade 3/5 at hip and knee joint and 5/5 at ankle joint, 3/5 at shoulder and elbow and 4/5 at wrist joint. Tone was normal, all DTRs were present and plantar was bilateral flexor. Sensory system was intact, and rest of neurological, cardiovascular, respiratory and abdominal examination was unremarkable. Investigations: Hb-9.8gm/dl, TC-5800/cu mm, Platelets: 1.72lakhs/cu mm, ESR- 65mm, electrolytes and basic metabolic profile were within normal ranges. CPK-9532, serum LDH- 1342 was remarkably high. Ca 19.9 was 90 (normal: <37U/ml) EMG showed myositis pattern. Muscle biopsy showed inflammatory myopathy. Myositis panel showed Mi2a, Mi2b, RO-52 positive.[3]

Based on the above findings of proximal muscle weakness, characteristic rash, elevated CPK, serum LDH, muscle biopsy findings, diagnosis of dermatomyositis was confirmed. She was started on pulse therapy of Methylprednisolone and subsequently shifted to oral maintenance steroid. However, there was no remarkable improvement. She was administered IVIG and Rituximab which also failed to provide a favorable outcome (Figure 1–3).

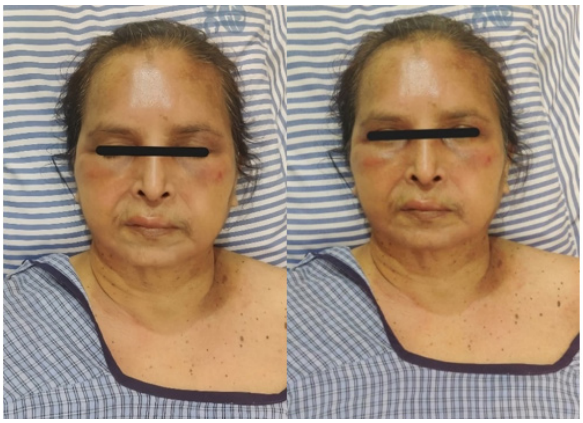

Figure 1 Heliotrope Rash in periorbital region.

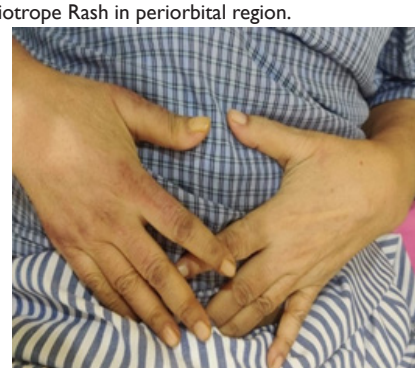

Figure 2 Gottrons's Papules: Inflammatory Violaceous papules over dorsal aspect of metacarpal and interphalangeal joints.

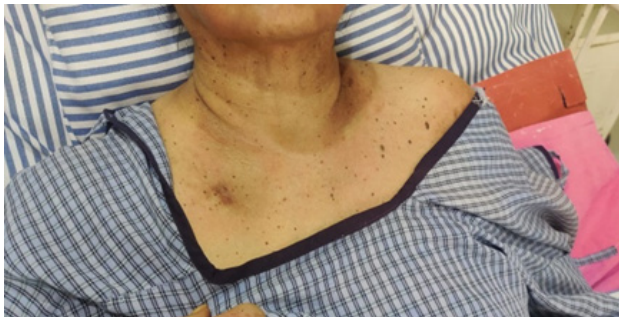

Figure 3 V sign is a patchy or discrete macular erythema over the sun-exposed parts of the anterior neck and upper chest in patients with dermatomyositis (DM).

Discussion

Dermatomyositis is an atypical inflammatory disease marked by proximal muscle weakness and characteristic skin rash. It is sometimes associated with underlying malignancy particularly pancreatic, lung, ovarian or colorectal. Anti-Mi-2B antibody is generally associated with favorable prognosis when treated with first line therapy, but in this case our patient inspite of getting stepwise management failed to show any response.[4] Old age and underlying malignancy are poor indicators for this disease. Patient had an increased level of Ca 19.9 and history of maternal aunt having pancreatic carcinoma which could have contributed to the poor prognosis. Mi-2 antigen, nuclear helicase protein, forms part of the nucleosome remodeling deacetylase (NuRD) complex involved in transcription regulation. Mi-2 is expressed highly in developing hair follicles and embryonic ectoderm which plays a critical role in the development of epidermis. Mi-2 is also essential for differentiation and renewal of the basal epidermis. Immune response to Mi-2 protein leads to the development of skin rashes and production of anti-mi-2 ab. Sunlight plays a significant role in anti-Mi-2 production through subcellular distribution, expression and metabolism of the components of Mi-2 antigens.[5–8]

Conclusion

Western literature often highlights the significance of the "heliotrope rash" and "Gottron's papule" in diagnosing dermatomyositis. However, it's crucial to acknowledge that these features may not be as easily discernible in individuals with darker complexions in our subcontinent. Therefore, vigilance is essential when a patient presents with proximal muscle weakness and facial edema, as Dermatomyositis should be considered as a potential differential diagnosis. While the presence of Anti Mi2b antibody is generally associated with a good prognosis, it's important to recognize that the disease course may change, especially if there is an undetected underlying malignancy, particularly of paraneoplastic origin.

Acknowledgments

None.

Conflicts of interest

The authors declare there is no conflicts interest.

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