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Case Report

Successful Treatment of Dermatomyositis in an Obese Female Patient

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Abstract

Dermatomyositis (DM) is a rare autoimmune illness that impacts youngsters and adults and is one of the idiopathic inflammatory myopathies (IIM) with skin involvement and the musculoskeletal system. Systems like, cardiac, respiratory, and gastrointestinal may additionally be affected up to a smaller degree. The predictable occurrence of DM is fewer than 10 cases per million inhabitants. Polymyositis has similar pathogenesis to dermatomyositis particularly targeting the musculoskeletal system without skin involvement.

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Introduction

Dermatomyositis (DM) is an uncommon autoimmune illness that impacts youngsters and adults and is one of the idiopathic inflammatory myopathies (IIM) with skin involvement and the musculoskeletal system. Systems like, cardiac, respiratory, and gastrointestinal may additionally be affected up to a smaller degree. The predictable occurrence of DM is fewer than 10 cases per million inhabitants. Juvenile DM is a subgroup of dermatomyositis and is normally detected between 5to15 years of age, while the analysis in adults can also manifest in the fourth and fifth decades. The disease shows purplish red skin rashes, which are particularly seen on the front and back of the chest, around the neck, cheeks, and knuckles.[1]

There are 6 specific kinds of dermatomyositis are

1. Classic dermatomyositis (CDM)
2. Amyopathic dermatomyositis (ADM)
3. Hypomyopathic dermatomyositis (HDM)
4. Clinically amyopathic dermatomyositis (CADM)
5. Basic dermatomyositis
6. Juvenile dermatomyositis.[2]

The definitive etiology of dermatomyositis is unclear. It is more commonly found in young females that are supposed to have high-intensity sunlight and ultraviolet radiations, though different inherited, ecological, infectious, and immunologic features influence dermatomyositis. The pathogenesis of dermatomyositis is an outcome of antibody-dependent immunologic reaction against the muscle vasculature. The process begins due to the activation of the complement system to its active fragments.[3]

The accurate test for the diagnosis of dermatomyositis is a muscle biopsy. The muscle biopsy shows neutrophilic infiltrates in the epimysium and perimysium of muscle. In dermatomyositis the infiltrates are grouped within muscle fascicles having B lymphocytes, CD4+ T lymphocytes, antigen-presenting cells like macrophages, and dendritic cells. Contrary to polymyositis, CD8+ T lymphocytes cells and natural killer cells are hardly present. Electromyography helps out in the involvement of a particular muscle group and its biopsy for diagnosis differentiates dermatomyositis from other neuropathies as well with low specificity. The characteristics findings of dermatomyositis comprise increased insertional motion, impulsive fibrillations, prominent sharp waves, Low-amplitude, and quick polyphasic motor unit potentials.[4]

Polymyositis has similar pathogenesis to dermatomyositis particularly targeting the musculoskeletal system without skin involvement. It is a subacute state that took 16 weeks to distinct. It is prevalent in adulthood affecting the proximal muscles of the body.[5]

Case Report

A 50 years old female tailor by profession (known hypertensive and diabetic for 10 years using medications but non-adherent with poor glycemic control) presented in the outpatient department with weakness and genera-

lized muscular pain involving the whole body for 2 months. Her weakness was the gradual onset and progressively increased. She felt difficulty in standing from sitting and squatting positions and was also unable to raise her both arms above her head, especially while combing her hair. With time she was unable to walk or climb stairs also. For the last 15 days, she noticed reddish, scaly, non-itching skin rashes over her face (butterfly), front and back of the chest, around the neck, and eyes sensitive to sunlight. Her muscular pain was aggravated by daily activities but was not associated with joint swelling or morning stiffness. The associated symptoms present were unilaterally temporal headache, hair loss, fingers discoloration, and swellings. There was no associated history of fever, oral ulcers, jaw claudication, dysphagia, dysphonia, or respiratory symptoms. Her bowel and bladder habits were normal. She was neither smoker nor an alcoholic with an increased appetite. She had a history of statin use for about 20 days prescribed by a local practitioner.

On general physical examination, she was obese, her weight was 148kg, and her BMI was 54 (Class III OBESITY). She had a heliotrope rash around her eyes, gottron's papules on her knuckles, a v-shaped rash on the front of her chest, a shawl-like pattern on the back of her chest, and a butterfly rash on her cheeks. GCS was 15/15, proximal myopathy in both upper and lower limbs with tenderness. Her muscle tone was normal; power is 3/5 in proximal muscles, and 5/5 in distal muscles of both upper and lower limbs with normal reflexes. Plantars were downgoing.

Labs included baseline investigations, Hb was 9.4, CPK was markedly raised 7783(normal 26-192IU), aldolase raised 200(normal 8-15IU) ANA+, anti dsDNA-, ENA profile showed markedly raised anti-Mi-2 antibodies while all other antibodies were insignificant.

The treatment given during admission was an injection of solumedrol (methylprednisolone) 1g IV OD for 5 days, after that shifted to oral steroids 80mg/day then tapered off. Immunosuppressant along used was azathioprine 50mg OD, Methotrexate 5mg once a week.

Figure 1: *Heliotrope rash present around the eyes, malar rash on cheeks, Rash on front of chest*

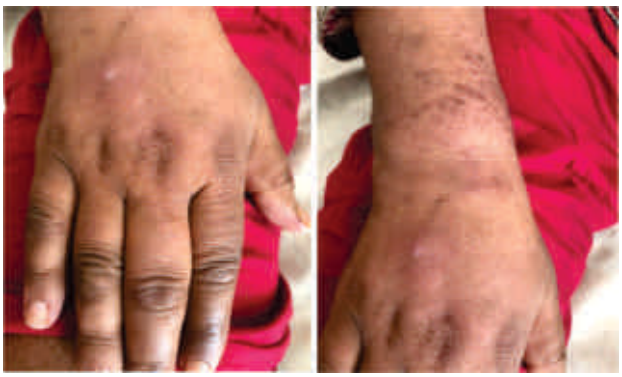
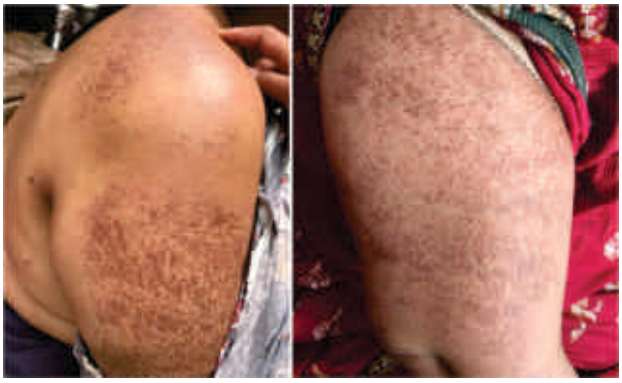
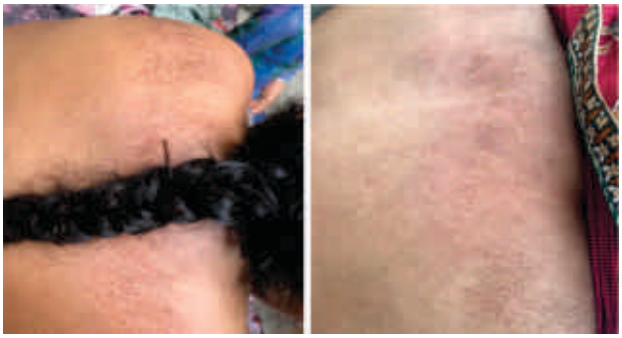


Figure 2: *Gottron's papules over the knuckles, rash on forearm*



Before Treatment *After Treatment*

Figure 3: *Rash on arm*



Before Treatment *After Treatment*

Figure 4: *Shawl like pattern on back of chest*

Discussion

Dermatomyositis is one of the infrequent myopathies. A longitudinal study done in the period duration 1967 -2007 in Olmsted county, Minnesota, shows an approximate dermatomyositis occurrence rate of 9.63/million population, including its amyotrophic subtype of 21% frequency rate. Dermatomyositis usually affects individuals in the fourth decade with a mean age at diagnosis of 44.0 ± 18.3 years. The disease is more prevalent in females as compared to males with an occurrence rate of 3.98 and 4.68 per million population respectively.[6]

The evaluated occurrence rate of all types of DM was

9.6/million rectifying for age and gender. A study shows the incidence rate of dermatomyositis subtypes is about 21.42% per million. One of the records shows the rate of new cases ranges from 1.1-17/per million population. Dermatomyositis has a stronger association with malignancy having an incidence rate ranging from 6% to 60% primarily involving the senile population but less risk of neoplasia as compared to polymyositis.[7]

Dermatomyositis primarily manifests as cutaneous problems. It is mandatory to cover the exposed body parts with clothing and apply sun blocks because of the risk of photosensitive reactions. Antimalarial drugs like chloroquine and hydroxychloroquine have a high-yielding result in the resolution of dermatomyositis with few drug eruptions which can be overcome by methotrexate administration.

The therapy choices for resistant cases comprise monoclonal antibodies, immunosuppressant, intravenous immunoglobulin (IVIG), and pulse therapy. Azathioprine and IVIG have fruitful results in treating various types of myopathies. A study was conducted to compare the outcome of the use of conventional immunosuppressive therapy alone versus intravenous immunoglobulins (IVIG) as an add-on therapy. After 6 months, there is marked resolution in musculocutaneous manifestation with the combine use of immunosuppressive therapy and IVIG i-e. dual therapy as compared to isolated conventional immunosuppressive therapy.[8]

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