Dermatomyositis – A Rare Case with Excellent Outcome

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Abstract

Dermatomyositis and Polymyositis are autoimmune rheumatologic diseases with prominent feature of inflammatory myopathy. When the characteristic cutaneous manifestations coexist with myositis it is called dermatomyositis, otherwise it is polymyositis. There are also differences in histological findings. A 25-years-old lady presented with characteristic skin rash and progressive inflammatory proximal myopathy. The patient had difficulty in Activity of Daily Living (ADL) and ultimately she was unable to walk and became bed bound. Her muscle enzymes were very high, MRI and Electromyography (EMG) showed features of inflammatory myopathy (myositis). Muscle biopsy showed typical findings of dermatomyositis. She responded well with steroid and immunosuppressive drug along with exercise, physiotherapy and rehabilitation therapy. Now she is leading almost normal life with minimum maintenance dose of prednisolone and azathioprine.

Key-words: Dermatomyositis, skin rash, myositis, EMG, MRI, Muscle biopsy, prednisolone, Azathioprine.

Introduction

Dermatomyositis (DM) is an idiopathic inflammatory myopathy with characteristic cutaneous findings. It is a systemic disorder that most frequently affects the skin and muscles, but may also affect the joints, esophagus, lungs and heart. In majority of cases the cause is autoimmune. Malignancies such as carcinoma of stomach, pancreas, breast, ovary, colon, lymphoma etc may produce dermatomyositis as paraneoplastic manifestation. Autoimmune dermatomyositis must be differentiated from polymyositis (PM), Systemic lupus erythematosus (SLE), MCTD (mixed connective tissue disease), paraneoplastic dermatomyositis/polymyositis, myasthenia gravis, inclusion body myositis, hereditary and drug and toxin induced myopathies[1]. If untreated the disease is fatal due to respiratory failure, aspiration pneumonia, cardiac complications, AKI (Acute kidney injury), GI bleeding and associated malignancy. But treatment is rewarding with excellent outcome. Early diagnosis and appropriate management strategies are the key to success. In CMH Dhaka, since 2010, we have diagnosed and successfully treated 5 cases of dermatomyositis. Here one case is reported who was admitted to CMH Dhaka in March 2017. The main objective of publishing this case report is to generate awareness amongst the medicos regarding the early diagnosis and proper treatment of dermatomyositis[2].

Case Report

A 25-year-old lady presented to Department of Rheumatology, CMH Dhaka, on March 2017 with the history of skin rash for 3 months and gradual and progressive weakness of all four limbs for 2 months. Initially she developed erythematous rash on forehead which subsequently spread over face, nose, chin, neck and extensor surface of fingers and knuckles. She noticed weakness and pain in muscles with difficulty in climbing stairs, sitting from lying and standing from sitting or squatting posture for 2 months. She felt difficulty in combing hairs, raising the arms above the head and lifting heavy objects. She also noticed weight loss and fatigability for the same duration. She gave no history of fever, joint pain, oral ulcer, hair loss, drooping of eyelids, difficulty in breathing, swallowing and speech. There was no history of cold or heat intolerance and no history of skin color change on exposure to cold. Her bowel and bladder habit were normal.

She had no past history of thyroid disorder, malignancy, myasthenia gravis, Cushing's Syndrome, diabetes, hypertension, bronchial asthma or any rheumatological diseases. There was no history of taking steroid, statins, penicillamine, antiepileptic or any other offending drugs.

Clinically she was ill looking, anxious and mildly anemic. Vitals were normal. She had heliotrope rash in eyelids (Figure-1), erythematous rashes in face, nose, chin, upper chest (necklace area: "V" sign) (Figure-2), back of the neck and shoulder (shawl sign) (Figure-3), extensor surface of distal interphalangeal (DIP), proximal interphalangeal (PIP) and metacarpophalangeal (MCP) joints (Gottron's sign) (Figure-4). Examination of locomotor and nervous system revealed tenderness in muscles of arms and thighs with muscle power 2/5 in upper limbs and 3/5 in lower limbs. Her cranial nerves, sensory functions, coordinations and deep tendon reflexes and plantar reflexes were normal. She could walk unaided with some difficulty.

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Basing on history and clinical findings she was provisionally diagnosed as a case of dermatomyositis. She was extensively investigated to confirm the diagnosis, to rule out other possibilities and underlying malignancy. Her CRP was positive, serum CPK - 20800 U/L (normal 24-190 U/L), serum Aldolase – 32 U/L (normal < 7.6 U/L), serum LDH – 3080 U/L (normal 225-280 U/L), ALT – 299 U/L (normal up to 45 U/L), urine myoglobin was positive (50 mg/dl). Her Hb was 11.7 gm/dl, ESR – 26 mm in 1st hour, TLC, DLC, platelet were within normal limit. RBS, serum creatinine, electrolytes, serum bilirubin, alkaline phosphatase etc were also within normal limit. RA, ANA, anti-ds DNA, anti Jo1 antibody, HBsAg and anti HCV were negative. Common tumor markers were negative. ECG, chest x-ray, USG of breasts and whole abdomen were normal. Upper GI endoscopy showed chronic gastritis and colonoscopy was normal. MRI of both thighs was done which revealed symmetric diffuse high signal intensity change in the muscle, skin and subcutaneous tissues of the thigh which is suggestive of dermatomyositis (Figure-5). Cross limbs EMG showed features of myositis (short amplitude, fibrillary waves, excessive spontaneous activity, polyphasic potentials of short duration and low amplitude, fibrillary waves) (Figure-6). Biopsy from affected muscles (quadriceps) revealed infiltration of lymphocytes and macrophage in perivascular, perimysial, perifascicular distribution along with myonecrosis, degenerative and regenerative fibers with phagocytosed muscle fiber (Figure-7).

Therefore, she was diagnosed as a case of Dermatomyositis. After adequate education and counselling she was started with high dose oral prednisolone and tablet azathioprine as immunosuppressant and steroid sparing agent. She was vaccinated against influenza and pneumococcus. She was also supplemented with calcium and vitamin D. She was advised for passive and active exercise, physiotherapy and rehabilitation therapy.

Before treatment (during the period of investigations) and initial few days of treatment her condition further deteriorated. She was unable to perform ADL and was totally incapacitated and became bed bound due to severe muscle weakness (Muscle power 0-1/5). Then her condition improved gradually. She could walk with assistance within 2 months and could walk independently after another 2 months. But during hospital stay she developed steroid psychosis and iatrogenic Cushing's which was managed by antipsychotic and gradual tapering of steroid.

After satisfactory improvement she was discharged on August 2017 with tapered dose of prednisolone and azathioprine. She was advised to continue medicine, exercise, take a high protein diet and follow up periodically. She was followed up at 1, 3, 6 and 12 months. Her improvement was progressive and sustained. Now she is leading a normal life with minimal maintenance dose of prednisolone and 50 mg of azathioprine daily. Currently she is pregnant for 7 months and her pregnancy is uneventful till date. There are no untoward effects of medication. Screening at yearly follow up revealed no evidence of malignancy.

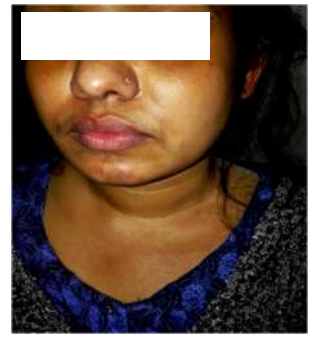

Figure-1*: Heliotrope rash in the case.

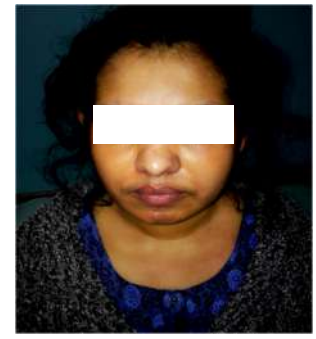

Figure-2*: "V" sign in the case

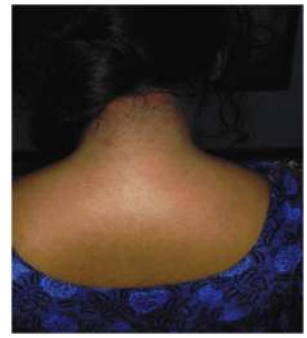

Figure 3*: Shawl sign

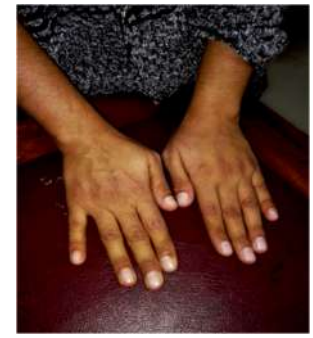

Figure-4*: Gottron's papules.

*Photo is published upon taking permission from the patient

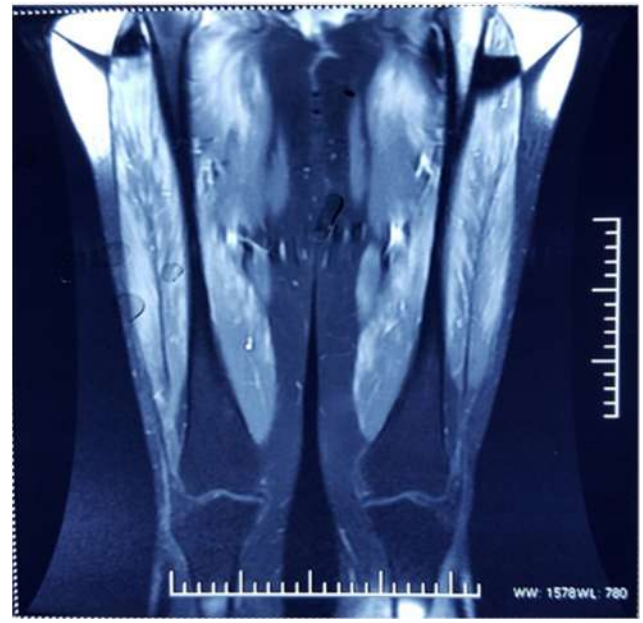

Figure-5: MRI of thigh showing symmetric diffuse high signal intensity change in the muscle, skin and subcutaneous tissues

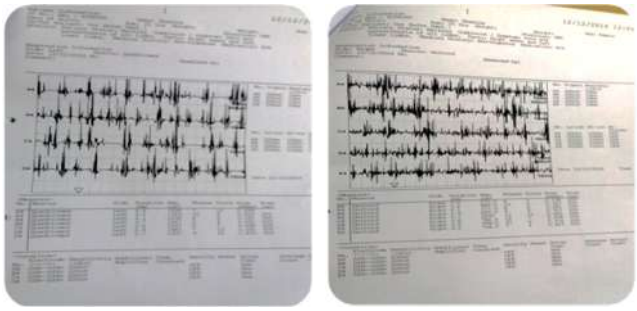

Figure-6: EMG of left and right deltoid showing polyphasic potentials of short duration and low amplitude

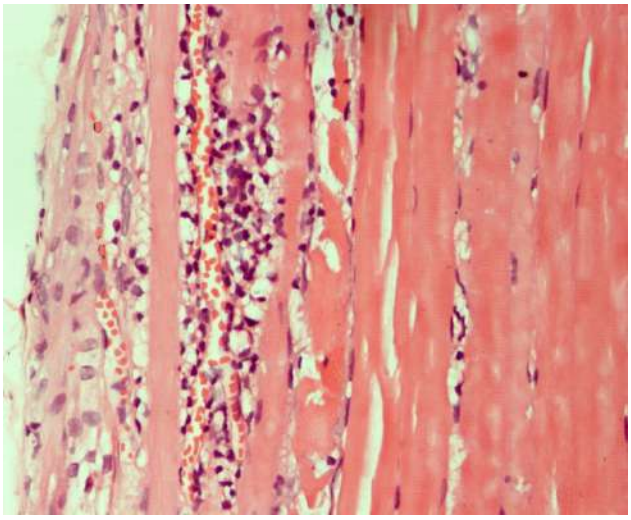

Figure-7: Muscle biopsy of rectus femoris showing perivascular and perimysial infiltration of lymphocytes with myonecrosis

Discussion

Dermatomyositis is an idiopathic inflammatory myopathy with characteristic cutaneous findings. It is a systemic disorder that most frequently affects the skin and muscles, but may also affect the joints, esophagus, lungs and heart[3].

Diagnostic criterion[4] for dermatomyositis as follows:

a. Progressive proximal symmetrical weakness
b. Elevated levels of muscle enzymes
c. An abnormal finding on electromyography
d. An abnormal finding on muscle biopsy
e. Cutaneous disease

Definite dermatomyositis is defined when there is presence of rash + 3 other criteria. Probable dermatomyositis means rash + 2 other criteria and possible dermatomyositis is rash + 1 other criteria.

Patients with dermatomyositis presents with proximal myopathy, skin rash, systemic features such as fever, malaise, weight loss, arthralgia, dyspnea, dysphagia, dysphonia, raynaud's phenomenon, arrythmia etc. Skin manifestations include heliotrope rash, "V" neck sign (Figure 8), Gottron's papule (Figure 9), shawl sign, mechanics hand (Figure 10), hyperpigmentation and periungual telangiectasia (Figure 11)[5]. Association with different types of malignancy, specially lung cancer in men and ovarian cancer in female may occur. Other malignancy like colonic, nasopharyngeal, gastric carcinoma and lymphoma may also occur. Connective tissue disease like SLE, Systemic sclerosis, Rheumatoid Arthritis etc can be present as well[6].

An array of investigations helps to diagnose dermatomyositis. Muscle enzymes like CPK, Aldolase, ALT, AST, LDH, Carbonic anhydrase lll are raised. Antibodies like ANA, Anti Jo-1 (anti synthetase antibody), Anti MI-2 Ab, Anti RNP Ab, Anti SRP Ab are often positive[7]. MRI of muscles identify areas of muscle inflammation, atrophy and fatty replacement. EMG shows abnormal electrical irritability (fibrillation, positive sharp waves), increase in the percentage of polyphagic motor unit potentials and decrease in the mean duration and amplitude of motor unit potentials[8]. Muscle Biopsy shows perivascular, perimysial and interfascicular lymphocytic infiltration, mainly CD4 T-cell and occasionally B-cell and phagocytes with adjoining group of muscle fiber necrosis, degeneration/ regeneration[9].

Corticosteroids are the 1st line drug. Immunosuppressive agents like Azathioprine, Methotrexate, Cyclosporine, Cyclophosphamide, Mycophenolate mofetil, Tacrolimus are used. Hydroxy-chloroquine may be used for persistent skin rash. Biologics like Rituximab and anti TNF are effective in case of resistant myositis. IV immunoglobulin and plasmapheresis are reserved for fulminant cases. Calcium, Vitamin D and bisphosphonates are also needed as supplements[10]. Exercise and Physiotherapy, Assistive devices, Orthotics are advised to patients according to their requirements[11]. Surgical excision of calcinosis can be done when it is large, painful or there is recurrent infection. Corrective surgery for contracture and deformities are also considered in advanced cases[12].

Without proper treatment a wide range of complications may occur. Respiratory complications like respiratory failure, aspiration pneumonia, pulmonary fibrosis, pulmonary hemorrhage can occur. Cardiac complications such as myocarditis, arrhythmias, conduction defect, cardiomyopathy; GIT complications like intestinal vasculitis and perforation, large bowel infarction and renal complications like acute kidney injury, focal mesangial proliferative glomerulonephritis may occur[13,14]. Malignancies such as gastric carcinoma, ovarian carcinoma, lung carcinoma, breast carcinoma and lymphoma can occur. Muscle may become atrophied and skin calcinosis may occur[15].

Natural history of the disease shows 20% spontaneous remission. 5 years survival in 85% cases is seen. 5% have fulminant progression and eventual death. Poor prognostic factors are recalcitrant disease, older age, malignancy, fever, pulmonary interstitial fibrosis, dysphagia and leukocytosis. Prognosis is better if presentation is on isolation, early presentation, young age, without having malignancy and absent

anti synthetase antibody. Mode of death in dermatomyositis is usually due to cardiac and pulmonary dysfunction, malignancy, acute kidney injury and infection[16].

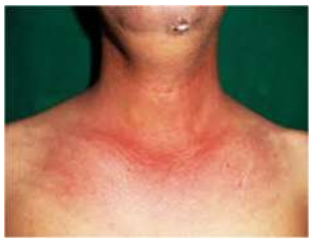

Figure-8: Typical "V" Sign

Figure-9: Typical Gottron's papule

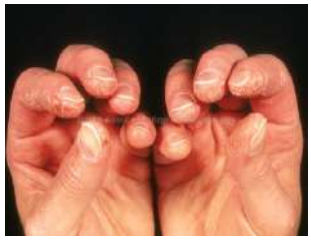

Figure 10: Typical Mechanics hand

Figure 11: Typical Periungual telangiectasia

Conclusion

Dermatomyositis is a progressive inflammatory myopathy with characteristic skin rash. The outcome is fatal if remains untreated. Mortality is mainly due to cardiorespiratory complications. High index of suspicion is required to diagnose the case. Adequate knowledge and experience is also needed to manage the case successfully. It has positive association with malignancy. So, malignancy screening is mandatory during initial workup and future follow-up. Early diagnosis and appropriate treatment is very much rewarding and can prevent disability and fatality.

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