Case Report

A Case of Dermatomyositis : Diagnostic Challenge in Low Resource settings

Richmond Ronald Gomes[1], Sanjoy Saha[1], Khan Shakil Ahmed[2], Masud Imtiaz[3], Serajul Islam[4], Mir Moyeedul Islam[5]

Abstract

Dermatomyositis is one of the idiopathic inflammatory myopathies. It is characterized clinically by progressive symmetrical proximal muscle weakness and a characteristic rash. Although the process primarily attacks the skin and the muscles, it is a systemic disease with frequent manifestations in the gastrointestinal tract and pulmonary system. Dermatomyositis has been linked to internal malignancy in somewhere between 15% and 25%. This case report involving a 30 years old fair female nurse who presented with extensive erythema involving face, upper neck, upper back and deteriorating proximal muscular weakness for three months. Later muscle enzymes, EMG and biopsy were done from BSMMU, Dhaka, confirmed the diagnosis of Dermatomyositis. Oral prednisolone along with azathioprine, hydroxychloroquine and photo protection with sun screen were initiated and showed good response both clinically and biochemically.

Key words : Dermatomyositis, heliotrope rash, shawl sign, poikilodermic changes

Introduction

Dermatomyositis is one of the idiopathic inflammatory myopathies[1-3]. It has two peaks of occurrence, one in childhood and one between the age of 45 and 65 years[4-5]. The estimated annual incidence rates ranged from 2 to10 cases per million6. In 1975, Bohan and Peter[4] published a classic article that suggested a set of criteria to aid in the diagnosis and classification of Dermatomyositis and polymyositis (PM). Of the 5 criterias, 4 related to the muscle disease: (1) progressive proximal symmetrical weakness (2) elevated muscles enzymes, (3) an abnormal electromyogram, and (4) an abnormal muscle biopsy, while the fifth was the presence of compatible cutaneous disease.

It was felt that DM differed from PM only by the presence of cutaneous disease. Recent studies of the pathogenesis of the myopathy have been controversial, some suggesting that the myopathies in DM and PM are pathogenetically different with DM being due to a vascular inflammation[5], whereas other studies of cytokines suggest that the processes are similar[6-9]. There has been a renewed interest in the pathogenic mechanisms involved in the myopathy with recent studies revealing abnormal levels of nitric oxide, elevation of circulating tumor necrosis factor (TNF) receptors, elevated soluble CD40 expression, and increased expression of major histocompatibility complex class I and interleukin 1a within the muscle. The pathogenesis of the cutaneous disease is poorly understood.

1. Assistant Professor & Head, Department of Medicine, Ad-din Sakina Medical College, Jessore.
1. Assistant Professor & Head, Department of Pharmacology & Therapeutics, Ad-din Sakina Medical College, Jessore and PhD fellow, Bangladesh University of Professionals, Dhaka.
2. Professor (CC) & Head, Department of Forensic Medicine, Ad-din Sakina Medical College, Jessore.
3. Associate Professor & Head, Department of Physiology, Khulna City Medical College, Khulna.
4. Assistant Professor & Head, Department of Community Medicine, Ad-din Sakina Medical College, Jessore.
5. Assistant Professor, Department of Pharmacology & Therapeutics, Ad-din Sakina Medical College, Jessore.

Correspondence : Richmond Ronald Gomes
email : richidmck56@gmail.com

Case Presentation

On 15th January 2015, a 30 years old fair female married nurse came to the department of Medicine of Ad-din Sakina Medical College & Hospital, Jessore, Bangladesh with extensive photo sensitive erythema involving face, neck, upper chest, upper back; alopecia and deteriorating proximal muscular weakness for three months. But patient denied any joint pain, oral ulcer, history of abortion, haematuria, features supporting raynaud's phenomenon, any respiratory and gastrointestinal complaints. On examination there was wide spread violaceous erythema involving face, "V" of neck and upper back (shawl sign). There were some poikilodermic changes (atrophy, hypopigmentation, talangiectesia) over upper aspect of the back with focal alopecia. Some periorbital oedema was also noted with heliotrope rash. But gottron's papules, hyperkeratosis of hands (mechanical hands), periungual talangiectesia and holsters sign (poikiloderma of upper

lateral thigh) were absent. Neurological examination revealed reduced proximal muscle strength (3+/5) in both upper and lower limb compared to distal power (5/5). No other focal neurological signs were elicited. Cardiovascular, respiratory and abdominal examinations were unremarkable.

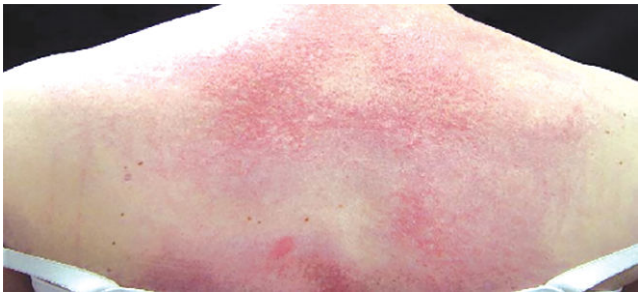

Figure-1 : Poikiloderma on the upper aspect of back is typical of the shawl sign.

Baseline haematological and biochemical investigations revealed normal complete blood count, Hb%-12.4gm/dl, WBC-7600/cmm, Neutrophil-50%, Lymphocyte-42%, Eosinophil-4%, total platelet count 393000/c mm. Inflammatory markers were elevated with ESR-42 mm in Ist hour and CRP 45. Lactate dehydrogenese (LDH), aspartate amino transferase (AST), levels were elevated at 2290U/L and 243U/L respectively. Her Creatin phosphokinase (CPK) level was significantly raised with 13738/L (Normal: 30-135U/L). Antinuclear antibody (ANA), Anti double-stranded DNA, Rheumatoid factor, ENA (extractable nuclear antigen) profile, HBsAg were negative. electromyography (EMG) findings were consistent with inflammatory myositis. Skin biopsy from upper chest revealed hyperkeratosis, keratotic plugging, mild atrophy of the epidermis, basal liquefaction and lympho histiocytic infiltration at dermoepidermal junction as well as
perivascular region in the upper dermis. Cancer screening including chest X-ray and ultrasonography of whole abdomen were normal. Based on the clinical features and investigations, the patient was diagnosed as a case of Dermatomyositis.

Her treatment was started with oral prednisolone along with azathioprine, hydroxychloroquine and photo protection with sun-screen. With treatment there was a clear response to treatment with resolution of skin lesions, gradual improvement of her muscular weakness and reducing level of CPK and LDH (780U/L and 417U/L respectively after one month of treatment). She was advised for regular follow up.

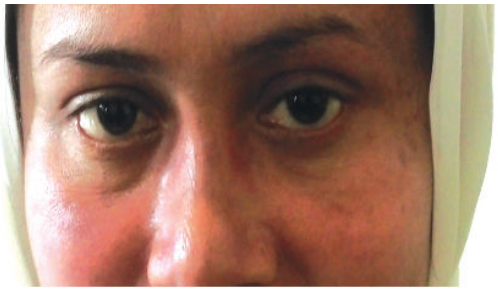

Figure-2 : Mild periorbital oedema with marked facial Erythema

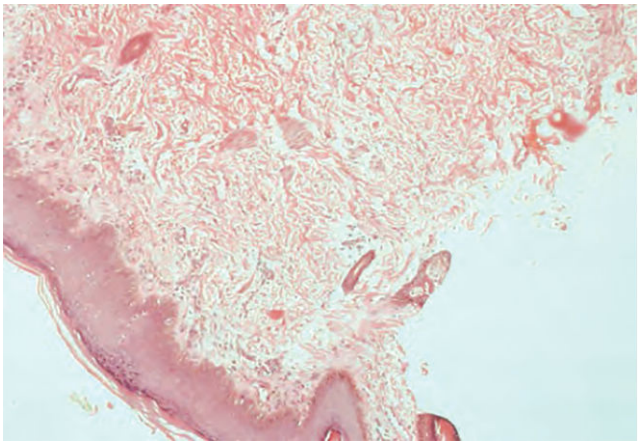

Figure-3 : Histopathology slide showing hyperkeratosis, keratotic plugging, basal cell liquefaction and lymphohistiocytic infiltration in to the upper dermis

Muscle/ Slide	Inser. Act	Fibs	Pos. wave	Fasc.	MYO Disch	Normal MUP	Poly	Low Amp	High Amp	Dur	Rec	Int. Patt
TA L	Inc	+2	+2	0	0	0	N	+3	0	Sh	F	F
Q L	Inc	+1	+3	0	0	0	N	+3	0	Sh	F	F
Q R	Inc	+3	0	0	0	0	+++	+3	0	Sh	F	F
GM L	Inc	+3	0	0	0	0	++	+2	0	Sh	F	F

TA: Tibialis Anterior, Q: Quadriceps, GM: Gluteus Medius, Inc: Increment, Sh: Short, F: Full
EMG findings Summary: Inflammatory myopathy

Discussion

Dermatomyositis (DM) is one of the idiopathic inflammatory myopathies[1-3]. Bohan and Peter[4] suggested 5 subsets of myositis—DM, PM, myositis with cancer, childhood DM/PM, and myositis overlapping with another collagen vascular disorder. In a subsequent publication, Bohan et al[10] noted that cutaneous disease may precede the development of the myopathy; however, it was only recently recognized that another subset of patients with disease that only affects the skin (amyopathic DM [ADM] or DM-sine myositis) may occur[11]. A seventh subset known as inclusion body myositis has been recognized in 1979[12,13]. Perhaps there is an eighth group in which characteristic cutaneous disease is drug-induced[14]. Finally, Sontheimer[15] has proposed that other subsets exist for patients with cutaneous disease including classic DM, ADM, and at least two additional subsets known as hypomyopathic DM, when the skin disease is present with subtle muscle disease, evident with studies other than enzymatic analysis, and finally, a subset known as post-myopathic DM when patients with previous classic DM have the myositis resolve, but the skin disease remains active.

DM is autoimmune in pathogenesis and results from a vasculopathy. Both cell mediated immunity to muscle antigen and immune complex disease may play role in the pathogenesis[16]. Along with symmetrical proximal muscle weakness the characteristic and possibly pathognomonic cutaneous features of DM are the heliotrope rash and Gottron's papules. Other skin manifestations of DM include, erythematous malar rash, confluent macular violaceous erythema overlying the extensor aspect of the upper extremity, V area of anterior neck and chest, central aspect of the face, periorbital areas, forehead of the scalp, lateral aspect of the hip and thigh, periungual telangiectasia, poikiloderma, hyperkeratosis or mechanical hands, cuticular over growth, panniculitis, cutaneous vasculitis. The skin lesions of DM are probably photoaggravated. Clinical observations suggest that not only is the skin disease exacerbated by light, but muscle disease may be worsened after sun exposure[17-20]. Phototesting has however not been able to reliably reproduce the skin lesions; thus, the wavelength of light that is responsible for the clinical manifestations (action spectrum) is not known. Scalp involvement in DM is relatively common and is manifested by an erythematous to violaceous, psoriasiform dermatitis[21]. in our case there was focal non scaring alopecia.

Nitsche et al first published a case of widespread subcutaneous edema as part of the dermatomyositis syndrome in 1988[22] in contrast to some periorbital oedema noted in our case. The underlying pathogenesis for the subcutaneous edema remains to be elucidated. It has been thought that increased vascular permeability in the tissues and muscles leads to extensive leakage of fluid into surrounding structures[23]. This implies that subcutaneous edema may be a result of severe inflammation and an indirect indicator of aggressive disease. On the other hand, there may also be a role for an immune complex mediated vasculitis[24].

Recently, evidence has emerged linking the pathogenesis of dermatomyositis to type I interferons. A case report of severe dermatomyositis exacerbated/induced by interferon beta therapy was published in 2008, supported by in vitro evidence of enhanced type 1 interferon signaling in response to interferon beta[25]. The optimal treatment of dermatomyositis associated remains unclear. The mainstay of therapy involves glucocorticoids, which are thought to act through anti-inflammatory and immunosuppressive effects[25,26]. However, additional immunosuppressive agents such as azathioprine, hydroxychloroquine, mycophenolate-Mofetil and methotrexate are often employed as a more aggressive attempt to gain control of disease. IV Ig has also been administered in severe, life threatening cases; eight out of nine patients given IVIg eventually recovered from their illness[26-33]. Newer biological agents have shown great promise in refractory cases of dermatomyositis. Rituximab has been successfully used in the treatment of refractory dermatomyositis and other inflammatory myopathies[34, 35].

Conclusion

Dermatomyositis is a condition primarily of skin and muscle, but other systemic features may occur. The pathogenesis of the muscle disease is becoming better understood but the cutaneous disease mechanisms remain enigmatic. Dermatomyositis in adults is associated with malignancy. So that, to exclude malignancy proper evaluation of each patient is necessary during initial and follow-up assessments. Patients should also be evaluated for the presence of esophageal, pulmonary and cardiac disease. Corticosteroids, immunosuppressive agents, biologic agents, and/or immune globulin are effective therapies for the myopathy of DM, whereas the skin disease is best managed with sun protection, topical corticosteroids, antimalarials, methotrexate, and/or immune globulin.

The prognosis is good except for patients with malignancy, those with severe weakness, and those with cardiac dysfunction, interstitial lung disease, or the presence of a myositis-specific autoantibody other than Mi-2.

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