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Case Report

An interesting case of dermatomyositis with classical presentation

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ABSTRACT

Dermatomyositis, a connective tissue disorder, is an idiopathic inflammatory myopathy characterised by skin manifestation. The diagnosis of dermatomyositis is based on rashes on the skin, progressive muscle weakness, elevated serum muscle enzymes, abnormal electromyogram, and abnormal findings on muscle biopsy. Hereby presenting this rare case of a 57-year-old female with dermatomyositis with all the typical clinical findings with interstitial lung disease.

Keywords: Dermatomyositis, Classical, Tissue disorder

INTRODUCTION

Dermatomyositis is an idiopathic inflammatory myopathy.[1] The prevalence of dermatomyositis is 1 in 100,000 while women are more affected than men.[2] Most of the patients are diagnosed at 40 years of age, with females getting affected almost twice than male. The diagnosis of dermatomyositis is based on rashes on the skin, progressive muscle weakness, elevated serum muscle enzymes, abnormal electromyogram, and abnormal findings on muscle biopsy.[3] This disease is associated with high rate of high rate of malignancy especially in the older age group. Diagnostic criteria include typical cutaneous features, progressive proximal symmetrical muscle weakness, elevated muscle enzymes and abnormal findings from muscle biopsy.[4] Classic skin manifestations of DM include the heliotrope rash, Gottron's papules, the V-sign, and shawl sign.

In approximate 40% of patients with inflammatory myopathies, Interstitial lung disease can also occur.

CASE REPORT

A 57-year-old female was brought to the hospital with the complains of multiple ulcerative lesions over the dorsum of the fingers since past 4 months. Complains of multiple joint pain and swelling for the past 4 months. Patient had shortness of breath for 10 days and Past history revealed alopecia (Figure 1) and weight loss since past 10 months.

On examination of the neck, v neck sign was seen (Figure 4), on examination of the chest, shawl sign was seen (Figure 5), Gottrons papules and contractures were seen over the left hand (Figure 3), bilateral Velcro crepitation over the base of lungs were present, PET CT showed diffuse active inflammatory poly myositis (Figure 2) and fibrotic changes in B/L lungs, CPK levels were 3690, muscle biopsy showed inflammatory myopathy, suggestive of dermatomyositis. Anti MI 2 antibody was found to be strong positive.

Figure 1: Alopecia.

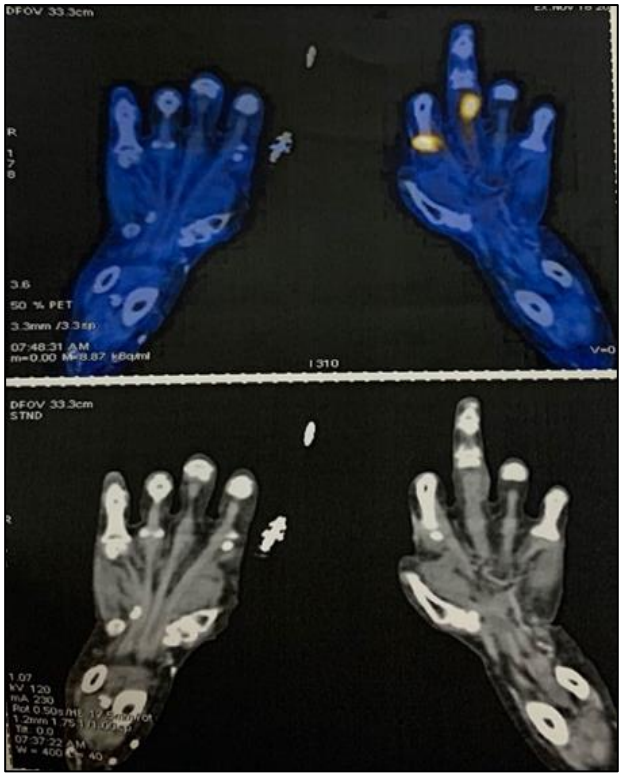

Figure 2: Diffuse active inflammatory poly myositis.

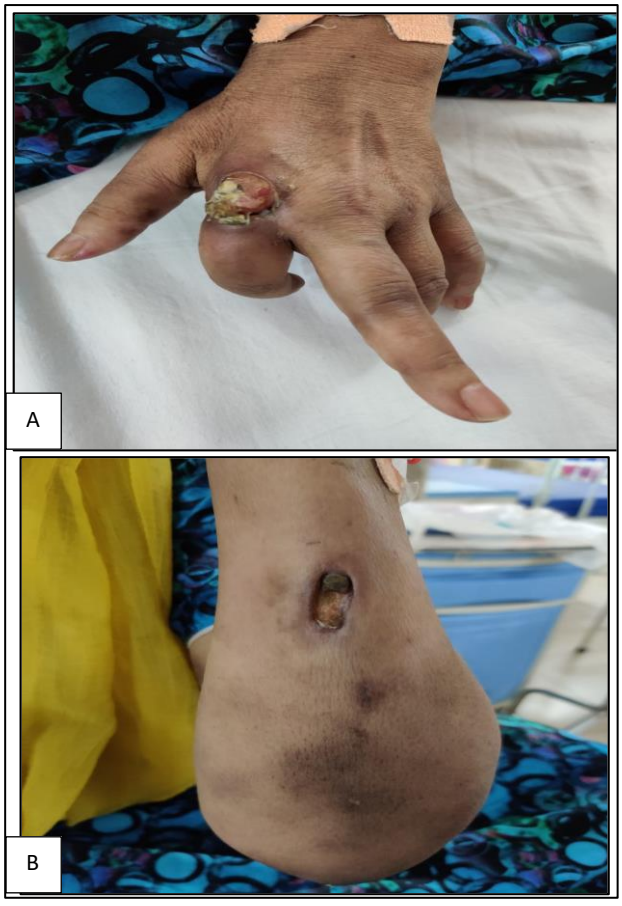

Figure 3 (A and B): Contractures and gottron's papules.

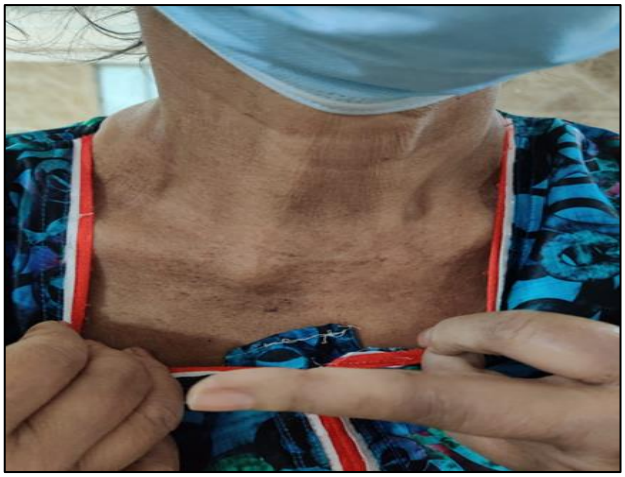

Figure 5: V neck sign.

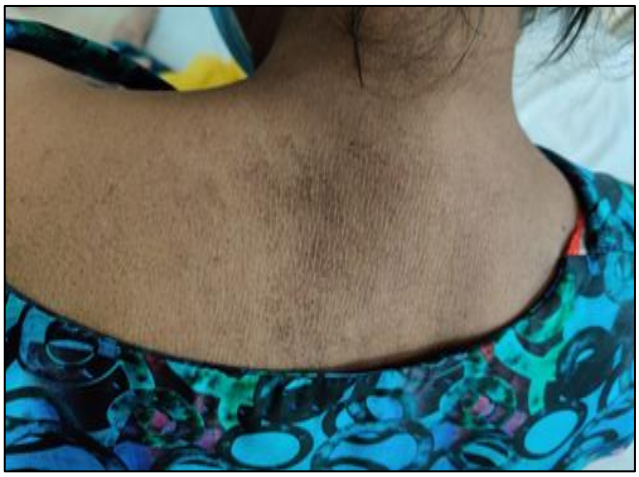

Figure 6: Shawl sign.

Course in the hospital

The patient was initially treated with inj methyl prednisolone 500 mg 3 days, inj cyclophosphamide 700 mg monthly pulse, tab. prednisolone 30 mg od, inj. Mesna 400 mg before and 400 mg after cyclophosphamide.

The patient improved clinically and was discharged with the following advice: Tab Prednisolone 30 mg od, Tab Aspirin 75 mg od and Tab Pantoprazole 40 mg od. The patient was asked to review monthly for cyclophosphamide cycles.

DISCUSSION

Dermatomyositis is an uncommon inflammatory disease marked by muscle weakness and a distinctive skin rash. Dermatomyositis is associated with high degree of malignancy particularly ovarian, lung, pancreatic, stomach and colorectal.[5] The patient showed typical features for diagnosis of dermatomyositis such as progressive symmetrical muscle weakness, elevated muscle enzyme and erythematous rashes all over the body with positive

anti-Mi-2 antibody, which is suggests dermatomyositis. Mi-2 antigen, nuclear helicase protein, forms part of the nucleosome remodeling deacetylase (NuRD) complex involved in transcription regulation.[6] Mi-2 is expressed highly in developing hair follicles and embryonic ectoderm, which plays a critical role in the development of epidermis.[7] Mi-2 is also essential for differentiation and renewal of the basal epidermis.[8] Immune response to Mi-2 protein leads to the development of skin rashes and production of anti-mi-2 antibody.[9] Sunlight plays a significant role in anti-Mi-2 production through subcellular distribution, expression, and metabolism of the components of Mi-2 antigens.[10] Bohan and peter classification was used to confirm the diagnosis.

CONCLUSION

This dermatomyositis patient showed typical clinical manifestation with increased muscle enzymes along with cutaneous manifestations.

Treatment consists of high-dose prednisone, along with steroid-sparing agent and IVIg. It is important to diagnose and start immunosuppressive treatment early in the natural history of the disease in order to reduce the burden of long-term complications.

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