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Erythrodermic Dermatomyositis

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Abstract:

There are protean skin manifestations of dermatomyositis such as Gottron's papules, shawl sign, V sign, and heliotrope rash. In this case report we describe a patient with a rare skin manifestation of dermatomyositis of which there are few reports worldwide.

Key words: Dermatomyositis, Exanthema, Autoimmune Diseases, Muscle weakness, Myositis.

Introduction

Dermatomyositis is an inflammatory myopathy characterised by muscle weakness along with characteristic skin changes. In this case report we describe an unusual skin manifestation of dermatomyositis-erythrodermic dermatomyositis of which there are only a handful of case reports worldwide.

Case Report

A 40 year old lady presented to the emergency with a gradually progressive proximal muscle weakness involving both her upper limbs as well as her lower limbs. She also noticed a generalised redness of her whole body and a butterfly rash on her face.

On examination she had a temperature of 100°F and a malar rash along with erythroderma involving almost her entire body [Fig.1]. The other clinical features which are pathognomonic

Fig.1: Malar rash in erythrodermic dermatomyositis.

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of dermatomyositis such as shawl sign, V sign, heliotrope rash and Gottrons papules were absent. Nail bed capillary dermatoscopy revealed haemorrhage in the nail fold, a feature which is pathogonomic of dermatomyositis. Muscle examination revealed power of 1/5 in proximal muscles of both upper limbs and lower limbs with grade 3/5 power in the distal muscles of the same. Reflexes were preserved.

The differential diagnosis considered were dermatomyositis, paraneoplastic syndrome of proximal myopathy and erythroderma secondary to internal malignancy, necrotising myositis secondary to infection or autoimmune disease, SLE with dermatomyositis overlap syndrome.

CPK (creatine phophokinase) was elevated at 29,746 U/L (normal range: 45-195). EMG/NCV (electromyography and nerve conduction velocity) were suggestive of a myopathic pattern. Muscle biopsy showed presence of myophagocytosis but no endomysial inflammatory infiltrate. Muscle antibodies as tested by immunoblot were negative. Our patient fulfilled 4/5 of the Peter and Bohan criteria for diagnosis of dermatomyositis.

Once the diagnosis was established an extensive search for malignancy was undertaken with a bone marrow examination, CT thorax and abdomen along with both upper and lower GI scopies. These were all negative for malignancy.

Discussion

Dermatomyositis is an inflammatory myopathy with involvement of both the skin and the muscle. In about 20-25% of cases it is associated with internal malignancy [1]. Common skin manifestations include Gottrons papules, heliotrope rash, poikiloderma (small areas of atrophy and telangiectasia with pigment anomalies) and occasionally ulcers and scarring [2]. The mean age at presentation as per Indian data is 35.03 years with a slight female preponderance [3].

The patient described in this report did not have any of the classical skin features of dermatomyositis but fulfilled almost all criteria for diagnosis of the same. This condition is a variant of dermatomyositis called erythrodermic dermatomyosis. There are only about 6 case reports of the same worldwide [4]. Its association with malignancy is not much different from the non-erythrodermic form dermatomyositis.

Our patient was treated with prednisolone at 1 mg/kg along with methotrexate 15 mg once a week following a pulse of IV methylprednisolone for 3 days. Her muscle power improved to 3/5 in all proximal muscles and 5/5 in distal muscles. The erythroderma also decreased considerably.

Conclusion

Erythrodermic dermatomyositis is a rare variant of dermatomymyositis with only a handful of cases described worldwide.

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