GOTTRON'S PAPULES WITH RAISED CPK LEVEL WITHOUT OTHER MANIFESTATIONS OF DERMATOMYOSITIS

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Dermatomyositis without myositis has been reported rarely and has been given several names such as dermatomyositis siné myositis and amyopathic dermatomyositis. We report a 19-year-old male patient with Gottron's papules and raised CPK level. The patient has been followed for 10 months and has not developed other features of myositis.

Key Words : Amyopathic dermatomyositis, Dermatomyositis siné myositis, Gottron's papules

Introduction

Dermatomyositis is a connective tissue disease with inflammation of muscles and skin. When only muscle inflammation is there, the term polymyositis is used. Sometimes in patients the initial presentation is with typical skin rashes only. Most such patients go on to develop muscle disease 3 to 6 months later. But there also exists a very samll subset of patients who despite having the typical rashes for months or years never develop clinical and/or laboratory evidence of myositis and this is referred to as the amyopathic form of dermatomyositis.

Case Report

A 19-year-old male presented to us with asymptomatic, slowly progressive, dull pinkish plaques on the skin overlying the periungual area, interphalangeal joints of all fingers, knuckles and the extensor aspect of both the elbows present for the last 6 months. Small, flat-topped, hypopigmented or pinkish, discrete papules were found overlying these plaques, the changes being more marked on the right hand (Fig. 1). The "candle grease" sign and the Auspitz sign were negative. These

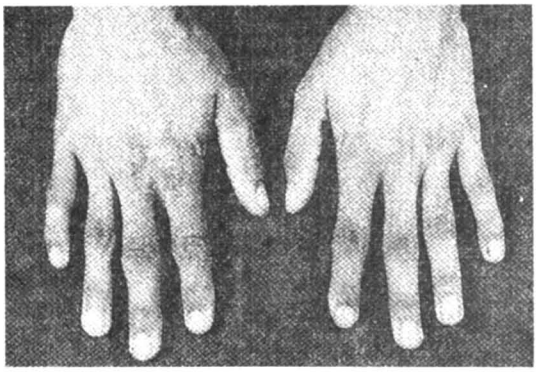

Fig. 1. Gottron's papules.

was no history of muscle weakness or pain, fever, photosensitivity, swelling or any other systemic complaint. The family history was also insignificant.

The blood counts, urea, sugar, electrolytes and creatinine levels were all within the normal limits. The CPK level was raised (312 U/L, normal range 30 to 200 U/L), the LDH, SGOT and SGPT levels were within the normal ranges. The histopathological examination of the biopsy specimen was compatible with dermatomyositis showing hyperkeratosis and focal perivascular lymphohistiocytic infiltrate in the dermis.

Keeping in view the strong clinical resemblance to Gottron's papules and the raised CPK level, it was decided to carry out an EMG of the quadriceps femoris, despite

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there being no muscular symptoms. The EMG was within normal limits and showed no evidence of myositis.

Since the appearance of the skin lesions, the patient has been followed up for 10 months. Though the skin lesions are still persisting despite topical applications of corticosteroid and sunscreen, he hasn't developed any clinical evidence of muscle weakness.

Discussion

There have been reports of typical skin lesions of dermatomyositis occurring without or with minimal evidence of myositis, the cases being labelled variously as dermatomyositis siné myositis, and amyopathic dermatomyositis, etc.[1] However, the original classification of dermatomyositis by Bohan and Peter did not include such an entity.[2] Euwer and Sontheimer have divided amyopathic dermatomyositis into 3 types :[3]

Type I: Pure amyopathic dermatomyositis patients who have only skin disease.

Type II: Patients with skin disease who have subjective myalgias and weakness but not laboratory evidence of muscle diseae.

Type III: Patients with no muscle weakness clinically but who have evidence of abnormal laboratory tests at some time during their course.

Our patient falls in the third type and as 10 months have already elapsed since the onset of skin disease without the developement of muscle weakness, he can in all probability be labelled as a case of dermatomyositis siné myositis. To our knowledge, this is the first such case report from India.

References

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