

CUTANEOUS MANIFESTATIONS OF DERMATOMYOSITIS IN MALE PATIENT: A RARE REPORT

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Source of Support:

Nil

Competing Interests:

None

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Our Dermatol Online. 2013; 4(2): 168-171

Date of submission: 19.10.2012 / acceptance: 19.11.2012

Abstract

Introduction: Dermatomyositis is an idiopathic inflammatory myopathy. It is a systemic disorder that most frequently affects the skin and muscles, but may also affect the joints, the esophagus, the lungs, and, less commonly, the heart.

Case report: It is presented with characteristic cutaneous findings like skin rash as well as progressive symmetrical proximal muscle weakness. Its prevalence rate is approximately one per 100,000 in the general population with a female to male predominance of about 2:1.

Conclusion: We report a male patient with the classical features of dermatomyositis in whom cutaneous changes preceded muscle weakness.

Key words: dermatomyositis; Gottron's sign; heliotrope sign; poikiloderma; shawl sign

Cite this article:

Ganesh Dhavalshankh, Archana Dhavalshankh, Vaishali Masvekar: Cutaneous manifestations of dermatomyositis in male patient: a rare report. *Our Dermatol Online.* 2013; 4(2): 168-171.

Introduction

Dermatomyositis is an idiopathic inflammatory myopathy (IIM) [1,2]. It is a systemic disorder that most frequently affects the skin and muscles, but may also affect the joints, the esophagus, the lungs, and, less commonly, the heart [3,4]. It is presented with characteristic cutaneous findings like skin rash as well as progressive symmetrical proximal muscle weakness. It has extramuscular manifestations such as joint contractures, dysphagia, cardiac disturbance, pulmonary symptoms, and subcutaneous calcifications. DM also has an association with malignant disease, and various autoimmune and connective tissue diseases. The average age at diagnosis is 40 yr, and almost twice as many women are affected as men [3,4]. Its prevalence rate is approximately one per 100,000 in the general population with a female to male predominance of about 2:1. DM is usually associated with an underlying malignancy, and its prevalence is even rarer without coexistent cancer [5,6]. Dermatomyositis is a connective tissue disorder constituting inflammatory myopathy along with characteristic cutaneous markers. The diagnostic criterias for the disease has been defined [7]. The various cutaneous manifestations in dermatomyositis, may precede or follow myositis [8,9]. However dermatomyositis can present without muscle weakness [10].

Objectives

We report a male with the classical features of dermatomyositis in whom cutaneous changes preceded muscle weakness. The presenting cutaneous lesions in dermatomyositis include a heliotrope rash with edema, photosensitivity, Gottron's papules and poikiloderma.

Case Report

A 32 years male patient with previous healthy condition presented to us 2 months back with red colored rash & swelling around both eyes and cheeks. He noticed similar type of rash on both hands, both thighs and both shoulders extending up to mid back. He also complained of severe muscle weakness & pain in both shoulder joints. He noticed difficulty in combing his hair and aggravation of rash when exposed to sun. The initial maculopapular erythematous rash started on dorsal aspect of both hands and then spread to involve face, back and abdomen since 6 months. Since last two months he noticed difficulty in climbing stairs, getting up from squatting position and combing his hair. The muscle weakness was bilateral and gradually progressive without any fasciculations. The clinical examination revealed bilateral periorbital erythematous rash and edema covered with fine white scales suggestive of heliotrope rash (Fig. 1) which is highly suggestive of dermatomyositis.

Similar type of rash was also present on both malar prominences, dorsii of both hands, abdomen and V of neck mostly covering sun-exposed areas suggestive of poikiloderma (Fig. 2). The rash extended on upper part of the back suggestive of shawl sign (Fig. 3). There were hyperpigmentation papules found on bony prominences particularly the metacarpophalangeal and interphalangeal joints suggestive of Gottron papules and Gottron's sign (Fig. 4).

The systemic examination revealed mild pallor. There was weakness of various group of muscles up to grade II. None of the muscles were atrophic but tenderness was present in the muscles of extremities. No fasciculations were seen in any of the muscles. Deep tendon, abdominal and plantar reflexes were normal. The breasts, genitalia and gastrointestinal tract were normal. No periungual erythema and nailfold telangiectasia or cuticular dystrophy observed. No mechanic hand lesions (fissured scaly hyperkeratosis) were found. Laboratory data revealed haemoglobin 13.88 gm/dl, total leukocyte count 5900/mm³, P73L19M4E4B0, platelets 1.9 lacs/mm³ and ESR 16 mm/hr. Urine analysis, blood glucose, blood urea, serum creatinine and serum uric acid were within normal limits. LE cells and antinuclear factors (by indirect immunofluorescence) were not detected (0.66) and anti-dsDNA was negative (19.13

IU/ml). CPK was 195 units/dl (n= 10-70), CPK (MB) 36.78 units/dl (n<5 percent of total), LDH 299 units/dl (n=200-450) and SGOT was 23 units/ml (n=15-45). RA test was negative. Ultrasonography of abdomen including pelvis, and ECG were normal. A skin biopsy showed mild hyperkeratosis, patchy parakeratosis, spongiosis, focal thinning, hydropic degeneration of basal cell layer with lymphocytic exocytosis. The upper dermis showed band-like lymphocytic infiltrate, incontinence of pigment and dilated capillaries (Fig. 5).

The patient received 10mg of oral prednisolone twice daily along with 200mg of hydroxychloroquine twice a day and once weekly 10mg of methotrexate. The therapy was supplemented with oral antioxidants, folic acid and topical diluted mometasone furoate at night and broad spectrum sunscreen during daytime. The muscle weakness was gradually improved over a period of four weeks and the muscle enzymes also reduced. The prednisolone was then gradually tapered to 5mg over a span of four weeks. Presently he is off the steroids and is being maintained on 5mg of methotrexate once a week and same dose of hydroxychloroquine. The patient has regained his muscle power and there is a drastic improvement in his rash.



Figure 1. Heliotrope rash with scales involving periorbital skin



Figure 2. Poikiloderma - Erythematous rash on sun exposed area



Figure 3. Shawl sign - Erythematous rash on upper part of back



Figure 4. Gottron sign - Erythematous and thickened skin at joints

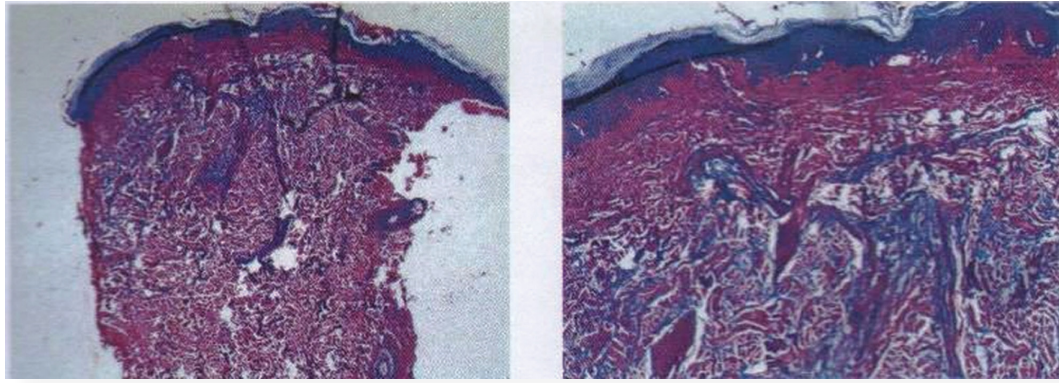


Figure 5. Histopathological changes

Discussion and Conclusion

Bohan and Peter (1975) suggested a set of criteria which aid in diagnosing dermatomyositis. Dermatomyositis and polymyositis is diagnosed in patient with typical cutaneous features, progressive proximal symmetrical muscle weakness, elevated muscle enzyme levels and abnormal findings from muscle biopsy. Patient of dermatomyositis often present with skin diseases as an initial manifestations. In nearly 40% patients skin disease may be the sole manifestation as the onset. This patient is presented and having mostly all characteristic skin manifestations, including Gottron's sign, the shawl sign, the heliotrope rash, poikiloderma and a generalized erythroderma. This patient manifested all classic skin signs with his flare up of DM [1,2,5,7]. Muscle disease may occur concurrently or it may precede the skin disease or it may follow the skin disease by weeks to years (Bohan 1975). Skin rash often precedes the onset of weakness by weeks to months [11]. Early in disease course, rash and muscle enzyme elevations may be the sole manifestations of DM [12]. In our study, patient had proximal muscle weakness since 1 week but the rash preceded muscle weakness and started since 1 year. Rockerbie NR et al [8] reviewed 50 patients of dermatomyositis retrospectively and was found that cutaneous changes sometimes preceded muscle weakness more than a year before the onset of muscle weakness. These findings suggest that the characteristic dermatomyositis eruption without muscle weakness should not preclude a diagnosis of dermatomyositis. This patient presented to us with similar complaints that rash followed by muscle weakness. Our case satisfied all the criteria for the definite diagnosis of dermatomyositis. The various cutaneous lesions, in dermatomyositis, may precede, occur simultaneously, or follow the onset of muscle weakness. Dermatomyositis sine myositis has also been documented [2]. The cutaneous lesions usually precede the onset of weakness by 3 to 6 months, [6] however, Pearson [11] has described one case, with skin involvement for 13 years. It is known that DM has a bimodal age distribution: one peak occurs in children between 5-14 yr of age and a second, larger peak occurs between 45-64 yr of age [13,14]. In our study, onset age was 32 years. It is reported that females outnumber males by 2:1 [13,14]. We reported this in male patient. The association between dermatomyositis (and possibly polymyositis) and cancer has long been recognized [15-19], but no any clinicopathological findings seen

relevant in this case. Though etiology of the disease is unknown, it is believed to be initiated by viral infection and altered immune response. Lymphocyte mediated muscle cell damage and small vessel damage are important central pathogenetic factors. Usually vascular deposits of immune complexes and complement are associated with endothelial cell injury and small vessel obstruction. Dermatomyositis is usually associated with CD4 T cells and B cells infiltrating the muscles whereas polymyositis is associated with CD8 cytotoxic T cells. This case shows similar biopsy changes like lymphocytic infiltrate, incontinence of pigment and dilated capillaries.

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