Late-onset Polymyositis in a Case of Poikilodermatomyositis


Abstract
A case of poikiloderma developed polymyositis ten years after the onset of skin changes. This rare case of poikilodermatomyositis, hitherto not reported from Asian continent, is documented.

INTRODUCTION

Dermatomyositis is a connective tissue disorder constituting inflammatory myopathy along with characteristic cutaneous markers. The diagnostic criteria for the disease has been defined. The various cutaneous manifestations in dermatomyositis, may precede or follow myositis. However dermatomyositis can present without muscle weakness. The presenting cutaneous lesions in dermatomyositis include a violaceous or heliotrope periorbital eruption or edema, periungual telangiectasia, photosensitivity, vasculitis, Gottron’s papules, Gottron’s sign and poikiloderma. The term poikiloderma atrophicans vasculare or poikiloderma in short, is applied to lesions that in the early stage show erythema with slight superficial scaling, a mottled pigmentation and telangiectases. In the late stage the skin appears atrophic and the erythema is less pronounced than in the early stage but the mottled pigmentation and the telangiectases are more evident. Dermatomyositis seen in association with poikilodermatous skin is termed as poikilodermatomyositis.

We report a rare case, in whom poikiloderma was present for ten years before she developed polymyositis. The treatment does not affect these skin changes. Its prognostic significance has not been elucidated. This is the first case report from this part of the world.

CASE REPORT

A 45 years female presented with history of asymptomatic, dry, scaly, hypo and hyperpigmented skin changes (Fig. 1) on sun-exposed (face and back of hands) and sun-protected (trunk and buttocks) areas of 10 years duration. The initial erythematous patches on the trunk and face spread to involve extremities in five years. In the next five years, reddish, tortuous lesions appeared on the trunk. Photosensitivity was present. From last two months, she had noticed difficulty in climbing stairs, getting up from squatting position and combing her hair. The muscular weakness was gradually progressive. There was no history of fasciculations. During this period, she also noticed bluish pigmentation and puffiness around her eyes. The examination revealed mild pallor. There was unequivocal weakness of various groups of muscles. The power of pelvic and pectoral girdle muscles was grade 4, while it was grade 3 in the flexors and extensors of upper and lower limbs. None of the muscles were atrophic or tender. 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. The cutaneous findings included a heliotropic rash around the periorbital area and dry, scaly, atrophy of skin with reticulate pigmentation of the face, neck, shoulders, chest, abdomen, back, upper arms and thighs. Telangiectasia was present over the face, neck, chest, abdomen and periungual region.

Laboratory data revealed haemoglobin 11.2gm/dl,total leukocyte count 7400/mm³, P55L41M3E1, platelets 2 lacs/mm³.

Fig. 1 : Poikilodermatous skin changes.
and ESR 40 mm/hr. Urine analysis, blood glucose, blood urea and serum creatinine were within normal limits. LE cells and antinuclear factors (by indirect immunofluorescence) were not detected. CPK was 195 units/dl (n=10-70), CPK (MB) 20 units/dl (n<5 percent of total), LDH 299 units/dl (n=200-450) and SGOT was 45 karmen units/dl (n=15-45). 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. A muscle biopsy from the vastus lateralis revealed occasional necrotic muscle fibres with collection of a few histiocytes, atrophic fibres, and focal replacement by fibrous and adipose tissue. Some fibres showed internalisation of the nuclei. No significant inflammatory infiltrate was seen in this biopsy and blood vessels were normal. EMG showed small motor unit potentials which were brief in amplitude and duration. Percentage of polyphasia was increased in all the muscles sampled. The findings suggested myopathic pattern.

The patient received 60 mg of oral prednisolone in once morning daily dosage. The muscle weakness gradually improved over a period of four weeks and the muscle enzymes also reduced. The prednisolone was then gradually tapered to 20 mg in eight weeks. It was then changed to 20 mg and 10 mg alternate days schedule and presently being maintained on 10 mg and 5 mg alternate day for last four months. Though the patient had regained her muscle power, her poikilodermatous skin remained unchanged.

**DISCUSSION**

Poikilodermatous skin is a feature of various genodermatosis like poikiloderma congenitale of Rothmund-Thomson, Bloom’s syndrome and dyskeratosis congenita as well as an early stage of mycosis fungoides, dermatomyositis and rarely in association with systemic lupus erythematosus. The poikilodermatous skin of Rothmund-Thomson and Bloom’s syndrome start in early infancy or early childhood, are autosomal recessively inherited and associated with mental and growth retardation and severe photosensitivity. Dyskeratosis congenita is inherited as X-linked recessive disorder that occurs in males only. Mycosis fungoides is a form of T-cell lymphoma and poikiloderma in these patients arises in patches from thin pinkish red plaques in unexposed areas of skin like, trunk and proximal extremities.

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. The cutaneous lesions usually precede the onset of weakness by 3 to 6 months, however, Pearson has described one case, with skin involvement for 10 years. Rockerbie, in his series of 50 cases, has documented maximum duration of skin involvement as four years and three months before the onset of muscle weakness. In this report, five of the 50 cases had poikiloderma but neither the duration nor the type of initial skin changes has been specified. Further, the time of onset of the muscle weakness in these patients is not mentioned. Also, the number of cases of poikiloderma developing polymyositis has not been described. Our patient had poikilodermatous changes for 10 years prior to the onset of muscle weakness. Therefore, except for one confirmed case where muscle weakness developed late (13 years after the skin rash - type not described), our case with late onset myositis had poikilodermatous skin changes for 10 years, before clinical muscle involvement. Besides, this case also confirms the previous reports that the skin involvement usually precedes the muscle weakness in most circumstances, and the poikilodermatous skin slowly evolves from initial erythematous patches to dry, scaly, atrophic and mottled skin. Further, the treatment remains directed towards polymyositis because the poikilodermatous skin remains unchanged. Thus, it is essential to recognise early skin manifestations and suspect dermatomyositis, so as to investigate and institute the therapy early and thus avoid irreversible changes in the form of poikiloderma.

**REFERENCES**