A Case of Amyopathic Dermatomyositis

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We report a 33-year-old woman with a 1-year history of typical skin features of dermatomyositis without any evidence of muscle involvement.

Her skin eruption of the face (heliotrope erythema) and hands (Gottron's papules) and skin biopsy findings were typical of dermatomyositis. Levels of serum muscle enzymes were within normal ranges and electromyography showed no signs of muscle involvement. During a 1-year follow-up, she had no signs of muscle weakness. Based on these clinical and laboratory findings, our case can be diagnosed as amyopathic dermatomyositis.

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Key Words : Amyopathic dermatomyositis, Heliotrope erythema, Gottron's papules

Amyopathic dermatomyositis (ADM) is an uncommon variant of dermatomyositis (DM) defined in 1979 by Pearson[1] to describe those patients who had typical cutaneous findings of DM but did not have any clinical and laboratory signs of muscle disease. In order to make this diagnosis it has been proposed that a patient must have one or more pathognomonic cutaneous features in association with one or more characteristic cutaneous lesions[2]. Furthermore, the possibility of systemic lupus erythematosus, contact dermatitis, drug eruption, and other disorders needs to be excluded. In Korean literature, Lee et al[2] reported a case of ADM, but in their case the follow-up period of 32 weeks was rather insufficient for the diagnosis of ADM. We report a case of ADM with typical cutaneous manifestations such as Gottron's papules, periungal nailfold telangiectasia and violaceous periorbital erythema of the face, lasting for 2 years, which were not accompanied by any sign of muscle involvement.

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CASE REPORT

A 33-year-old woman presented with a 1-year history of progressive skin eruption involving the face, V area and nape of the neck, upper portion of the back, and extensor aspects of forearms, arms, elbows, hands and knees. There was no history of myalgia, arthralgia or Raynaud's phenomenon. Examination revealed periorbital violaceous erythema and edema, erythematous and edematous patches on the face (Fig. 1), violaceous papules over the elbows and proximal interphalangeal and metacarpophalangeal joints (Fig. 2), and poikiloderma atrophicans vasculare-like lesions on the back (Fig. 3). Nailfold changes with periungal telangiectasia were prominent (Fig. 2). She had no signs of muscle weakness on physical examination.

Laboratory investigations including complete blood cell counts, renal function test, urinalysis were within normal limits. Serum levels of creatinine phophokinase, lactic dehydrogenase, and aldolase were within the normal range. Antinuclear antibody was positive (1:40, speckled pattern), whereas all other immunological parameters (anti-DNA, anti-Ro, anti-La, anti-Sm, anti-RNP, lupus anti-

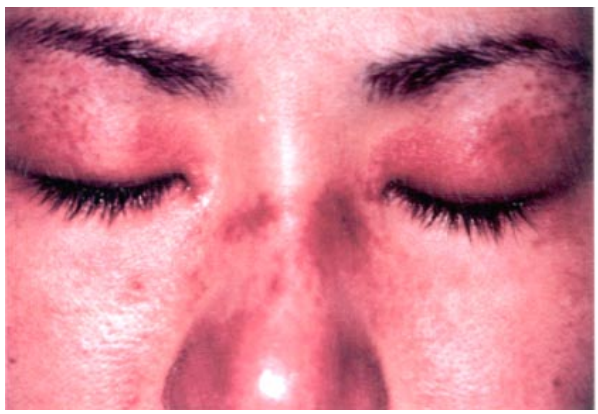

Fig. 1. Periorbital violaceous erythema and edema on the face.

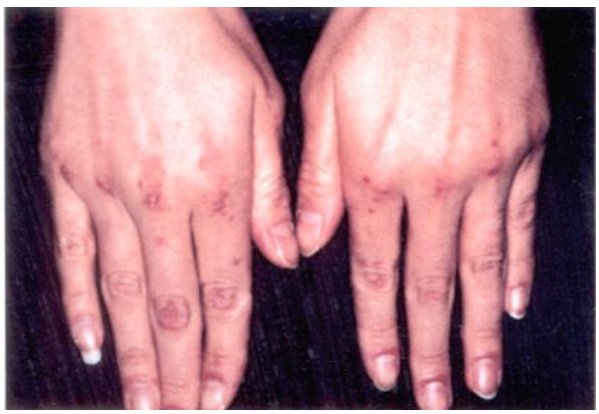

Fig. 2. Typical Gottron's papules and periungual nail-fold telangiectasia.

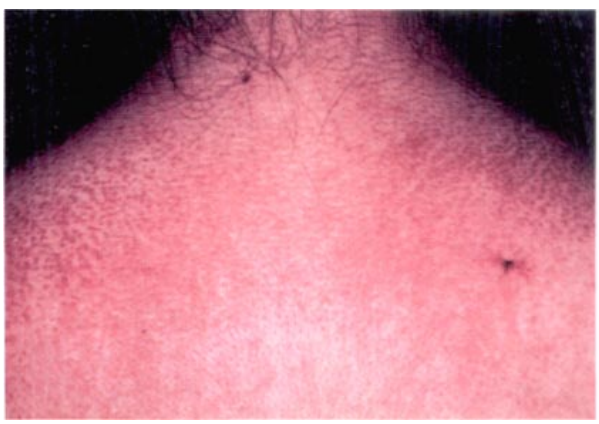

Fig. 3. Poikiloderma atrophicans vasculare-like skin lesions on the back.

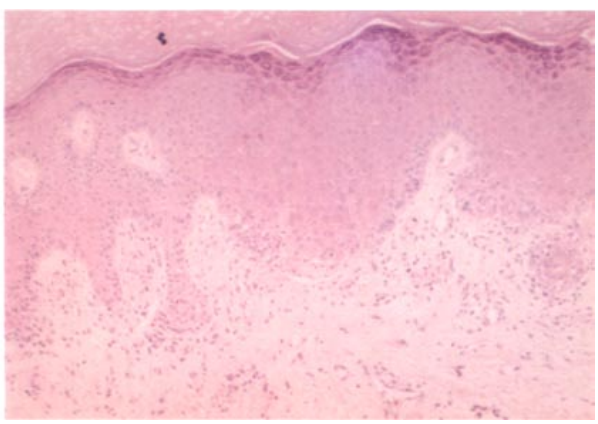

Fig. 4. A superficial perivascular lymphocytic infiltrate with focal vacuolar change involving the keratinocytes along the dermoepidermal junction. There is a slightly compact orthokeratosis (H & E, ×100).

coagulant, anti-cardiolipin) were negative.

Electromyography of proximal muscle groups did not reveal a myositis of the arms and legs.

Histopathologic examination of biopsy specimens obtained from two papular lesions on the dorsum of the hand and the elbow revealed hyperkeratosis, exocytosis, and focal liquefactive degeneration of the basal layer in the epidermis, and perivascular mononuclear cell infiltration in the upper dermis (Fig. 4). Direct immunofluorescence revealed slight deposits of fibrinogen at the dermoepidermal junction, but showed no deposits of immunoglobulins or complements.

She was treated with an application of a topical steroid ointment. The skin lesions were slightly improved with this treatment, but recurred after withdrawl. During a 1-year follow-up period, there was no evidence of muscle weakness.

DISCUSSION

ADM was originally defined as the presence of biopsy confirmed classic cutaneous findings of DM in the absence of any clinical or laboratory signs of muscle disease for at least 2 years after onset of skin pathology[1]. Euwer and Sontheimer[3] reported that the prevalence of ADM during a 15-year period at their institution was 11% (6/54). They suggested that the diagnosis of ADM need not be limited only to patients with a skin disease for 2 years. Rather, any patient with only skin disease or with minimal muscle disease at presentation would warrant this diagnosis. Recently, Caproni et al[4] defined ADM as an abortive and/or transient form of DM, which is, however, important for the

low incidence of malignancies and for its slow and long benign course.

The cutaneous manifestations of DM are classified as pathognomonic, characteristic, and compatible[3]. Gottron's papules and sign are considered as pathognomonic. Periorbital violaceous heliotrope erythema, periungal telangiectasia and symmetrical macular violaceous erythema of the face, shoulder area and upper chest are characteristic. Compatible skin lesions are poikiloderma atrophicans vasculare, and subepidermal bullous lesions and superficial erosions. To diagnose ADM, there needs to be one or two pathognomonic cutaneous features in association with one or more characteristic cutaneous features[3]. Our patient had one pathognomonic sign (Gottron's papules), two characteristic signs (heliotrope erythema and periungal telangiectasia), and one compatible sign (poikiloderma atrophicans vasculare), which were consistent with ADM.

The cutaneous histopathologic features of DM are not pathognomonic. However, there are certain histopathologic features that favor the diagnosis of DM, including hyperkeratosis, lymphocytic perivascular infiltrates with vacuolar interface changes and thickening of the walls of cutaneous blood vessels[3]. Immunofluorescent findings are also not specific and not helpful in making the diagnosis of DM but may help rule out lupus erythematosus[3]. In our case, we also found these findings in the biopsy specimens.

Establishing the diagnosis of ADM requires rigorous evaluation to exclude muscle involvement. For this purpose, diagnostic methods such as electromyography and muscle biopsy are available. However, the opinions on the need of extended investigations such as electromyography and muscle biopsy for establishing the diagnosis of ADM have remained controversial. Some authors emphasize that electromyography and muscle biopsy are less sensitive than muscle enzyme studies in detecting active myositis[5]. The major disadvantage of muscle biopsy is its invasive character. Currently, newer imaging techniques such as magnetic resonance imaging and muscle ultrasound have been proposed for this purpose[6-8]. In this case, there was no clinical or laboratory signs of muscle disease, although muscle biopsy was not performed.

The mainstay of therapy in DM is systemic cortocosteroids. Other agents are azathioprine, methotrexate, and cyclophosphamide, which all carry a substantial risk of side effects[3,9,10]. The skin is notoriously unresponsive to many of these treatment modalities. In some cases, however, hydroxychloroquine has been claimed to be successful[11,12]. It is as yet undetermined, whether early aggressive immunosuppressive treatment of ADM might prevent the development of myositis at a later date or influence the course of the skin disease[13]. Some authors[3] suggested that a more aggressive treatment might prevent the development of muscle disease in patients who initially had only skin involvement. In contrast, Others[6,9,14] suggested withholding systemic corticosteroid or other immunosuppressive therapy in the absence of muscle disease. Cosnes et al[15] also documented that unless frank weakness develops, systemic corticosteroid treatment can be avoided. From our experience, we recommand a more expectant attitude with careful and regular monitoring for possible development of muscle disease.

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