A Case of Wong-type Dermatomyositis with Anti-transcriptional Intermediary Factor1-Gamma Autoantibody

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ABSTRACT:

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Dermatomyositis (DM) belongs to the spectrum of the idiopathic inflammatory myopathies characterized by muscle weakness and skin rashes. Wong-type dermatomyositis is one of rare variants of DM.

To the best of our knowledge, there are a few reported cases of Wong-type dermatomyositis. We herein report a case of DM presented with atypical cutaneous manifestations typified by hyperkeratotic follicular papules. Histologic features were similar to those of pityriasis rubra pilaris. Treatment with systemic prednisolone and hydroxychloroquine led to well control of cutaneous and muscle symptoms.

Key words: Dermatomyositis, pityriasis rubra pilaris, wong-type dermatomyositis

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Introduction

The diagnosis of dermatomyositis (DM) relies on cutaneous findings including pathognomonic, characteristic or compatible skin signs, muscle weakness and evidence of inflammatory myopathy. The onset of DM is bimodal, being juvenile and adult form[1]. We herein report a case of DM presented with atypical cutaneous manifestations.

Case report

A 48-year-old woman presented with erythematous rashes at the upper extremities for 1 year. One month prior to this visit, the lesions progressed and spread to the scalp, eyelids, chest wall, trunk and knees. She noticed that her limbs were weak without numbness. On examination, ill-defined, erythematous to violaceous and edematous patches at periorbital areas were seen. Ill-defined scaly erythematous follicular papules, patches, and plaques at the scalp, retroauricular areas, upper chest, torso, upper extremities, metacarpophalangeal joints, and along tendons were also detected (Figure 1). Oral lesions were not observed. Nail fold dermoscopic examination showed dilated capillary loops and capillary dropout. On neurological examination, there was a decrease in the motor power of all proximal limbs. Skin biopsy specimen taken from the affected area showed mild papillomatosis of the epidermis with subtle areas of checker-board orthokeratosis and parakeratosis. The basement membrane was thicken and there was also vacuolar alteration of the dermal-epidermal junction and pigmentary incontinence (Figure 2). Alcian blue stain revealed mucin deposition in the dermis. Direct immunofluorescence showed no deposition of immunoreactants. Laboratory tests showed muscle enzyme elevation (CPK 472 U/L), antinuclear antibody positivity (1:160, homogenous and fine speckled pattern) and elevation of anti-transcriptional intermediary factor 1-gamma (TIF1-γ). Occult neoplasia was excluded by appropriate investigations (mammogram, chest -abdominal computed tomography). The pelvic examination and pap smear were performed. The ear, nose and throat examination revealed normal. She was treated with oral prednisolone (60 mg/day) and hydroxychloroquine (200 mg/day). The lesions disappeared completely after 3 months of the treatment leaving post-inflammatory hyperpigmentation and the serum creatine kinase level decreased to 197 U/L with full recovery of muscle strength. Then prednisolone was tapered off and methotrexate (7.5 mg/week) was added. The patient had no recurrence of rashes or weakness after discontinuing corticosteroid.

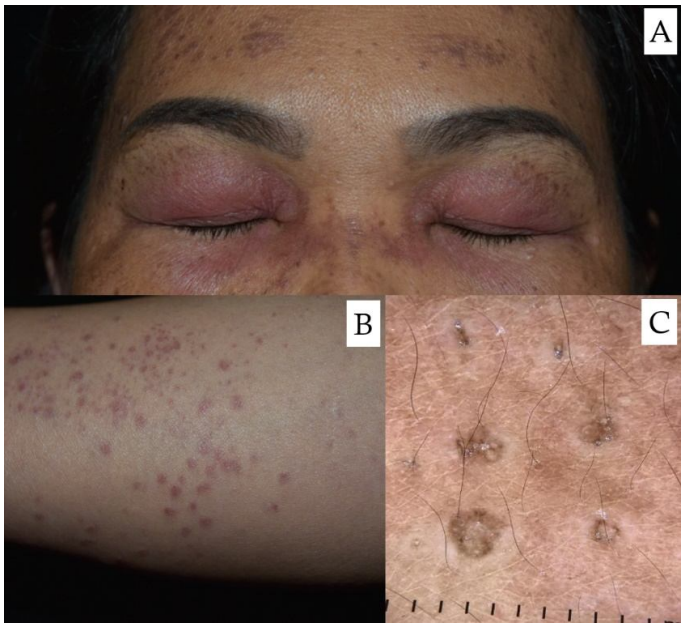

Figure 1 A) Ill-defined, erythematous to violaceous and edematous patches at periorbital areas
B) Ill-defined scaly erythematous follicular papules, patches, and plaques at the upper extremities, metacarpophalangeal joints, and along tendons
C) Dermoscopic examination showed scaly follicular papules

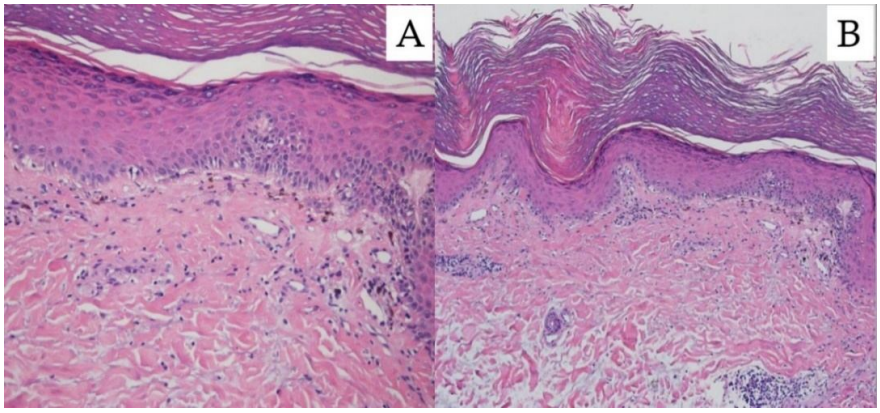

Figure 2 Hematoxylin and eosin stain showed vacuolar alteration of the dermal-epidermal junction and pigmentary incontinence, mild papillomatosis of the epidermis with subtle areas of checker-board orthokeratosis and parakeratosis (X200 [A] and X100 [B])

Discussion

Wong–type dermatomyositis is a rare subgroup of DM, characterized by the mixture of clinical features of pityriasis rubra pilaris (PRP) and DM[2]. Our patients presented with hyperkeratotic papules, follicular hyperkeratosis and palmoplantar keratoderma. The lesions were similar to those of PRP[2]. Wong–type DM was described originally by O'Leary in 1953 and were found in 11 patients in Wong's study[3]. The histopathological findings of Wong–type DM reveal follicular and non-follicular hyperkeratosis as well as arrector pilorum myositis. Basal vacuolization and mucin deposition resemble to those of typical DM may be present. Moreover, alternating parakeratosis and orthokeratosis, which is typically found in PRP, can be seen in Wong-type DM as well with a less frequency[2,3].

The association of DM with malignancy is well known. However, the relation between Wong-type DM and malignancy are not well established[4,5]. These patients have good responses to glucocorticoid therapies. Nevertheless, some reports show the relapse of skin lesions during the immunosuppressive drugs tapering[5].

Interestingly, our patient has anti-TIF1-γ positivity which is one of myositis-specific autoantibodies. Anti-TIF1-γ is found more often in Caucasian than Asian and in patients with clinically amyopathic dermatomyositis (CADM) subtype[6-8]. The term CADM is used when there is cutaneous involvement without evidence of weakness by history or clinical examination. However, CADM can be further classified into 2 subgroups. First, hypomyopathic DM, when there are at least one of the abnormal finding; laboratory, biopsy or imaging. Second, amyopathic DM, when those tests are normal. The presence of anti- TIF1-γ autoantibodies is associated with an increasing risk of malignancy[9]. Some reports show a lower risk for development of interstitial lung disease, Raynaud phenomenon and arthritis[8-9]. Severe cutaneous manifestations are commonly observed. Characteristic skin findings include psoriasiform lesions, hyperkeratotic palmar papules, hypopigmented patches mixed with telangiectatic macules or patches ("red on white") and palatal erythema (ovoid patch)[8]. Our patient did not displayed any of anti-TIF1-γ cutaneous features.

In summary, we herein present a case of Wong-type DM with hyperkeratotic papules and follicular keratosis. Although the relation between this DM subtype and malignancy is unclear; medical histories, complete physical examination and cancer-screening are mandatory, especially in patients with a positive test of anti-TIF1-γ.

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