Wong-Type Dermatomyositis: A Report of Two Cases

Dear Editor,

Wong-type dermatomyositis (DM), a variant of DM, is characterized by the presence of pityriasis rubra pilaris (PRP)-like cutaneous lesions in addition to findings of DM.[1] Herein, we describe two patients of this rare variant, who had varying systemic involvement.

An 18-year-old girl presented with an itchy red rash on her face, trunk, and limbs, along with fever and malaise for 1 month. She had proximal muscle weakness in all four limbs for 2 weeks. She had progressive dyspnea (modified Medical Research Council (mMRC) grade I to IV), and for a week, she had difficulty in swallowing solid food. Examination revealed palmoplantar keratoderma, erythematous scaly rash on the face prominently involving the upper eyelids, linear telangiectasia over eyelids, erythematous scaly plaques over the trunk, and extensor aspects of limbs suggestive of shawl sign, and Gottron's sign, respectively [Figure 1a-c]. The rash on the trunk and limbs was notable for its folliculocentric and keratotic nature. She also had periungual erythema. She had proximal muscle weakness in all the limbs (power 2/5). Due to respiratory distress and pooling of secretions, she was electively intubated and mechanically ventilated. The baseline investigations showed elevated liver transaminases [alanine aminotransaminase (ALT) - 160 U/L; aspartate aminotransferase (AST) - 121 U/L], serum lactate dehydrogenase (LDH-400 IU/L), N-acetyl-cysteine-activated creatinine kinase CK-NAC (4200 IU/L), and positive antinuclear antibody (ANA) in a nucleolar pattern. High-resolution computed tomography (HRCT) of the chest revealed nonspecific interstitial pneumonitis (NSIP) pattern. She was treated with intravenous methylprednisolone and cyclophosphamide pulse therapy followed by oral cyclophosphamide, and hydroxychloroquine. Her symptoms resolved after 20 days and she was extubated. She was discharged with oral medications and is doing well on follow-up.

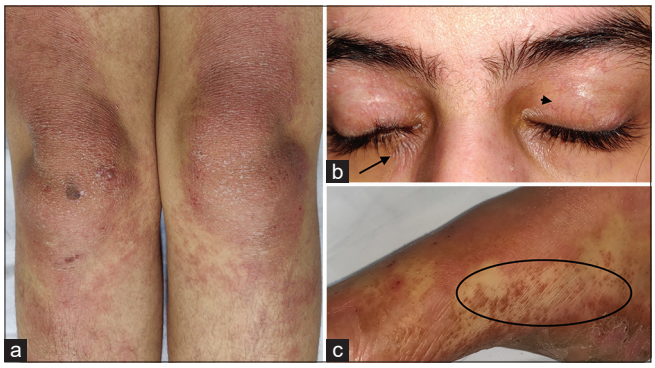

Figure 1: (a) Confluent erythematous follicular keratotic papules on extensors of lower limbs, in a distribution suggestive of Gottron's sign. (b) Erythematous to violaceous scaly rash involving the upper eyelids and peri-orbital region (Long black arrow). Telangiectasias are visible on the upper eyelid (short arrow). (c) Discrete follicular keratotic papules (black oval) are noted at the periphery of plaques on the dorsum of the foot. Keratoderma of soles, with lesional and perilesional erythema.

The second patient, a 25-year-old man presented with fever, photosensitivity, and a generalized rash of 2 months duration. He had been treated previously for dermatomyositis with oral prednisolone, methotrexate, and hydroxychloroquine but was non-compliant. Examination revealed an erythematous scaly follicular rash involving his face, trunk, and limbs [Figure 2a and b]. Gottron's papules, Gottron's sign, nail fold erythema, and ragged cuticles were also present [Figure 2c]. Palmoplantar involvement was absent. Investigations revealed elevated CK-NAC (736 IU/L), LDH (364 IU/L), and a positive anti-mi2 antibody. HRCT of the chest and serum for ANA was unremarkable. He was treated with oral prednisolone, hydroxychloroquine, and azathioprine with improvement in skin rash and myositis. Both patients had histopathological findings of follicular hyperkeratosis and parakeratosis [Figure 3a and b]. Screening for common malignancies were negative for both patients.

Wong's DM is an uncommon presentation of DM and is defined by the presence of follicular cutaneous lesions closely resembling the PRP lesions in a patient with DM. The most constant histopathological feature reported is follicular hyperkeratosis, with keratotic plugs filling dilated follicular infundibula as seen in the index cases.[1]

Apart from keratotic follicular papules of varying extents, the occurrence of diffuse erythema (as seen in patient 2) or spinulosis and palmoplantar keratoderma (Patient 1) is also described in Wong's DM. Cutaneous lesions in Wong-type DM may occur simultaneously (Patient 1),

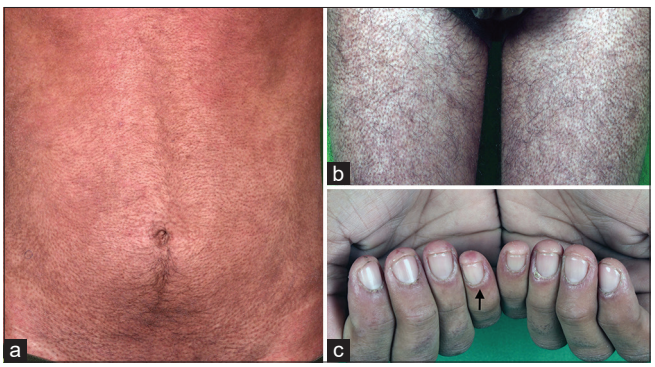

Figure 2: (a) Erythematous follicular keratotic papules diffusely present on the trunk and (b) thighs. (c) Periungual erythema (black arrow), nail fold hemorrhages, and ragged cuticles were noted on both hands

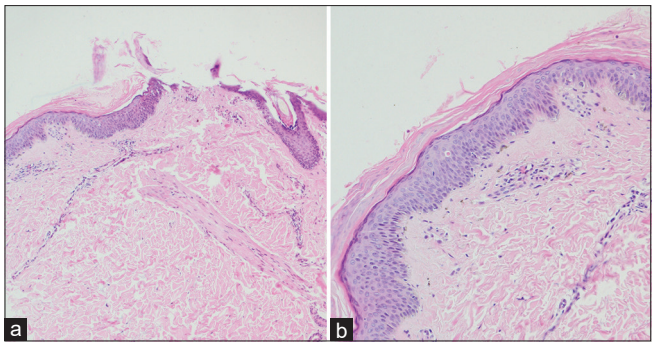

Figure 3: (a) Hyperkeratosis and follicular plugging (Hematoxylin and Eosin ×100); (b) alternative hyperkeratosis and parakeratosis, focal basal cell vacuolar degeneration, and presence of apoptotic keratinocyte (Hematoxylin and Eosin ×200)

before or after myositis (Patient 2) as in classic DM.[2] Wong-type DM, like typical DM, may have associated myositis and interstitial lung disease (ILD), however, with varying severity.[3] Patient 1 had life-threatening myositis with NSIP pattern ILD, while patient 2 had limited myositis without ILD. Some authors suggest the need for prolonged monitoring of patients with PRP, to exclude the development of myopathy, which has also been seen to develop after a lag period of 1 year of developing cutaneous lesions.[4]

The relationship between Wong's DM and malignancy is not well established. In Wong's series, whether the reported neoplasms were associated with classic DM or Wong's variant was not specified.[1] Though the age range of cases reported varied (9–66; mean 36 years), the involvement of the predominantly younger age group explained a lesser association with malignancy.

Treatment of Wong's DM depends on the severity of cutaneous and systemic involvement. Systemic steroids, hydroxychloroquine, acitretin, azathioprine, mycophenolate mofetil, cyclosporine, and rituximab have been used with variable responses.[5]

In conclusion, Wong's DM should be ruled out in all patients with PRP-like skin lesions. This has clinical relevance as treatment for PRP is with oral retinoids, and Wong-DM responds to systemic immunosuppressive agents.

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Conflicts of interest

There are no conflicts of interest.

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