



## CASE REPORT

## Overlap syndrome of type Wong variant dermatomyositis and rheumatoid arthritis

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## ABSTRACT

Wong type dermatomyositis describes a small group of patients with dermatomyositis clinically presented with pityriasis rubra pilaris-like eruptions, including diffuse palmoplantar hyperkeratosis and follicular hyperkeratosis. Histopathologic findings include follicular hyperkeratosis and arrector pili myositis. There have been 20 cases reported since Wong's large series. We report a 56-year-old male with overlap syndrome of Wong type dermatomyositis and rheumatoid arthritis. He also fulfilled the criteria of anti-tRNA synthetase syndrome due to the presence of anti-Jo-1 antibody and clinical features of both rheumatoid arthritis and myositis. No internal malignancy was found after a complete systemic survey and within 1 year follow-up period. There was a good response to oral steroid therapy.

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## Introduction

Dermatomyositis is an idiopathic inflammatory myopathy with cutaneous manifestations. Characteristic findings include Gottron's papules, heliotrope rash, shawl sign on the nape, shoulders, and upper back, and a "V sign" on the V-shaped region of the neck and upper chest. Other cutaneous findings include nail changes, vasculitis, localized lipoatrophy, panniculitis, bullous lesions, and, rarely, hyperkeratotic skin eruptions.<sup>1</sup> Pityriasis rubra pilaris (PRP) describes a group of uncommon, idiopathic, chronic papulosquamous conditions that are characterized by orange-red scaling plaques with follicular plugging, perifollicular erythema, and palmoplantar hyperkeratosis. A PRP-like eruption has been described in patients with dermatomyositis.<sup>2</sup> We report a 56-year-old man with dermatomyositis and rheumatoid arthritis who developed skin eruptions clinically resembling PRP with histologic features of both dermatomyositis and PRP.

## Case report

A 56-year-old male patient was referred from the rheumatology clinic with symptoms of pain in multiple joints, morning stiffness, muscle weakness, and skin rash for several months. The abduction, adduction, flexion, and extension of the shoulders and the hips of the patient were tested, which showed muscle power of 4 for the shoulders and 3–4 for the hips bilaterally, according to the Medical Research Council scale. He had difficulty raising himself from the bed, climbing stairs, and was unable to hold heavy objects as usual. These symptoms were followed by the skin rash 6 months later. Examination revealed diffuse hyperkeratosis on the bilateral soles and palms with orange hue (Figure 1A and B), erythematous scaly patches on the shoulders, back, and knees, puffy fingers, erythematous patches over the metacarpal and proximal interphalangeal joints (Figure 1C), and follicular papules over the bilateral knee joints (Figure 1D). This patient did not have other cutaneous features characteristic of dermatomyositis, such as heliotrope rash, shawl sign, V sign, or periungual telangiectasia. Blood tests showed elevated: antinuclear antibody titer 1:640 (normal range: <1:40); anti-SSA: 509 AU/mL (normal range: <100 AU/mL); anti-SSB: 433 AU/mL (normal range: <100 AU/mL); rheumatoid factor: 436.0 IU/mL (normal range: <20.0 IU/mL); anti-Jo-1: 147 AU/mL (normal range: <100 AU/mL); serum creatine kinase: 606 IU/L (normal range: 38–397 IU/L); aspartate transaminase: 42 IU/L (normal range: 15–41 IU/L), erythrocyte

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**Figure 1** Diffuse hyperkeratosis with orange hue on (A) bilateral soles and (B) palms with sharp margins. (C) Scaly reddish patches on dorsum of fingers. (D) Follicular hyperkeratotic papules (arrow) on the knee.

sedimentation rate: 20 mm/hour (normal range: <12); and lactic dehydrogenase: 307 IU/L (normal range: 98–192 IU/L). Anticyclic citrullinated peptides and C-reactive protein were within normal limits, while HLA-B27 was negative.

A skin biopsy obtained from the palm revealed an acanthotic epidermis overlying with loosely arranged hyperkeratosis (Figure 2A). There were scattered dyskeratotic corneocytes and parakeratosis in horizontal and vertical arrays (Figure 2B). Vacuolar degeneration in the basal cell layer, lymphocyte exocytosis, and scattered dyskeratotic cells were noted, indicating interface dermatitis (Figure 2C). There were also focal cornoid lamellas demonstrated by the loss of the granular layer with a dell in the epidermis containing a stacked column of horny cells, some retaining their nuclei (Figure 2D). A mild to moderate lymphocytic perivascular infiltrate was present in the upper dermis. The overall pathological features presented an unusual combination of interface dermatitis, PRP, and porokeratosis.

A muscle biopsy specimen taken from the left thigh revealed perivascular lymphocytic infiltration and degenerative changes in the muscle tissue. The needle electromyography showed spontaneous activities and low amplitudes, with short duration and polyphasic motor unit action potentials in the right deltoid, vastus medialis, and tibialis anterior muscles, suggestive of myopathic change. A bone scan revealed arthritis involving the bilateral shoulder and wrist joints, carpi, metacarpal, and proximal interphalangeal joints of both hands.

Proximal muscle weakness, increased serum muscle enzyme, and typical findings on electromyography and muscle biopsy results in our patient were compatible with dermatomyositis. The unusual clinical and pathological presentations of PRP-like features indicated Wong type dermatomyositis. Furthermore, according to

2010 rheumatoid arthritis (RA) classification criteria,<sup>2</sup> the patient had a score of 9/10 (cut off point for definite RA: 6/10), compatible with RA. The patient was diagnosed as having overlap syndrome of dermatomyositis and RA. Due to the positive anti-Jo-1 antibody and clinical features of rheumatoid arthritis and myositis, our patient also fulfilled the criteria of anti-tRNA synthetase syndrome, a well-known overlap condition.

A complete malignancy workup was performed. The chest radiograph showed only increased bilateral lung markings; the upper gastroendoscopy revealed shallow ulcers in the lower-body of the stomach. There was positive occult blood, and therefore the patient had a colonoscopy examination and transrectal polypectomy. A tubular adenoma at the hepatic flexure, and a mixed tubular adenoma and hyperplastic polyp were removed. No carcinoma was found. Nasopharyngoscopy and serum tumor markers including carcinoembryonic antigen,  $\alpha$ -fetoprotein, CA19-9, prostate-specific antigen, Epstein–Barr virus-IgA and Epstein–Barr virus early antigen, and nuclear antigen IgA were normal.

He was treated with: daily oral prednisolone 30 mg, plaquenil 200 mg, and etoricoxib 60 mg; topical salicylic acid 2.5% ointment twice daily for the palmoplantar lesions and desoximetasone 0.25% ointment twice daily for the body. The skin lesions gradually improved, so the dosage of oral prednisolone was tapered to 10 mg/day. He did not report muscle weakness after the prednisolone being tapered. The laboratory data, including aspartate aminotransferase, lactate dehydrogenase, erythrocyte sedimentation rate, and serum creatine kinase, decreased following the treatment. Serum creatine kinase decreased from 606 IU/L to 66 IU/L. He is now being maintained on this regimen and regularly followed up at the rheumatology and dermatology clinics.

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