



INFLAMMATORY SKIN DISEASES (OTHER THAN ATOPIC DERMATITIS & PSORIASIS)

## TREATMENT OF PITYRIASIS RUBRA PILARIS WITH USTEKINUMAB: A CASE REPORT

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**Background:** Pityriasis rubra pilaris (PRP) is a rare, chronic, inflammatory skin disease characterized by follicular hyperkeratotic papules of unknown etiology. It may show islands of spared skin, palmoplantar keratoderma and nail dystrophy and may progress to generalized erythroderma. Recently, patients refractory to conventional therapies have been treated successfully with biologic drugs. Recent studies also revealed a role of the interleukin-23/T-helper 17 axis in PRP pathogenesis. Ustekinumab is a anti-IL12/23 IgG1 human monoclonal antibody that is approved for the treatment of psoriasis, but has been shown to be effective as an off-label treatment for PRP. Here we report a case of type 1 PRP that was unresponsive to conventional treatments, but demonstrated complete resolution with use of ustekinumab.

**Observation:** A 52-year-old woman presented with 4-months old, diffuse, pruritic, salmon-colored rash on her trunk, extremities and face. She did not have any other medical comorbidity. Cutaneous examination revealed sparse 'islands' of spared skin within the erythematous areas. Her palms and soles were noted to be hyperkeratotic with associated mild nail dystrophy. Skin biopsy was consistent with PRP. Clinical and laboratory testing ruled out any underlying immunodeficiency and malignancy. The patient was subsequently diagnosed with PRP type 1. She was started on cyclosporine A (3 mg/kg/day) and topical glucocorticoids. The patient's follicular papules persisted over the following 2 months. The treatment regime was changed to acitretin 0,5 mg/kg/day and methotrexate (MTX) 15 mg/week. In the following 3 months her lesions persisted and deterioration in her liver function was observed. MTX was discontinued and acitretin dose was reduced. Off-label ustekinumab treatment was initiated. She was administered with ustekinumab 45 mg injection subcutaneously at weeks 0 and 4, then quarterly. After 6 months of treatment, her lesions disappeared completely.

**Key message:** Ustekinumab appears to be a safe and effective treatment in cases of unremitting PRP.

