Woronoff Ring in Deficiency of Interleukin-36 Receptor Antagonist (DITRA)

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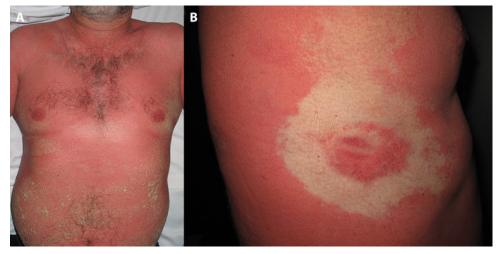
Case Presentation

A 54-year-old-man diagnosed with a deficiency of interleukin-36 receptor antagonist (DITRA) under anti-interleukin-1 therapy with anakinra (described elsewhere [1]) presented with an erythrodermic pustular psoriasis 5 months after suspension of his biological treatment (Figure 1A). Anakinra and topical corticoids were then initiated. Three days later, a halo of nonreddened skin circumscribing an erythematous pustular plaque was observed on his right side (Figure 1B). The patient showed a complete clinical response in the following weeks.

Teaching Point

DITRA is an autoinflammatory disorder caused by mutations in the *ILRN36* gene, characterized by abrupt-onset episodes of generalized pustular psoriasis, fever, and systemic

Figure 1. Woronoff ring in deficiency of interleukin-36 receptor antagonist (DITRA). (A) Widespread erythematous and pustular rash on the trunk. (B) Halo of nonreddened skin circumscribing an erythematous pustular plaque on the right side (3 days after initiating anakinra and topical corticosteroids).



involvement. The Woronoff ring is an underrecognized sign of psoriasis, a halo of nonerythematous skin surrounding psoriatic plaques. Its pathogenesis is not fully understood; a relative deficiency of prostaglandins and endoglin has been reported. The Woronoff ring can occur after topical corticoid or tar treatment, fumaric acid esters, phototherapy, biological therapy [2], or even in untreated psoriasis.

References

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