

Pityriasis Rubra Pilaris and Severe Hypereosinophilia



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PRACTICE POINTS

- Pityriasis rubra pilaris (PRP) can clinically mimic psoriasis. Look for islands of sparing and palmar and plantar hyperkeratosis to help diagnose PRP. A biopsy may be useful to help with this differentiation.
- Pityriasis rubra pilaris may be associated with eosinophilia, but one should rule out other causes of eosinophilia first.

To the Editor:

A 63-year-old man presented with a prior diagnosis of severe psoriasis affecting the extremities, neck, face, and scalp of 1 year's duration. He reported pain, itching, and swelling in the affected areas. He felt the rash was worst on the hands and feet, and pain made performing activities of daily living difficult. His treatment regimen at presentation included triamcinolone cream 0.1% and azathioprine 150 mg daily as prescribed by an outside dermatologist without any response. Physical examination revealed diffuse erythema with lichenification and thick, white, flaking scale on the arms and legs (Figure 1A), face, neck, palms, and soles with islands of sparing. Multiple salmon-colored, follicular-based papules topped with central hyperkeratosis were scattered on these same areas. The palms and soles had severe confluent keratoderma (Figure 2A). Histologic examination of a follicular-based papule showed foci of parakeratosis and hypergranulosis consistent with the patient's clinical picture of pityriasis rubra pilaris (PRP).

Baseline laboratory tests at the time of PRP diagnosis revealed 20.8% eosinophils (reference range, 0%–7%) and an absolute eosinophil count of

$2.17 \times 10^9/L$ (reference range, $0-0.7 \times 10^9/L$). Laboratory test results from an outside dermatologist conducted 10 to 12 months prior to the current presentation showed 12% eosinophils with a white blood cell count of $8.9 \times 10^9/L$ (reference range, $4.5-11.0 \times 10^9/L$) around the time of rash onset and before treatment with azathioprine, making a drug reaction an unlikely cause of the eosinophilia.

After consulting with the hematology department, a hypereosinophilia workup including erythrocyte sedimentation rate, lactate dehydrogenase, serum protein electrophoresis, urine protein electrophoresis, tryptase, double-stranded DNA antibody, human T-lymphotropic

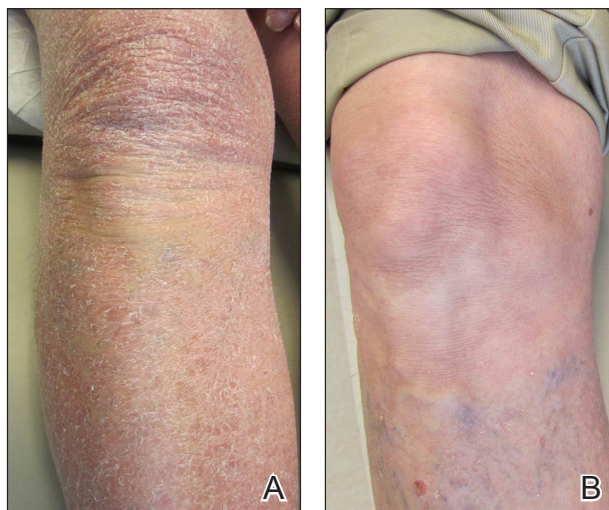


FIGURE 1. Pityriasis rubra pilaris on the leg with salmon-colored, follicular-based papules topped with central hyperkeratosis and islands of sparing below the knee before (A) and after 3.5 months of isotretinoin showing resolution of papules and hyperkeratosis (B).

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The authors report no conflict of interest.

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FIGURE 2. Pityriasis rubra pilaris with bilateral severe confluent plantar keratoderma before (A) and after 3.5 months of isotretinoin showing resolution of plantar keratoderma (B).

virus I/II, stool ova, and parasites, as well as a *Strongyloides* antibody titer, were performed; all were within reference range. His antinuclear antibody level was mildly elevated at 1:160, but the patient had no clinical manifestations of lupus. Given this negative workup, the most likely explanation for the hypereosinophilia was a reactive process secondary to the extreme inflammatory state.

The patient was started on isotretinoin 40 mg daily in addition to urea cream 40% mixed with clobetasol ointment at least once daily to the extremities. Hydrocortisone ointment 2.5% and petrolatum-based ointment were applied to the face, and hydroxyzine was used as needed for pruritus. One month after initiating isotretinoin, erythema had decreased and a repeat complete blood cell count with differential showed a decrease of eosinophils to 14.7% and an absolute eosinophil count of $1.56 \times 10^9/L$. After 2 months of therapy, the patient showed remarkable improvement. After 3.5 months of

therapy, the keratoderma on the palms and soles was almost completely resolved, the follicular-based papules disappeared, and the patient had no areas of lichenification (Figures 1B and 2B). After 5 months of therapy, the patient experienced resolution of the PRP, except for residual facial erythema. His eosinophil count continued to trend downward during these 5 months, reaching 7.6% with an absolute eosinophil count of $0.93 \times 10^9/L$. Three years after the initial onset of the rash and 2 years after completing isotretinoin, his eosinophil level was normal at 5.3% with an absolute eosinophil count of $0.7 \times 10^9/L$.

We present a case of PRP and severe eosinophilia. We initially considered a second disease process to explain the extremely elevated eosinophil count; however, a negative eosinophilia workup and simultaneous resolution of these problems suggest that the eosinophilia was related to the severity of the PRP.