Recalcitrant Solitary Erythematous Scaly Patch on the Foot

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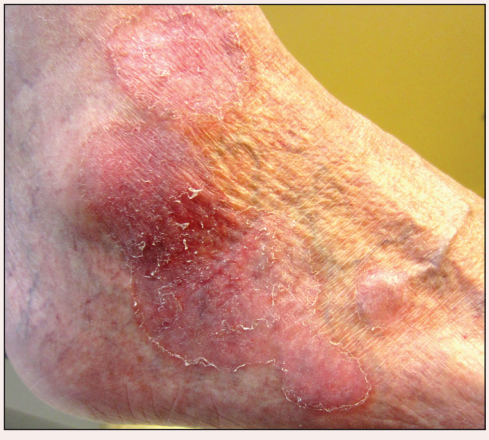

An 80-year-old man with a history of malignant melanoma and squamous cell carcinoma presented to the dermatology clinic with a chronic rash of 20 years' duration on the right ankle that extended to the instep of the right foot. His medical history was notable for hypertension and hyperlipidemia. Family history was unremarkable. The patient described the rash as red and scaly but denied associated pain or pruritus. Over the last 2 to 3 years he had tried treating the affected area with petroleum jelly, topical and oral antifungals, and mild topical steroids with minimal improvement. Complete review of systems was performed and was negative other than some mild constipation. Physical examination revealed an erythematous scaly patch on the dorsal aspect of the right ankle. Potassium hydroxide preparation and fungal culture swab yielded negative results, and a shave biopsy was performed.

What's the diagnosis?

a. allergic contact dermatitis
b. nummular eczema
c. pagetoid reticulosis
d. psoriasis vulgaris
e. tinea corporis

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The authors report no conflict of interest.

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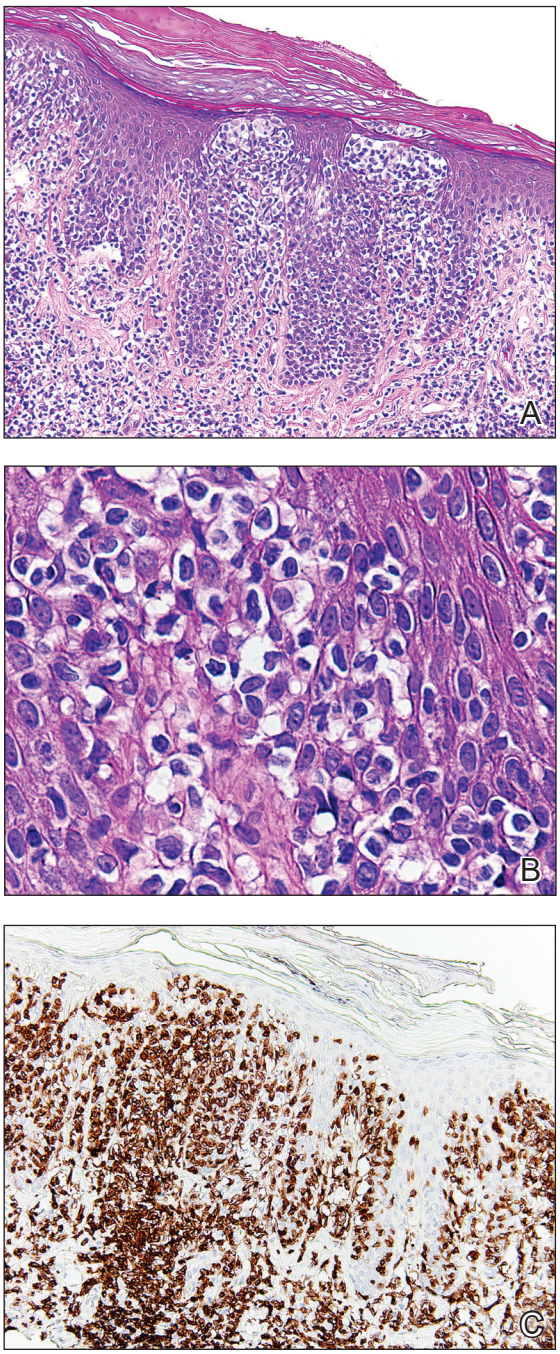
The Diagnosis: Pagetoid Reticulosis

Histopathologic examination demonstrated a dense infiltrate and psoriasiform pattern epidermal hyperplasia (Figure, A). There was conspicuous epidermotropism of moderately enlarged, hyperchromatic lymphocytes. Intraepidermal lymphocytes were slightly larger, darker, and more convoluted than those in the subjacent dermis (Figure, B). These cells exhibited $CD3^+$ T-cell differentiation with an abnormal $CD4^-CD7^-CD8^-$ phenotype (Figure, C). The histopathologic finding of atypical epidermotropic T-cell infiltrate was compatible with a rare variant of mycosis fungoides known as pagetoid reticulosis (PR). After discussing the diagnosis and treatment options, the patient elected to begin with a conservative approach to therapy. We prescribed fluocinonide ointment 0.05% twice daily under occlusion. At 1 month follow-up, the patient experienced marked improvement of the erythema and scaling of the lesion.

Pagetoid reticulosis is a primary cutaneous T-cell lymphoma that has been categorized as an indolent localized variant of mycosis fungoides. This rare skin disorder was originally described by Woringer and Kolopp in 1939[1] and was further renamed in 1973 by Braun-Falco et al.[2] At that time the term *pagetoid reticulosis* was introduced due to similarities in histopathologic findings seen in Paget disease of the nipple. Two variants of the disease have been described since then: the localized type and the disseminated type. The localized type, also known as Woringer-Kolopp disease (WKD), typically presents as a persistent, sharply localized, scaly patch that slowly expands over several years. The lesion is classically located on the extensor surface of the hand or foot and often is asymptomatic. Due to the benign presentation, WKD can easily be confused with much more common diseases, such as psoriasis or fungal infections, resulting in a substantial delay in the diagnosis. The patient will often report a medical history notable for frequent office visits and numerous failed therapies. Even though it is exceedingly uncommon, these findings should prompt the practitioner to add WKD to their differential. The disseminated type of PR (also known as Ketron-Goodman disease) is characterized by diffuse cutaneous involvement, carries a much more progressive course, and often leads to a poor outcome.[3] The histopathologic features of WKD and Ketron-Goodman disease are identical, and the 2 types are distinguished on clinical grounds alone.



Pagetoid reticulosis histopathologic findings from a lesion on the right ankle including a dense infiltrate and psoriasiform pattern epidermal hyperplasia (A)(H&E, original magnification ×40). At higher magnification, conspicuous epidermotropism of moderately enlarged, hyperchromatic lymphocytes was seen (B)(H&E, original magnification ×400). Immunohistochemical stain was positive for CD3 (C)(original magnification ×40).

Histopathologic features of PR are unique and often distinct in comparison to mycosis fungoides. Pagetoid reticulosis often is described as epidermal hyperplasia with parakeratosis, prominent acanthosis, and excessive epidermotropism of atypical lymphocytes scattered throughout the epidermis.[3] The distinct pattern of epidermotropism seen in PR is the characteristic finding. Review of immunocytochemistry from reported cases has shown that CD marker expression of neoplastic T cells in PR can be variable in nature.[4] Although it is known that immunophenotyping can be useful in diagnosing and distinguishing PR from other types of primary cutaneous T-cell lymphoma, the clinical significance of the observed phenotypic variation remains a mystery. As of now, it appears to be prognostically irrelevant.[5]

There are numerous therapeutic options available for PR. Depending on the size and extent of the disease, surgical excision and radiotherapy may be an option and are the most effective.[6] For patients who are not good candidates or opt out of these options, there are various pharmacotherapies that also have proven to work. Traditional therapies include topical corticosteroids, corticosteroid injections, and phototherapy. However, more recent trials with retinoids, such as alitretinoin or bexarotene, appear to offer a promising therapeutic approach.[7]

Pagetoid reticulosis is a true malignant lymphoma of T-cell lineage, but it typically carries an excellent prognosis. Rare cases have been reported to progress to disseminated lymphoma.[8] Therefore, long-term follow-up for a patient diagnosed with PR is recommended.

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