Woringer-Kolopp Disease of the Foot

A Case Report

James J. Lapolla, DPM*
Lawrence A. DiDomenico, DPM†
Robert T. Brodell, MD‡
Michael L. Casteel, DPM§

Woringer-Kolopp disease is a rare variant of mycosis fungoides, a type of cutaneous T-cell lymphoma. Described is a case of a small annular plaque on the foot diagnosed histologically as Woringer-Kolopp disease and treated successfully with topical and intralesional steroids. In addition, a brief review of the literature and treatment options is provided. (J Am Podiatr Med Assoc 110(6): 000-000, 2020)

Cutaneous lymphomas are classified based on their growth patterns and cytomorphology (cell lineage). In general, this leads to a division between B-cell, T-cell, and natural killer–cell cutaneous lymphomas, with T-cell involvement accounting for roughly 40% to 50% of cases.[1,2] Mycosis fungoides is a type of cutaneous T-cell lymphoma phenotypically defined by the involvement of effector memory T cells (compared to Sézary syndrome, a more aggressive form involving central memory T cells).[1,3] Mycosis fungoides involves lymphocytic infiltrates into both the dermis and epidermis, as compared to Woringer-Kolopp (WK) disease, which is a rare subset of mycosis fungoides involving dense infiltrates into only the epidermis with scant dermal involvement.[4] Typical phenotypic lineage of WK lymphocytes are either CD4 T-helper cells, CD8 T-cytotoxic/suppressor cells, or CD4/CD8 double-negative cells.[5,6] Clinically, WK disease presents as a solitary hyperkeratotic plaque or erythematous patch on the extremities that is slow growing in nature.

WK was first described by Frederick Woringer and Pierre Kolopp in 1939 and later termed "pagetoid reticulosis" by Braun-Falco et al in 1973, secondary to the histologic appearance of abnormal pagetoid cells within the epidermis.[7] Ketron-Goodman disease is the only other cutaneous condition defined as "pagetoid reticulosis." Although it is similar histologically to WK disease, it is more aggressive and typically presents with disseminated lesions localized to the extremities.[8,9] Woringer-Kolopp disease does not have a gender predilection and more commonly affects adults.[4] The differential diagnosis includes allergic contact dermatitis with epidermotropism, solitary plaque psoriasis, Bowen disease (squamous cell carcinoma in situ), unilesional mycosis fungoides, and the palmaris et plantaris variant of mycosis fungoides.[10-12] In general, WK disease has a good prognosis, with local recurrence rare after initial treatment. Our report describes a case of WK disease on the foot of a middle-aged woman successfully treated with topical and intralesional steroids.

Case Report

A 55-year-old white woman presented to our office with a chief complaint of right heel pain. In addition, she also stated that there was a nonpruritic plaque on her right foot that had been bothering her, located along the lateral forefoot at the border of the glabrous junction (Fig. 1). The patient related no history of trauma, recent travel, insect bite, chemical contact, or change in medication. She stated that the plaque had been slow growing and she could not recollect when it had appeared. Her medical history was unremarkable and she reported no history of skin cancer, psoriasis, or any other autoimmune disease. The plaque measured approx-

*Northeast Ohio Foot, Ankle and Wound Center, Warren, OH.

†Forum Health, Youngstown, OH. Dr. DiDomenico is now with Northern Ohio Medical Specialists (NOMS) Ankle and Foot Care Centers, Boardman, OH.

‡University of Mississippi Medical Center, Jackson, MS.

§The Villages Health, The Villages, FL.

Corresponding author: Michael Casteel, DPM, The Villages Health, 2910 Brownwood Blvd, The Villages, FL 32163. (E-mail: mikecasteeldpm@gmail.com)

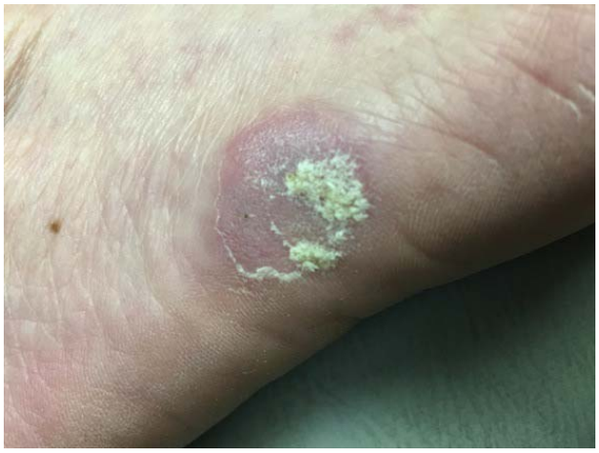

Figure 1. Clinical appearance of plaque on initial presentation.

imately 1.0 × 1.0 cm and was annular in appearance, with central hyperkeratosis and minimal surrounding erythema. Sublesional calor, induration, or fluctuance was not noted. Two 4-mm punch biopsies were subsequently performed on different locations within the plaque.

Histopathologic evaluation revealed an atypical epidermotropic lymphocytic infiltration with reactive lymphocytes localized in the dermis (Figs. 2 and 3). In addition, the reactive lymphocytes colonized the basal layer and produced parabasilar aggregates consistent with Pautrier microabscess formation. The epidermal response was primarily that of hyperplasia with overlying orthohyperkeratosis. Immunohistologically, the neoplastic cells stained positive for the CD3 and CD8 phenotypes and negative for the CD20 and CD30 phenotypes, with

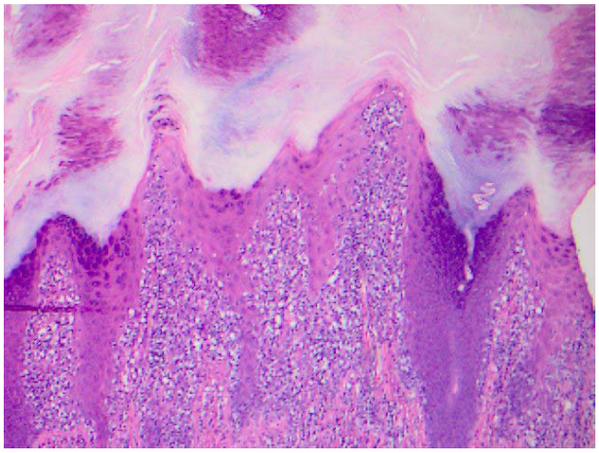

Figure 2. Photomicrograph of plaque.

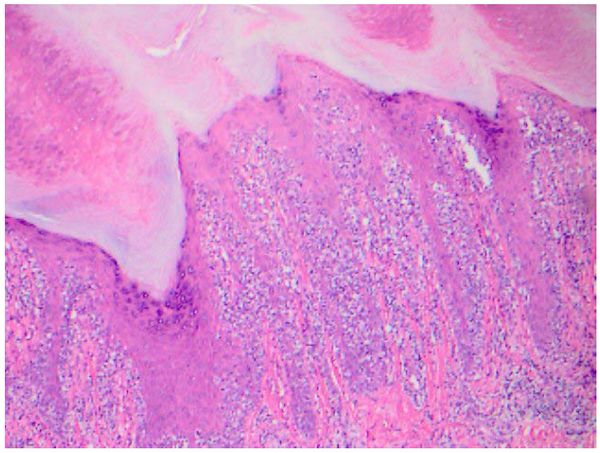

Figure 3. Additional photomicrograph of plaque.

further analysis showing the predominate lineage being the CD8 phenotype.

The plaque was subsequently diagnosed as WK disease and treated initially using triamcinolone 0.1% cream applied twice a day for 8 weeks (Fig. 4). An intralesional injection of 1 cc of 4 mg/ml dexamethasone phosphate was also performed at the conclusion of the 8-week topical therapy regimen and performed again 3 months later. To date, there has been no clinical evidence of plaque recurrence (Fig. 5).

Discussion

As with any skin lesion, an accurate diagnosis of WK disease is obtainable only by histopathologic evaluation. Because it is confined solely to the epidermis, diagnosis of WK disease obtained using

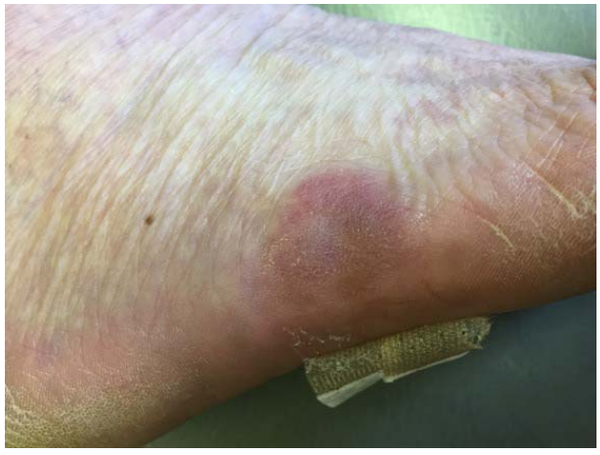

Figure 4. Clinical appearance after 8 weeks of topical triamcinolone 0.1% cream.

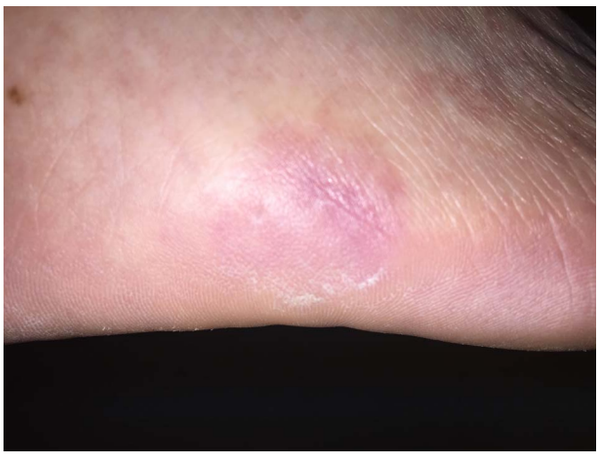

Figure 5. Final clinical appearance after treatment cessation.

either a punch or deep shave (saucerization) biopsy is adequate. Woringer-Kolopp disease has a good prognosis because of the indolent nature of its lesions with rare local recurrence after treatment.[1] In addition, although advanced lesions can become verrucous and hyperkeratotic in appearance, WK disease has little propensity for dissemination or visceral involvement.[6] Effective treatment options include topical nitrogen mustard, high-potency topical steroids, phototherapy, or topical photodynamic therapy.[1,4,13,14] Because of the excellent response to these methods, surgical excision is not required for most cases.

In this study, treatment was dictated by our senior author (J.L.), with topical steroids being the primary modality. Intralesional steroid injection was used as an adjuvant later in the treatment course and, clinically speaking, appeared to be effective in helping resolve the WK plaque. To our knowledge, this is the first reported case of using intralesional steroids as an additional treatment modality in the resolution of WK disease. Further research on the effectiveness of intralesional steroids versus topical steroids would certainly be warranted.

Conclusions

Woringer-Kolopp disease is a rare, slow-growing variant of T-cell lymphoma characterized by atypical lymphocytic infiltration into an acanthotic epidermis. Our case study shows that topical and intralesional steroids, when combined, can be an appropriate treatment regimen for this condition, with no side effects or recurrence demonstrated.

Financial Disclosure: None reported.
Conflict of Interest: None reported.

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