A Case of Keratosis Lichenoid Chronica

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Keratosis lichenoid chronica is a very rare disease. It was originally described by Kaposi and later named Nekam Disease after Nekam reported a case in 1938.1 This rare dermatosis is characterized by violaceous papular and nodular hyperkeratotic lesions with overlying gray scales. They are primarily found on the dorsum of hands and feet along with on the extremities and trunk. Initially discrete, the lesions often coalesce and form warty, lichenous-like lesions arranged in linear and reticulate patterns. Associated with the disease are erythematous telangiectasias of the face resembling rosacea (H. Foong, S. Ghosn, J. Bhawan, Virtual Grand Rounds in Dermatology online forum, December 11, 2004). Nail bed changes including thickening, discoloration, onycholysis, paronychia, and warty lesions of the periungual areas have also been reported.1

Hyperkeratosis, parakeratosis, and hypergranulosis of the outer epidermis is also seen histologically. The epidermis is acanthotic with liquefied degeneration of the basal cell layer. Noted in the upper dermis is lymphocytic infiltrate, containing few plasma cells in addition to perivascular and peri-appendageal involvement. The histology of keratosis lichenoid chronica mimics that of warty lichen planus. Differentiation is made clinically with lesion pattern distribution and the absence of pruritus.2

Although very resistant to treatment, the use of psoralen ultraviolet A therapy (PUVA), topical calcipotriol, and oral retinoids have been helpful treating the disease.2–6

History and Physical Examination

A 58-year-old male was seen in consultation at a rehabilitation center for which he was admitted status postcerebrovascular accident. He relates a history of more than 15 years of intermittently painful hyperkeratotic plaques and nodules diffusely located over his trunk and extremities bilaterally (Fig. 1). The plaques began on the elbows and knees then progressed to involve the trunk and distal extremities. Plaques located at the ankle level progressed into intensely raised nodules over a period of a few years.

Previous treatments included topical calcipotriene; cryotherapy, which was temporarily helpful; and oral retinoids, which were discontinued because of painful xerosis.

Laboratory Data and Histopathology

Biopsies in 1998 and 2009 revealed epidermal hyperplasia with alternating epidermal atrophy and overlying neutrophil-rich parakeratotic scale, lichenoid inflammation, and spongiosis with exocytosis of lymphocytes. Also, lichenoid infiltrate composed of lymphocytes, histiocytes, eosinophils, and plasma cells was seen (Fig. 2). All of the above features are consistent with keratosis lichenoid chronica. A nail biopsy taken in 2010 revealed only fungal elements and no nail involvement of keratosis lichenoid chronica.

Treatment

The patient was started on 40% urea cream, which proved helpful on the smaller, nonviolaceous regions but did not relieve the larger, nodular
lesions (Figs. 3 and 4). The patient was offered PUVA therapy but refused due to inability to comply with strict schedule upon discharge. He was also offered surgical debridement of the large nodular regions of the lower extremities but also declined this treatment.

Conclusions

Keratosis lichenoid chronica is a rare and difficult-to-treat dermatologic disorder. PUVA therapy remains the preferred and most responsive treatment. However, due to its strict need for compliance and longevity of the treatment many patients withdraw from treatments. Other alternatives include oral retinoids; however, they can cause extremely xerotic skin and cause a problem with compliance.

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References


