JAMA Dermatology Clinicopathological Challenge

A Patient With a 10-Year History of Generalized Pruritus

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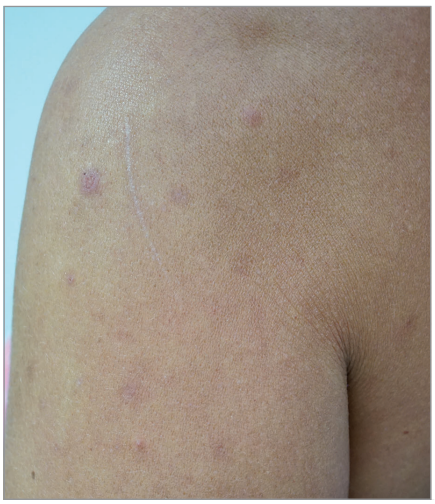
A Clinical image of shoulder



B Clinical image of lower lip

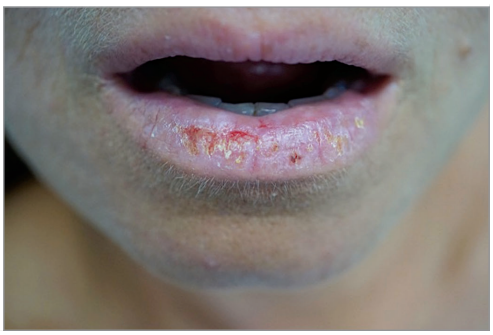


Figure 1. A, Clinical image of actinic prurigo in the skin of the patient showing unspecific erythematous papules on the arm. B, Clinical image of actinic prurigo cheilitis in the patient showing scales, crusts, and fissures on the lower lip.

A white woman in her early 40s presented with generalized pruritus of 10-years' duration. She had no other medical history. The patient provided the report of a skin biopsy performed 2 years before presentation. It was described as indicating spongiotic dermatitis. She had been treated with topical corticosteroids with no control of the symptoms. Physical examination revealed erythematous papules, some of them crusted, and some excoriations located over the trunk and upper extremities (Figure 1A). In addition, some scales and fissures were seen on her lower lip (Figure 1B), sparing the surrounding skin, as well as inner mouth and upper lip. A new biopsy sample was taken to rule out actinic cheilitis.

WHAT IS YOUR DIAGNOSIS?

- **A.** Herpes simplex virus–associated pseudolymphoma
- **B.** Actinic prurigo cheilitis
- **C.** Cutaneous lymphoid hyperplasia
- **D.** Marginal zone lymphoma

Diagnosis

B. Actinic prurigo cheilitis

Microscopic Findings and Clinical Course

The biopsy results showed spongiosis and erosions in the epidermis and a nodular lymphocyte infiltrate in the upper and mid dermis with a peripheral mantle zone surrounding a central germinal center with cells that were CD20 positive and BCL-2 negative (Figure 2). All these findings were diagnostic of follicular cheilitis, also known as actinic prurigo (AP) cheilitis. A phototest with a solar simulator (Saalman Multitester TYP SBC LT 400) was carried out with the immediate reading negative for UV-B, UV-A, and visible light. However, erythema appeared in the area irradiated with UV-A 48 hours afterwards. Blood test results including antinuclear antibodies and complement, along with urine porphyrin analysis were normal. A diagnosis of AP with associated cheilitis was made. Treatment with narrowband UV-B at a dose of 0.1 J/cm^2 was started. After 33 sessions and a cumulative dose of 4.5 J/cm^2 combined with sunscreen use when outdoors, the patient was completely asymptomatic.

Discussion

Actinic prurigo is an uncommon photosensitive disease that has generally been diagnosed in people native to Central and South America, although some series describe the disease in white patients.[1,2] Its pathogenesis is not well elucidated but it has been related to human leucocyte antigen (HLA) DR4 and in particular HLA DRB1*0407, and is considered to be a type IV hypersensitivity

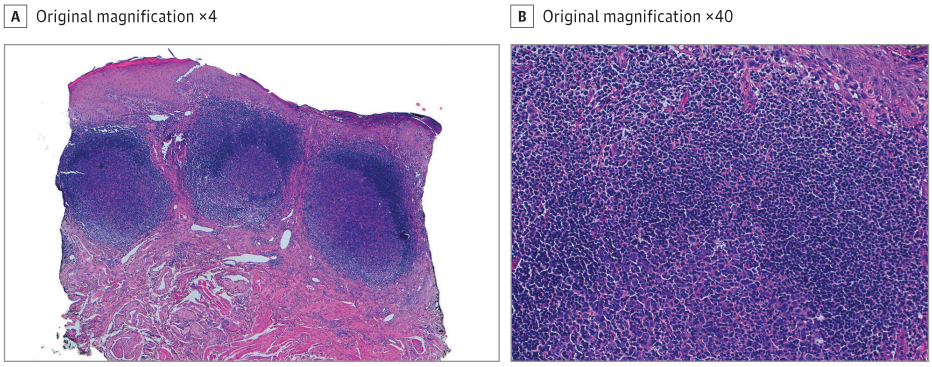


Figure 2. A, Hematoxylin-eosin staining demonstrates spongiosis and erosions over the epidermis and a nodular infiltrate in the upper and mid dermis composed of lymphoid aggregates with ample germinal centers. These enlarged lymphoid follicles coalesced into irregular shapes but maintained discernible mantle and marginal zones. B, Higher magnification shows the germinal center and mantle of the lymphoid follicles in the dermis.

reaction.[3] Actinic prurigo usually appears in childhood, although late onset in adulthood is well described.[4] The main clinical features of AP are itching and erythematous papules; vesicles and scarring can also be present. In general, the lesions appear on the face and forearms but in covered sites as well. Mucosal surfaces, such as the conjunctivae and lips, may also be affected. Some authors report a more severe course with frequent mucosal involvement in children compared with adults.[5] In the acute phase, AP cheilitis biopsy results are unspecific with acanthosis, spongiosis, edema, and an inflammatory infiltrate. In a chronic phase it characteristically shows well-formed dermic lymphoid follicles as described in this patient. This pattern is considered by some authors as a very characteristic finding of AP[5] and was the key to the diagnosis of this patient. Cutaneous lesions are less specific. Early lesions show epidermal spongiosis, acanthosis, and a dermal perivascular inflammatory infiltrate. Chronic lesions present lichenification, excoriations, and a heavier mononuclear infiltrate. Otherwise, the nonspecificity of symptoms and skin lesions, along with the rarity of this disease in our setting had been responsible for the diagnostic delay. Regarding other differential diagnoses for this patient, herpes simplex virus (HSV), including orolabial HSV infection, may demonstrate histologic features of pseudolymphoma without obvious viral cytopathic changes in some cases,[6] but the patient's clinical presentation was not suggestive of HSV infection. Cutaneous lymphoid hyperplasia can show a similar histopathology to this case; nevertheless it has a different clinical presentation, which is typically unilesional or regionally grouped lesions.[7] Marginal-zone lymphoma can also show a nodular infiltrate in the dermis, but characteristically the centers of the nodules show dark areas surrounded by a pale area, in which neoplastic Bcl-2–positive cells predominate.

Regarding treatment, many options such as strict photoprotection, steroids, antimalarials, and thalidomide have been tried with different outcomes. Macfarlane et al[1] presented UV-B desensitization as a treatment option and this is the treatment that showed the best results in this patient.

Conclusions

We describe an adult woman with a medical 10-year history of pruritus, who showed lesions of AP cheilitis that were biopsied and informed as follicular cheilitis giving us the diagnosis of AP, and who was treated with low doses of UVB with resolution of the pruritus.

ARTICLE INFORMATION

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