JAMA Dermatology Clinicopathological Challenge

An Atrophic and Spiny Eruption of the Palms

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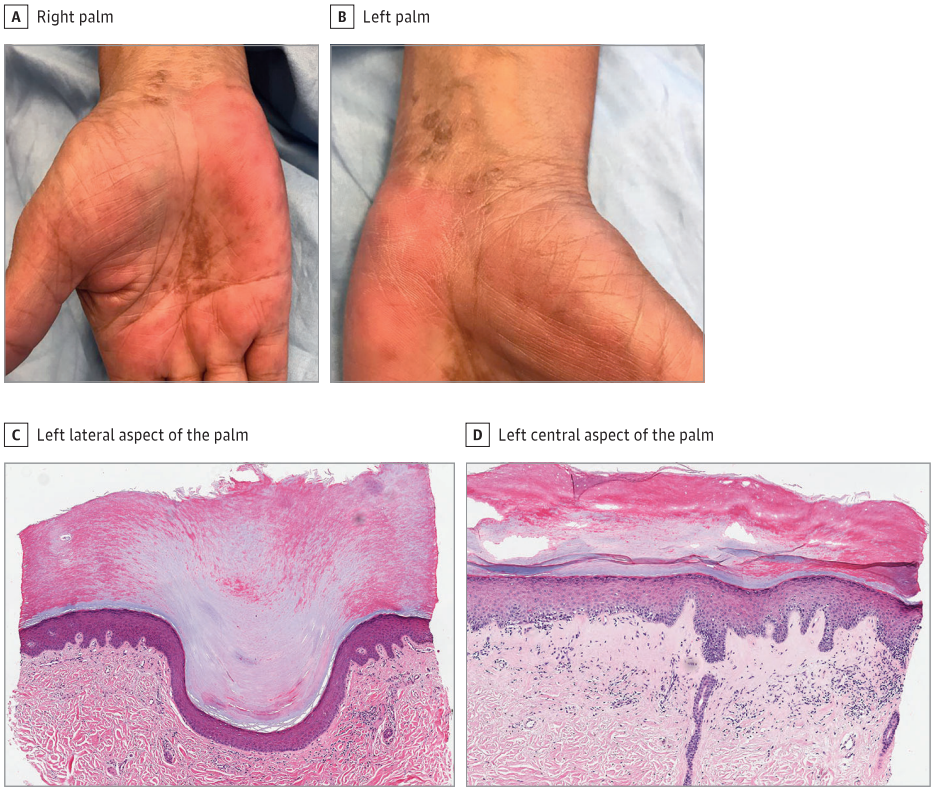

Figure. A, An atrophic hyperpigmented plaque with spiny papules in the palmar creases. B, Similar features extending onto the volar wrist. C and D, Histopathologic images (hematoxylin-eosin, original magnification ×100).

A woman in her 60s presented for initial dermatologic evaluation of a 2-year history of a pruritic and sometimes burning hyperpigmented eruption on the bilateral palms. The eruption had gradually spread from the left palm to the right palm and then focally onto the bilateral volar aspect of the wrists in recent months (**Figure**, A and B). She had not experienced involvement of the plantar surfaces. No additional concerns were reported, and there was no family history of similar lesions. There was no known history of travel in relation to onset of the lesions. Limited clinical examination revealed well-marginated brown atrophic patches on the bilateral palms extending focally onto the volar aspect of the wrist and centrally surmounted by spiny keratotic pits in the palmar creases. Two punch biopsy specimens were obtained for diagnostic clarification (Figure, C and D).

WHAT IS YOUR DIAGNOSIS?

A. Lichen sclerosus et atrophicus

B. Acrodermatitis chronica atrophicans

C. Acquired palmoplantar keratoderma

D. Sclerotylosis (Huriez syndrome)

Diagnosis

A. Lichen sclerosus et atrophicus

Microscopic Findings

The first specimen demonstrated compact orthohyperkeratosis overlying an attenuated epidermis with a central dell and preserved granular layer. These features, in isolation, raised a histopathologic

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differential diagnosis of punctate palmoplantar keratoderma. The second specimen revealed similar orthohyperkeratosis and an effaced and atrophic epidermis; however, in the dermis, hyalinization of the papillary dermis above a thin band of lymphocytic inflammation was present. Close inspection and deeper levels of the first specimen revealed subtle interface changes and superficial dermal hyalinization. These features were diagnostic of lichen sclerosus of the palms.

Discussion

The patient returned to the clinic for discussion of biopsy results. When asked specifically, she reported significant vulvar pruritus, which had previously been treated unsuccessfully by her primary care physician and gynecologist; she realized in retrospect that this pruritus had developed around the same time as the palmar eruption. Examination of the external genitalia revealed a pearly white patch involving the anterior aspect of the labia minora and clitoral hood; no advanced scarring or fusion of anatomical structures was present. Treatment with an ultrapotent topical steroid and a calcineurin inhibitor was initiated at both sites of involvement.

Lichen sclerosus involving the palms has been reported 3 times in the English-language literature. It was first described in 1971 by Purres and Krull in the setting of disseminated disease, both genital and extragenital.[1] This report mentions small keratotic plugs on one of the palms, perceived to be located at the orifices of the sweat ducts. The presented histopathologic findings did not demonstrate the keratotic pits. Previously, the existence of palmar lichen sclerosus was the subject of debate; while Hallopeau's original report mentions a lesion on the volar aspect of the left forearm, it is unclear whether the palmar surface proper was involved.[2]

A second case was described in 1979 by Petrozzi et al.[3] This patient had small whitish papules on the palms and soles in the absence of genital involvement. The histopathologic findings were reported to include hyperkeratotic plugs at adnexal orifices, but were not depicted. In 1984, Tudino and Wong described a patient with bullous and hemorrhagic lesions of the palms and wrists.[4] Additional reports have appeared in non–English-language sources.[5-7]

Lichen sclerosus is an inflammatory sclerosing condition that most commonly affects the external genitalia. Recognition of this entity is crucial because of the risk of squamous cell carcinoma that may develop in untreated or undertreated cases. A recent study revealed a significant decrease in the risk of carcinoma with proper treatment.[8] Involvement of extragenital sites is not uncommon, but in general occurs on the trunk and proximal extremities. Involvement of the palms and soles is rare; less than one dozen cases have been reported, although this condition may be both underrecognized and underreported.

The features of extragenital lichen sclerosus have been well documented in other sources.[9] The histopathologic findings are similar to those found in the genital area but also include prominent follicular plugging at hair-bearing sites. The keratotic palmar pits, which may clinically simulate punctate keratoderma, may represent a recapitulation or forme fruste of this process at a glabrous site. While disease isolated to the palms and soles has been reported, most cases have presented with concomitant vulvar disease. Identification of palmar disease may lead to diagnosis of vulvar disease (as in this case), which may in turn reduce the patient's risk of malignant neoplasm with proper treatment. While malignant transformation of extragenital lichen sclerosus has been reported, none of the patients with volar acral involvement have developed squamous cell carcinoma.[10]

Acrodermatitis chronica atrophicans is a manifestation of chronic infection with Borrelial species, most commonly observed in persons who have visited or live in Europe. Nonvolar skin is more prominently affected clinically, and a prominent plasmacellular infiltrate is often seen histologically. Acquired palmoplantar keratoderma was considered in the initial differential diagnosis, and orthokeratotic epidermal dells with preserved granular layer typify the microscopic appearance of punctate keratoderma; however, the dermal infiltrate and sclerosis are not seen. Sclerotylosis is a rare variant of hereditary keratoderma characterized by diffuse inflammatory keratoderma of the palms and soles and scleroatrophic changes of the digits.

ARTICLE INFORMATION

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