Palmar lichen sclerosus et atrophicus

Jennifer Seyffert, DO,[a] Nathan Bibliowicz, DO,[a] Tanner Harding, MS,[b] Rajiv Nathoo, MD,[a] and Sadia Saeed, MD[a]
Orlando, Florida

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INTRODUCTION

Lichen sclerosus et atrophicus (LS&A) is a chronic inflammatory dermatitis that most commonly involves female anogenital skin.[1] Extragenital disease occurs exclusively in 10% of patients and classically manifests as ivory atrophic papules and plaques on the trunk and extremities, rarely involving the acral surfaces.[1] To our knowledge, we report the fifth case of extragenital LS&A with palmar involvement in the English-language literature.[2-5]

Abbreviation used:

LS&A: lichen sclerosus et atrophicus

CASE REPORT

A 69 year old white woman with a medical history of hypertension, diabetes mellitus type 2, smoking, chronic obstructive pulmonary disease, and hypothyroidism and weight of 78 kg presented with 2-year history of pruritic plaques distributed on arms, trunk, thighs, and palms. Physical examination found erythematous, ivory, and hyperpigmented atrophic plaques ranging in size from 0.5 cm to 4 cm on the proximal extremities and trunk. The patient's palms exhibited diffuse erythema with atrophic and bullous ivory-yellow plaques with dermatoglyphic and eccrine ostia accentuation extending onto the flexural wrist (Fig 1 and Fig 2). Atrophic plaques also overlaid the dorsal distal interphalangeal joints bilaterally. There was no involvement of the anogenital or plantar skin.

Four 4-mm punch biopsy sections were obtained from the left forearm, left posterior thigh, and bilateral palms. Histopathology found compact orthokeratosis overlying an epidermis with focal atrophy, vacuolar degeneration of keratinocytes, homogenous eosinophilic sclerosis of the papillary dermis, and chronic inflammatory infiltrate (Fig 3) consistent with LS&A.

Given the patient's history of smoking, the rare atypical presentation of this condition, and one case report of breast cancer preceded by extragenital LS&A,[6] age-appropriate malignancy screening was pursued to rule out an atypical paraneoplastic phenomenon. Paraneoplastic workup including mammogram, colonoscopy, and chest radiograph only found underlying chronic obstructive pulmonary disease. The patient had no subjective or objective clinical or functional improvement with topical keratolytics, super-potent topical steroids, and a 3-month trial of methotrexate titrated up to 25 mg weekly. As there was no functional or clinical improvement, and the patient complained of nonremitting gastrointestinal upset, methotrexate was discontinued. Given the expense of acitretin, the patient was started on isotretinoin, 30 mg twice a day, topical pimecrolimus 1% cream twice a day, and betamethasone dipropionate 0.05 % cream twice a day pulse therapy. Hypertriglyceridemia despite dietary modification and gemfibrozil resulted in reduction of isotretinoin dose to 20 mg twice a day. Currently, at 6 months of isotretinoin and topical therapy, the patient's pain and pruritus are subjectively improved, allowing for improved comfort, function, and quality of life. Objectively, extragenital lesions on the back, arms, and legs have cleared, and there has been a decrease in acral hyperkeratosis of the palms. Atrophic plaques and erythema remain on the palms and overlying dorsal distal interphalangeal joints with no reduction in plaque size. The patient has been offered physical therapy and excimer laser

From KCUMB-Advanced Dermatology and Cosmetic Surgery[a] and University of Central Florida College of Medicine.[b]

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Correspondence to: Jennifer Seyffert, DO, 260 Lookout Place, Suite 103, Maitland, FL 32751. E-mail: Jennifer.seyffert@adcsclinics.com.

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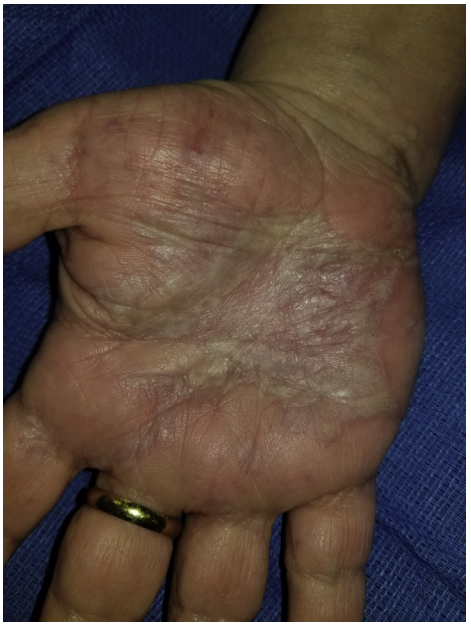

Fig 1. Palmar LS&A. Diffuse erythema with atrophic and bullous ivory plaques with follicular accentuation.

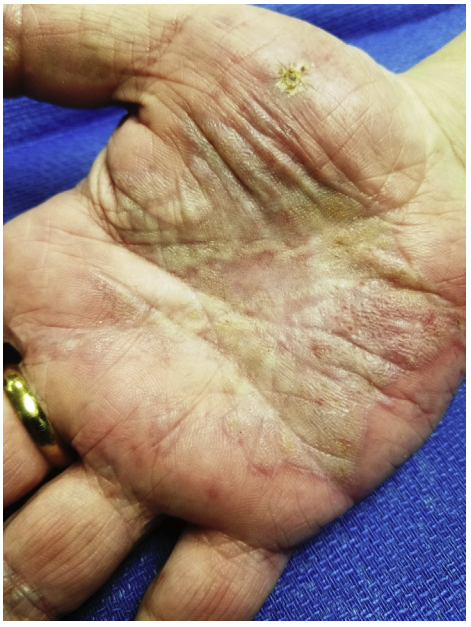

Fig 2. Palmar LS&A. Atrophic plaque with accentuation of dermatoglyphics and eccrine ostia.

therapy; however, difficulty in transportation limits the use of these modalities. Alternative therapies have been discussed based on previous case studies for genital and extragenital LS&A including intralesional steroids, hydroxychloroquine, and mycophenolate mofetil.

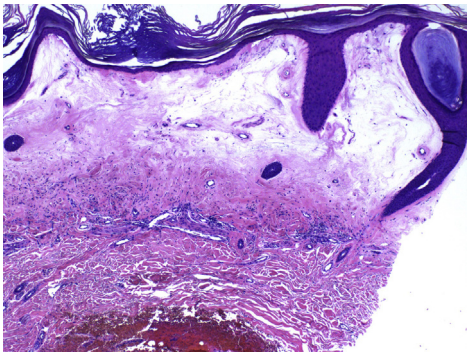

Fig 3. Histopathology. Punch biopsy (4 mm) from left thenar eminence shows hyperkeratosis, epidermal atrophy, subtle vacuolar change, sclerotic papillary dermis, and striking papillary dermal edema, consistent with LS&A. (Hematoxylin-eosin stain; original magnification: ×4.)

DISCUSSION

LS&A is a chronic inflammatory condition that occurs in all ages and races, with a female predominance.[7] An association with major histocompatibility complex class II HLA-DQ7 has been identified.[8] Eighty percent of patients with LS&A have IgG autoantibodies against ECM-1, which may act as an autoantigen.[9] LS&A can affect both anogenital and extragenital skin, with rare involvement of the oral cavity, palms, and soles.

Palmar involvement of LS&A is rarely documented; reported cases are summarized in Table I. The first reported patient with palmar involvement of LS&A was described by Purres and Krull[2] in 1971; their patient also had widespread cutaneous involvement and was resistant to treatment. In 1979, Petrozzi et al[4] reported a case of LS&A limited exclusively to the palms and soles that was resistant to salicylic acid and topical steroids. Tudino and Wong[3] in 1984 reported a female patient with extragenital LS&A with involvement of the palms and flexural wrists with hemorrhagic bullae. Most recently in 2003, Aggarwal et al[5] described a male patient with diffuse extragenital involvement including the palms and soles.

Extragenital disease presents as sclerotic scar-like papules and plaques with ivory coloration, a wrinkled shiny surface, and surrounding erythematous halo. Telangiectasia and follicular plugging can be seen in the more advanced stages. Keratotic plugging can be evident in lesions involving hair-bearing skin, whereas accentuation of eccrine ostia and dermatoglyphics can be prominent on acral skin, as seen in our patient. In general, extragenital LS&A is asymptomatic except for dryness and associated

Table I. Comparison of reported cases of palmar lichen sclerosus

	Purres and Krull[2]	Petrozzi et al[4]	Tudino and Wong[3]	Aggarwal et al[5]	Seyffert et al
Sex	Female	Female	Female	Male	Female
Age, y	71	57	54	58	69
Comorbid conditions	Healthy	Healthy	N/A	N/A	Smoker HTN DM2 Hypothyroidism COPD Hyperlipidemia
Genital involvement	+	-	-	-	-
Plantar involvement	-	+	-	+	-
Generalized cutaneous involvement	+	-	+	+	+
Treatment	Topical and intralesional steroids	Keratolytics, Topical steroids	Topical Steroids	N/A	Topical steroids, topical calcineurin inhibitors, methotrexate, isotretinoin
Treatment response	Refractory	Refractory	Partial success	N/A	Partial success

COPD, Chronic obstructive pulmonary disease; *DM2,* diabetes mellitus type 2; *HTN,* hypertension.

pruritus; this was not the case for our patient, as her extensive palmar disease was painful and limited function of her hands. Based on limited reported cases, acral LS&A presents with ivory-yellow papules which coalesce into plaques with follicular accentuation; bullous variants have been described.[3] Increased fragility of the dermoepidermal junction can result in the development of bullae, which can become hemorrhagic.

Classic histologic findings in LS&A include orthokeratosis, epidermal atrophy, vacuolar degeneration of the basal layer, pale homogenized superficial dermis, loss of elastic fibers, and dermal edema.[10] Hyperkeratosis is pronounced, particularly at the follicular openings leading to follicular plugging; this classic finding may not be present on acral skin given the lack of hair follicles in this anatomic region.

First-line therapy for LS&A is historically potent topical steroids.[1] Topical and oral retinoids, topical and systemic immunomodulators, phototherapy, and surgical procedures have also been reported as therapeutic options. A report detailed plantar LS&A improvement with hydroxychloroquine, 400 mg/d.[11]

Exclusive acral lesions are rare in LS&A; therefore' this diagnosis should prompt further physical examination, particularly of the anogenital skin. Acral LS&A should be considered in the differential diagnosis of ivory atrophic or bullous palmar-plantar lesions. Physicians should be familiar with this unusual entity and management options.

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