Recalcitrant erosive plaques on the palms and soles: A rare manifestation of eccrine syringofibroadenoma

Olivia Chukwuma, BS, Addie Walker, MD, Kiran Motaparthi, MD, and Marjorie Montanez-Wiscovich, MD, PhD
Gainesville, Florida

Key words: acrosyringeal nevus; eccrine syringofibroadenoma; eccrine syringofibroadenomatosis.

INTRODUCTION
Eccrine syringofibroadenoma (ESFA) is a rare cutaneous adnexal lesion of eccrine duct origin, first described by Mascaro in 1963.1 It remains controversial as to whether it is a true neoplastic process, hamartoma, or reactive eccrine hyperplasia. Multiple ESFA, also known as eccrine syringofibroadenomatosis, is an unusual variant of ESFA that has been primarily described in reported cases as palmoplantar hyperkeratosis.2-6 Because of its rarity, ESFA is not often recognized and may be difficult to distinguish from more common inflammatory dermatoses, which may lead to inappropriate therapeutic interventions. We present a rare manifestation of ESFA that presented as progressive erosive plaques on the palms and soles of an elderly woman, which had been unresponsive to several therapies.

CASE
A 98-year-old African-American woman was referred to the dermatology clinic for evaluation of nonhealing lesions on both palms and soles for a year and a half. Her medical history was significant for hypertension, stage 3 chronic kidney disease, and peripheral artery disease. The lesions first developed on her heels as erythematous patches, which slowly progressed to painful, eroded plaques over the course of several months.

After initially presenting to the emergency department for evaluation, she was treated with oral prednisone tapered over 3 weeks (10-40 mg/d) and betamethasone cream with little success. Courses of oral doxycycline, cephalexin, and fluconazole for secondary infections were also administered over the following weeks without improvement. The palms subsequently became involved with asymptomatic redness and scaling, and she was referred to the dermatology department. On questioning, the patient denied a family history of similar symptoms or a personal history of abnormalities of the hair, teeth, and nails.

Physical examination found prominent, well-demarcated eroded plaques on the bilateral plantar heels with bright red granulation tissue and areas of friable, macerated skin (Fig 1, A and B). The bilateral proximal palms were remarkable for hyperkeratosis, scaling, and fissuring plaques (Fig 1, C). Onychoschizia was noted on multiple fingernails. Otherwise, there were no abnormalities of the hair or teeth. The remainder of the cutaneous examination was normal.

The clinical differential diagnosis included keratoderma climacterum, erosive lichen planus, discoid lupus erythematosus, and localized bullous pemphigoid. She was initially treated with oral prednisone (10-20 mg/d for 2 weeks), halobetasol ointment 0.05% with and without occlusion, and soaks with aluminum sulfate and calcium acetate for the following 4 months, all without success. Two punch biopsies were performed from the right heel, revealing epidermal acanthosis with hyperkeratosis, hypergranulosis, and proliferation of superficial...
sweat ducts. The dermis showed fibrosis, reactive vascular proliferation, and sparse chronic inflammation. Direct immunofluorescence was negative. Changes were subtle and thought to be nonspecific. She was then treated for presumptive erosive lichen planus with acitretin for 4 months followed by mycophenolate mofetil for 2 months—neither with improvement. In addition, the plaques on her palms had begun to erode (Fig 1, D) and a new area of scaling and erythema had developed on her left anterior leg and left dorsal foot (Fig 1, E).

Because there was no improvement, 3 additional punch biopsies were performed from the left anterior leg and right palm. Direct immunofluorescence was negative. Histopathology found multiple thin, branching strands of epithelium with ductal lumina extending into the papillary dermis among a background of dermal fibrosis and lymphoplasmacytic inflammatory infiltrate (Fig 2). Taking both sets of biopsies into consideration, these findings were consistent with those of ESFA.

**DISCUSSION**

There are 5 recognized subtypes of ESFA that are classified according to clinical presentation and associated findings: (1) solitary ESFA, which often presents as a single nodule on the extremities of elderly individuals; (2) multiple ESFA without associated cutaneous findings, also known as syringofibroadenomatosis, characterized by multiple lesions with linear and symmetrical arrangement or with a palmoplantar distribution; (3) multiple ESFA associated with hidrotic ectodermal dysplasia (ie, Schöff-Schulz-Passarge and Clouston syndromes), characterized by individual and/or familial abnormalities of the hair, nails, and teeth; (4) nonfamilial unilateral...
linear ESFA, described as unilateral plaques and papules in a linear arrangement; and (5) reactive ESFA, characterized by reactive epithelial and eccrine ductal changes secondary to inflammatory or neoplastic processes, including erosive lichen planus, bullous pemphigoid, leprosy, chronic diabetic foot ulcer, burn scar, venous stasis, ileostomy stoma, squamous cell carcinoma, and nevus sebaceous.1,2,7

The diagnosis of ESFA is made by its unique histopathology, which is shared among all subtypes. Histologically, ESFA is characterized by proliferating cords of epithelial cells with ductal lumina surrounded by an inflammatory fibrovascular stroma, which often contains plasma cells.2 Importantly, given that this pattern may reflect a secondary reactive phenomenon, a primary inflammatory or neoplastic process must be excluded. In this case, multiple biopsies were performed over time to exclude more common inflammatory disorders, adjacent associated malignancy, or malignant transformation.

Previously, 5 cases of eccrine syringofibroadenomatosis with palmomplantar distribution have been reported in the literature. In these reports, the lesions were primarily described as hyperkeratotic papules and plaques.2-6 Our case represents a unique clinical manifestation of syringofibroadenomatosis characterized by progressive erosive palmoplantar plaques.

ESFA generally follows a benign clinical course; however, malignant transformation has been reported, especially with solitary forms.8 Surgical excision is recommended in these cases.8 Once malignant transformation has been excluded, close monitoring of active lesions is preferable. ESFA may resolve spontaneously; however, alternative treatments for refractory ESFA have been reported with varying degrees of success, including radiotherapy, photodynamic therapy, 5-fluorouracil, and imiquimod.9,10

For our patient, after the diagnosis of ESFA was made, we continued halobetasol ointment 0.05% for 1 month with minor improvement. Because of the patient’s concerns regarding the cost of the medication, we replaced halobetasol with triamcinolone ointment 0.1% twice daily, with home wound care 3 times a week using petrolatum gauze and fabric gauze bandages. Over the following 6 weeks, the eroded plaques continued to gradually improve and re-epithelialize (Fig 1, F).

REFERENCES
1. Mascaro JM. Considerations on fibro-epithelial tumors: exocrine syringofibroadenoma. Ann Dermatol Syphiligr. 1963; 90:143-153.
2. Starink TM. Eccrine syringofibroadenoma: multiple lesions representing a new cutaneous marker of the Schöpf syndrome, and solitary nonhereditary tumors. J Am Acad Dermatol. 1997;36(4):569-576.
3. Temnithikul B, Jerasutus S, Sudtikoonaset P, Voravutinon N, Kootiratrakarn T, Kattipathanon P. Eccrine syringofibroadenoma (ESFA): a report of two cases. Dermatol Pract Concept. 2016;6(1):5-8.
4. Lui H, Stewart WD, English JC, Wood WS. Eccrine syringofibroadenomatosis: a clinical and histologic study and review of the literature. J Am Acad Dermatol. 1992;26:805-813.
5. Ochonisky S, Wechsler J, Marinho E, Revuz J. Eccrine syringofibroadenomatosis (mascaro) with mucous involvement. Arch Dermatol. 1994;130(7):933-934.
6. Polat M, Ustun H. Eccrine syringofibroadenomatosis of the soles. J Am Pod Med Assoc. 2016;106(2):141-143.
7. French LE. Reactive eccrine syringofibroadenomatosis: an emerging subtype. Dermatology. 1997;195:309-310.
8. Bjarke T, Ternesten-Bratel A, Hedblad M, Rausing A. Carcinoma and eccrine syringofibroadenoma: a report of five cases. J Cutan Pathol. 2003;30:382-392.
9. Sirikham T, Rojhirunsaokool S, Vachiramon V. Reactive eccrine syringofibroadenoma associated with neuropathy, venous stasis, and diabetic foot ulcer. Case Rep Dermatol. 2016;8(2):124-129.
10. Morganti AG, Martone FR, Macchia G, et al. Eccrine syringofibroadenoma radiation treatment of an unusual presentation. Dermatol Ther. 2010;23:520-523.