Case 25
A 43-year-old Thai man from Bangkok
Chief complaint: Asymptomatic plaques on left foot for 4 years

Present illness: The patient developed a slowly progressive asymptomatic plaques on left foot for 4 years. He was treated by topical corticosteroids with minimal improvement. He has no weight loss, prolonged fever, or night sweat.

Past history:
- Diabetes treated with insulin injection and metformin.
- Neuropathic arthropathy S/P amputation of left 1st, 2nd, 3rd toes.
- Severe venous reflux left SFJ and both CFVs S/P venous stripping both legs.

Physical examination
T 37°C
HEENT: Not pale, no jaundice
Lymph node: Not palpable
Heart: Normal S1S2, no murmur
Lungs: Clear
Abdomen: Soft, no hepatosplenomegaly
Extremities: Amputation of 1st, 2nd, 3rd toes on left foot.

Skin examination
Two discrete well-defined, moist macerated erythematous verrucous plaques on left foot. No signs of venous stasis.

Histopathology (S15-013182, Left foot)
- Multiple foci of thin anastomosing strands of uniform cuboid cells emanating from the epidermis
- Epithelial strands show ductal differenting, embeddes within fibrovascular stroma
Microscopic diagnosis: Eccrine syringofibroadenoma

Diagnosis: Reactive eccrine syringofibroadenoma (ESFA)
Treatment:
Cryotherapy with liquid nitrogen 15 seconds/cycle x 2 cycles on lesion monthly
Fusidic acid ointment apply on lesion BID
Antiperspirant powder apply on left foot BID

Presenter: Thirawut Sirikham, MD
Consultant: Vasanop Vachiramon, MD

Discussion:
Eccrine syringofibroadenoma (ESFA) is a rare benign adnexal neoplasm considered to originate from the excretory portion of eccrine sweat glands. These tumors are believed to arise from the acrosyringium, dermal sweat duct or both. It was first described by Mascaro in 1963. ESFA predominantly affects the elderly between the seventh and eighth decades of life. The clinical appearance of ESFA is typically a slow-growing but varies considerably from a solitary nodules, erythematous plaques to multiple papules. The distribution of lesions is most often on the extremities. The other parts include face, back, abdomen, buttocks, and nails. ESFA is classified into 5 subtypes according to the clinical presentation.

I. Solitary ESFA (most common subtype) a single, non-hereditary, verrucous nodule
II. Multiple ESFA are described in association with Schöpf–Schulz–Passarge syndrome and Clouston syndrome which are caused by mutations in WNT10A and GJB6 respectively.
III. Multiple ESFA without associated cutaneous findings, also called syringofibroadenomatosis
IV. Non-familial unilateral linear ESFA (nevoid ESFA)
V. Reactive ESFA which associated with inflammatory or neoplastic dermatoses such as erosive lichen planus, bullous pemphigoid, burn scars, venous stasis, lymphedema, nail trauma, nevus sebaceous, epidermolysis bullosa, lepromatous leprosy, diabetic foot ulcer, secondary to primary cutaneous amyloidosis, squamous cell carcinoma, Bowen’s disease or clear cell acanthoma.

The differential diagnosis includes chronic eczema, infections (e.g., viral warts, tuberculosis verrucosa cutis, atypical mycobacterium, chromoblastomycosis), squamous cell carcinoma, eccrine poroma, porocarcinoma, fibroepithelioma of Pinkus or clear cell acanthoma.

The histopathological appearance of ESFA is characterized by anastomosing strands and thin epithelial cords, surrounded by abundant fibrous tissue. The epidermis shows long, thin projections, with confluent terminations and a peculiar appearance, similar to crab claws. Focal ductal differentiation was detected within the descending epithelium.

Malignant transformation is possible, mostly involved the non-reactive form of ESFA. Excision of ESFA to prevent malignant degeneration is recommended, although the risk of this occurrence is unknown. Recurrences and metastatic disease have not been reported.

The co-existence of ESFA with squamous cell carcinoma has been described but the incidence of SCC arising in ESFA is unknown. The malignancy developed on severely sun-damaged skin in one case but no other risk factors for developing SCC, such as radiation or immunosuppression.

The treatment of choice is surgical excision with skin grafts or flaps. Other reported treatments are cryotherapy, curettage, electrodessication, ablative laser therapy, radiotherapy, 5-fluorouracil, imiquimod, topical and systemic retinoids.
References