A case report of reactive solitary eccrine syringofibroadenoma

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Abstract

Eccrine syringofibroadenoma is a very rare benign tumour of acrosyringium of eccrine sweat duct. Based on the evidences of known etiological factors, two forms have been proposed; reactive and nonreactive. Reactive forms are rarer, and on even rarer occasions, trauma complicated by secondary nonspecific infections may lead to the development of reactive eccrine syringofibroadenoma, as in our case. Here, we are documenting a case of reactive solitary eccrine syringofibroadenoma in a 65-year-old male presenting with coalescing, firm, pinkish, verrucous nodules and painful deep ulceration on the right sole preceded by trauma and secondary infection. Histopathologic revelation of distinctive microscopic findings confirmed the diagnosis in our case.

Key Words: *Acrosyringium, eccrine syringofibroadenoma, ectodermal dysplasia, hamartomatous*

Introduction

Eccrine syringofibroadenoma (ESFA) is a very rare benign tumour of the acrosyringeal part of the eccrine duct. Since its first description by Mascaro,[1] in 1963, only around 50 cases have been reported globally. It usually involves distal extremities, presenting as solitary or multiple, coalescing, firm, pink or skin-coloured verrucous nodules of variable sizes in a "Streusel-bread"-like appearance at the margin of the hyperkeratotic area with ulceration. It is presently classified into five types depending on its number, pattern, and associated features. Diagnosis on clinical background is very difficult, and therefore, histopathological evaluation is imperative to confirm the diagnosis.

Case History

A 65-year-old male presented with single, painful, non-healing ulcer on the right sole with multiple, pink verrucous nodules at the lateral margin of the ulcer, involving the right lateral malleolus and lateral border of the right sole since two years. On detailed history taking, it was revealed that two years ago, patient had a traumatic injury on his right sole for which he did not consult any registered medical practitioner and applied some homemade chemicals on the wound. Surgical intervention was done to debride the wound by local, untrained, nonmedical personnel without proper antibiotic coverage, which led to secondary infections and nonhealing ulceration. Within the next 6 months, after developing a fully-formed ulcer, multiple small pinkish nodular lesions appeared at the lateral margin of the ulcer, which increased in size with time, coalesced with other nodular lesions, and became verrucous. On local cutaneous examination, a deep seated ulcer of 6 cm × 4 cm × 2 cm size was seen over the right sole, whose floor was clean (because of previous antiseptic cleaning) and the margin was hyperkeratotic and macerated [Figure 1]. Along the lateral border and on the lateral malleolus of right foot, multiple, coalescing, firm, pink-coloured verrucous nodules of varying sizes (largest measuring 3 cm × 2.5 cm) in a "Streusel-bread"-like appearance were present [Figure 2]. The size of the whole coalescing multinodular lesion was 9 cm × 6 cm. There was no regional lymphadenopathy, no loss of sensation, nerve thickening, or any other cutaneous signs of leprosy. There was no clinical or radiological evidence of osteomyelitis. The family history, past medical history, routine laboratory investigations, and systemic examinations were non-contributory.

To reach a final diagnosis, a wedge-shaped incisional biopsy was performed and the sample was sent for histopathological evaluation, which demonstrated vertically

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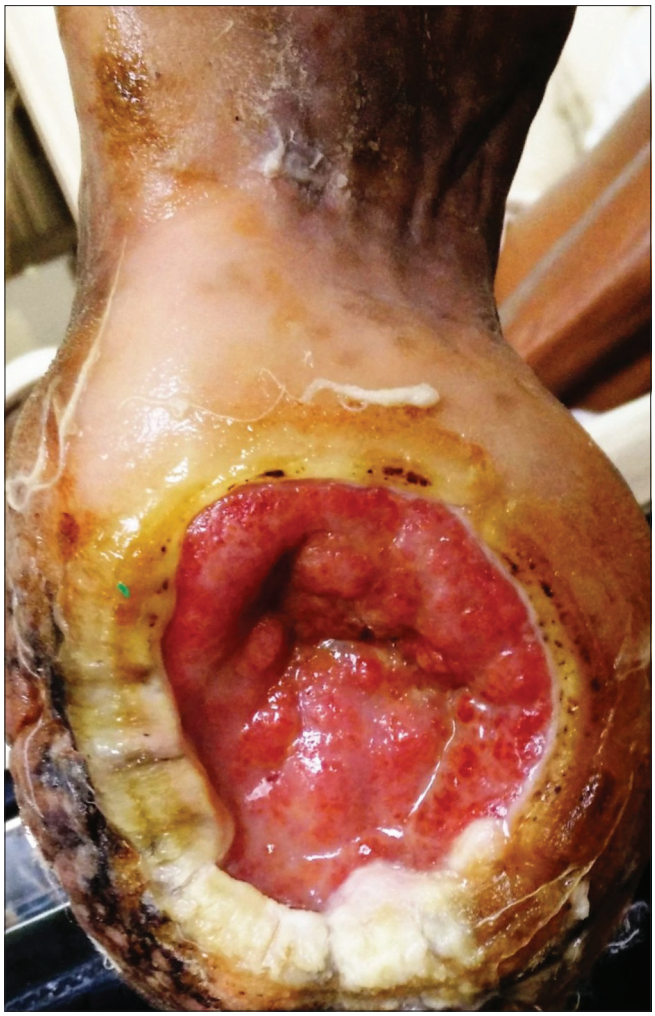

Figure 1: Deep ulcer over the right sole (hyperkeratotic and macerated margin with clean floor)

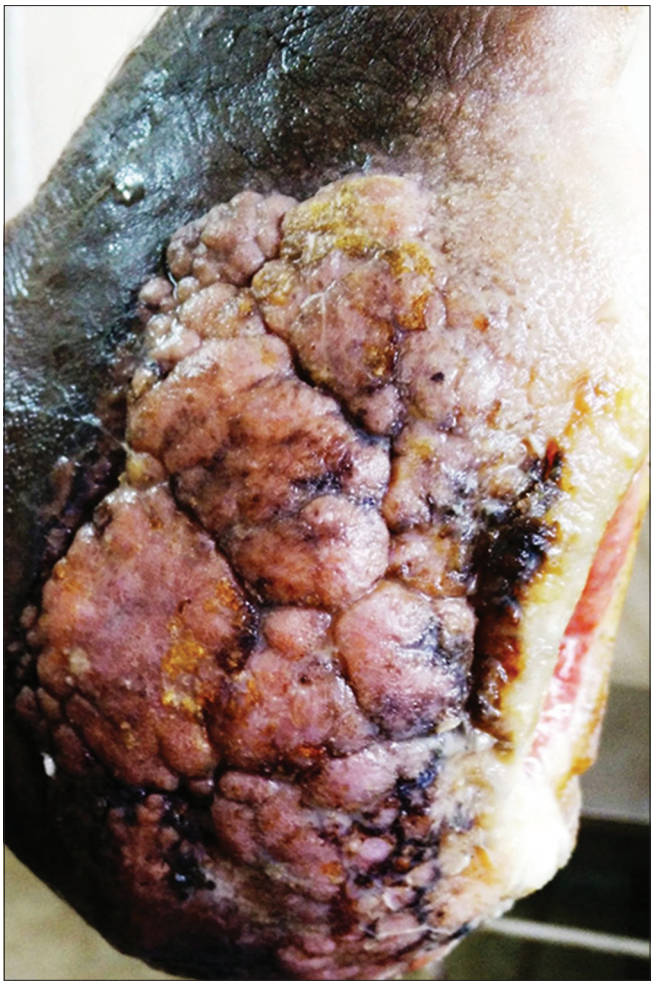

Figure 2: Multiple, coalescing, firm, pink-coloured verrucous nodules of varying sizes in a "Streusel-bread"-like appearance

oriented, multiple, thin, anastomosing strands centred around the acrosyringium [Figures 3 and 4] forming a lattice of double-layered acrosyringeal ductal structures embedded in delicate fibrovascular stroma, extending into the reticular dermis [Figure 5].

On the basis of detailed history, clinical presentations, and distinct histopathological features, diagnosis of ESFA was made, and it was considered to be reactive in nature due to the traumatic nonhealing ulcer.

Discussion

ESFA is a rare, benign, cutaneous adnexal neoplasm with eccrine acrosyringeal differentiation. It was first classified into the following four types by Starink: solitary, multiple ESFA associated with hidrotic ED-like Schopf–Schulz–Passarge syndrome (eyelid cysts, hypotrichosis, hypodontia, nail hypoplasia) or Clouston syndrome (palmoplantar keratoderma, patchy alopecia, nail dystrophy), multiple ESFA without associated cutaneous findings, and nonfamilial unilateral linear ESFA.[1-3] In 1997, the fifth type was recognized as reactive ESFA by French as a ductal hyperplastic or hamartomatous process initiated by repeated damage to the eccrine structures by chronic inflammatory or neoplastic dermatoses such as diabetic foot ulcer, leprous neuropathy, venous stasis or insufficiency, burn scar, BP, EB, naevus sebaceous, stoma of ileostomy, palmoplantar erosive LP, trauma, chronic plaque type psoriasis, epithelioid haemangioendothelioma, and squamous cell carcinoma.[4-6] Reactive ESFA is usually single and acrally located, but if multiple sites are involved, it is termed as "eccrine syringofibroadenomatosis." Apart from these five types, another clear cell variant was also reported by Hu *et al.* in 2005.[7]

Its exact pathogenesis is not yet completely understood but there must be some alteration in the cellular growth and differentiation of epidermal and adnexal structures. Based on few molecular studies, Wnt/β-catenin signalling pathway has been claimed to play the central role whose loss of inhibition or overexpression can lead to abnormal epidermal cellular differentiation and proliferation.[8] Further, strong evidence is required to fully elucidate its pathogenesis.

Clinically, ESFA may have a variable presentation but usually it presents as asymptomatic, solitary or multiple,

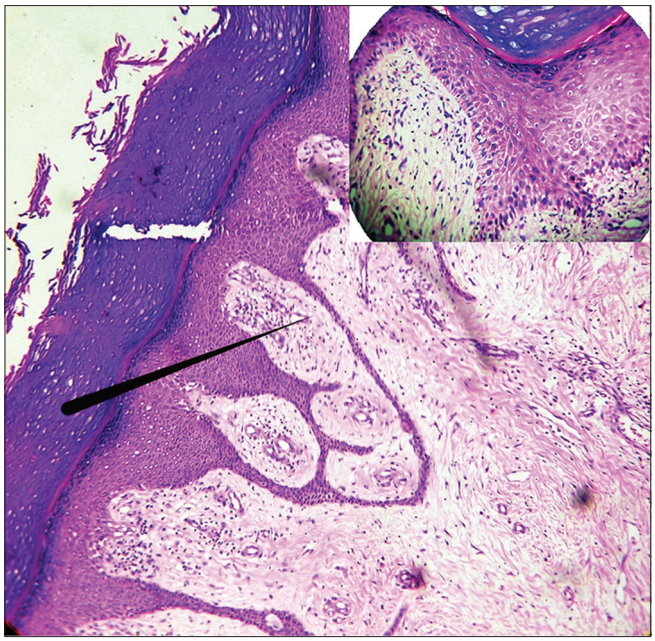

Figure 3: Vertically oriented proliferation of epidermis centred around acrosyringium extending into the reticular dermis (H and E, ×20)

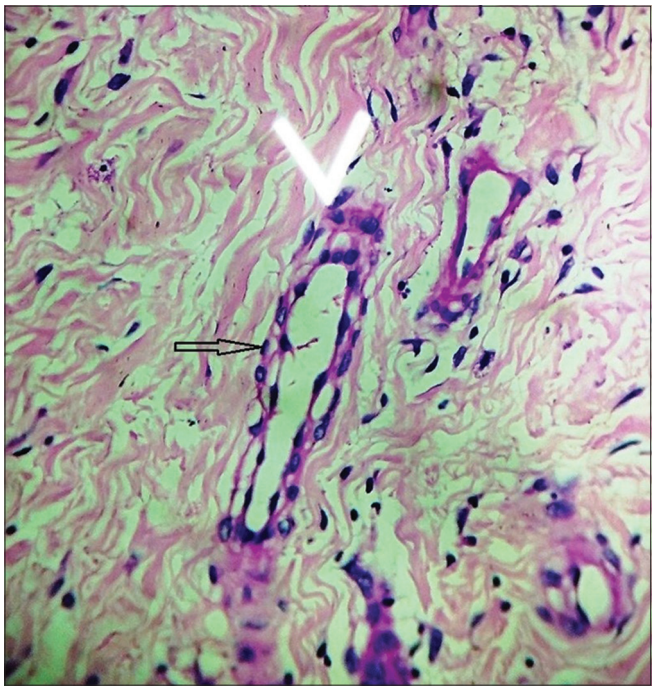

Figure 5: Well-formed eccrine ducts with double-layered cuboidal cells (H and E, ×40)

coalescing, firm, pink/skin-coloured verrucous nodules of variable sizes in a "Streusel-bread"-like appearance at the margin of hyperkeratotic area with or without ulceration. Most commonly affected sites are the feet or lower leg. Other less common sites of predilection are the face, back, abdomen, buttocks, palm, and nails. Most patients are in the 7th–8th decade of age.

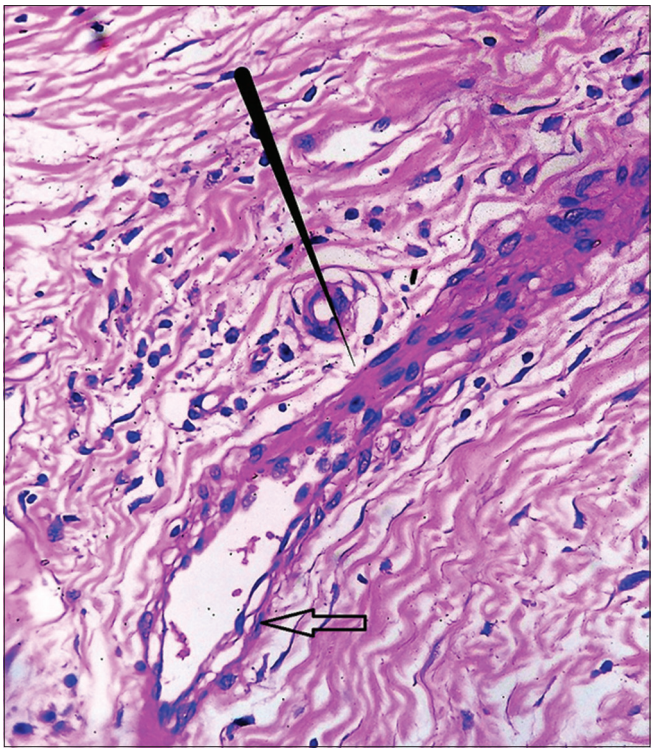

Figure 4: Thin, anastomosing, vertical strand comprising acrosyringeal structures including ductal cuboidal epithelial cells (arrow) and lumina embedded in fibrovascular stroma (H and E, ×40)

The clinical differential diagnoses are tuberculosis verrucosa cutis (TVC), other atypical mycobacterial infections, deep mycoses, and squamous cell carcinoma (SCC). Histopathological differential diagnoses will include eccrine poroma, acrosyringeal nevus, syringofibroadenocarcinoma, fibroepithelial tumor of Pinkus, SCC, and reticulated seborrheic keratosis (SK).

In case of acrosyringeal nevus, strong PAS positivity and plasma cell infiltrate are the distinguishing features, whereas eccrine poroma shows a more uniform small epithelial cell proliferation with vertical thick strands of cells extending into the dermis. In syringofibroadenocarcinoma, there is an area of transformation where a malignant phenotype emerges displaying cytological atypia. Fibroepithelial tumor of Pinkus shows focal changes, which are typical of basal cell carcinoma with peripheral palisading and clefting artefact and loose fibrous stroma.

Histopathologically, it is characterized by distinct diagnostic features, which are observed as multiple, thin, anastomosing strands of cuboidal epithelial cells with ductal structures forming a lattice embedded in loose fibrovascular stroma and connected to the undersurface of the epidermis extending into the dermis. On immunohistochemistry, ductal cells stain positively with S-100, carcinoembryonic antigen (CEA), and CK-19, but in the presence of scarring, CEA and CK-19 may be negative.

This disease generally pursues a benign course, although malignant transformation to eccrine syringofibroadenocarcinoma has been reported in nonreactive ESFA.[9] Moreover, spontaneous resolution can occur in the reactive type.[10] Complete surgical excision is the treatment of choice if the lesion is solitary, not very large, and not in close proximity to vital areas. Other treatment options include CO_2 laser ablation or radiotherapy for lesions at difficult to treat sites. Because of the risk of malignancy, regular follow up and close observation should be done if lesions are left without treatment.

Conclusion

In previously reported cases of reactive type of ESFA, neuropathic ulcer has been the most common underlying aetiology. Trauma complicated by wrong surgical intervention followed by nonspecific secondary infections is a very rarely reported inciting factor for reactive ESFA in the literature, as in our case.

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Conflicts of interest

There are no conflicts of interest.

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