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Research Article

Eccrine Syringofibroadenoma (Of Mascaro): A Rare Case Report

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ABSTRACT
Eccrine syringofibroadenoma is a very rare, benign, adnexal neoplasm derived from cells of the acrosyringium of eccrine sweat ducts and glands, with a propensity for extremities in elderly persons, especially palms and soles. It was first described by Mascaro in 1963. It has a variety of clinical manifestations and distribution of lesions, yet a characteristic histological appearance. In order to illustrate a typical presentation of the tumor, we report a case of eccrine syringofibroadenoma that occurred in a 69-year-old man. He presented to us with unilateral(right) distal lower limb lesions with marked trophic skin changes and extensive non-healing ulcerations associated with necrosis and erythematous nodular plaques, over a period of 1 year. Histopathological evaluation of skin biopsy from representative site revealed slender anastomosing network of monomorphous cuboidal cell cords extending from basal layer of epidermis infiltrating into dermis, with focal luminal differentiation resembling duct structures, embedded in a marked fibrovascular myxoid stroma, consistent with the diagnosis of eccrine syringofibroadenoma. Complete surgical excision is advised for these tumours in view of the risk of carcinomatous transformation.

Keywords: Eccrine Syringofibroadenoma, Benign Adnexal Neoplasm, Acrosyringeum, Ulceration, Erythematous Nodular Plaques, Cuboidal Cell, Fibrovascular Myxoid Stroma.

INTRODUCTION

Lower extremity/pedal dermatological disorders are commonly limited to superficial fungal, viral and bacterial infections, pressure induced and genetic hyperkeratotic disorders, psoriasis and eczemas. Neoplastic diseases are much less common. Eccrine syringofibroadenoma (ESFA) is a very rare adnexal tumor[1-5]. This tumor was described for the first time in 1963 by Mascaro[1]. It develops at the expense of the acrosyringium of the eccrine sweat glands[2-4]. Other name of this lesion is acrosyringeal adenomatosis[6]. The age of onset ranges from 16 to 80 years, but most patients present in the seventh and eighth decades. There is no clear racial or gender predilection[7]. The distribution is also equally variable and includes the face, torso, buttock and rarely the nail. The predilection of ESFA for the extremities has been reported[8].

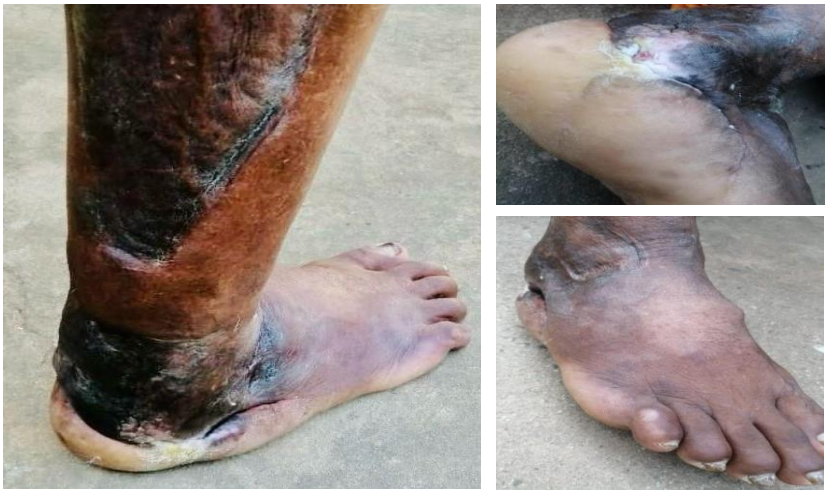
It is presently classified into five types reported in the literature[2-5], depending on its number, pattern, clinical features and associated features. However, its histopathological appearance is peculiar and common to all subtypes[9].

Patients usually present with 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[4]. The most common clinical presentation is solitary, often verrucous papules or nodules. Rarely it may present as disseminated lesions. Clinical course of ESFA is typically benign in nature[10]. However, recent reports suggest the possibility of an association of ESFA with squamous cell carcinoma or malignant transformation of ESFA[11,12]. About 75 cases of ESFA have been reported in the English literature to date, and 18 of the cases (24%) were diagnosed as reactive ESFA[13].

Clinical diagnosis is very difficult and, therefore, histopathological evaluation is essential[4]. To illustrate a typical presentation, we report here a case of eccrine syringofibroadenoma in a chronic leg ulcer setting.

Case Report: A 69 years old hypertensive diabetic male presented to us with chronic,

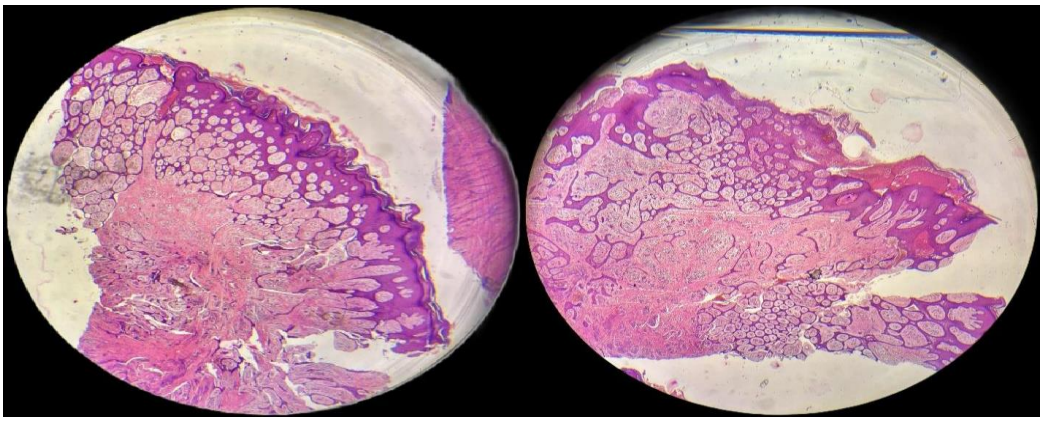
slow-growing, symptomatic, unilateral(right) distal lower limb lesions with marked trophic skin changes and extensive non-healing ulcerations, noticed over a period of 1 year. Also associated with these ulcers was necrosis and surrounding eczematisation. Patient reported of a history of varicose veins and lymphedema.



Cutaneous examination of the right lower limb revealed warty, erosive, ulcerative, flesh-coloured nodules and plaques in a 'Streusel-bread'-like appearance, associated with areas of crusting measuring upto 18 cms in largest dimension. The skin was hyperpigmented and indurated with no loss of sensitivity or any other cutaneous sign of leprosy. The lesion had gradually progressed to attain the current status. A clinical diagnosis of squamous cell carcinoma was considered.

To reach a final diagnosis, a punch biopsy was performed and the sample was sent for histopathological evaluation.

Histopathological evaluation revealed epidermis and dermis. Epidermis was lined by keratinized stratified squamous epithelium exhibiting acanthosis, orthokeratotic hyperkeratosis and papillomatosis. Vertically oriented, branching, slender, irregularly anastomosing strands and cords of proliferating small, monomorphous, cuboidal epithelial cells(acrosyringial cells) emanating from multiple sites of the basal layer of epidermis into the reticular dermis were observed. Some of these projections had a typical 'crab claw' appearance. The cords exhibited focal luminal or ductal differentiation lined with cuboidal cells resembling eccrine ducts but there were no cytological abnormalities. No mitotic figures or neural invasion was seen. The cells were embedded in a cellular and markedly fibrovascular myxoid stroma with mild perivascular lymphocytic infiltration, and exhibited a latticed pattern characteristic of ESFA. It was considered to be reactive in nature due to the association of traumatic nonhealing ulceration. There was no evidence of malignant transformation. Due to financial constraints, we could not perform immunohistochemistry.



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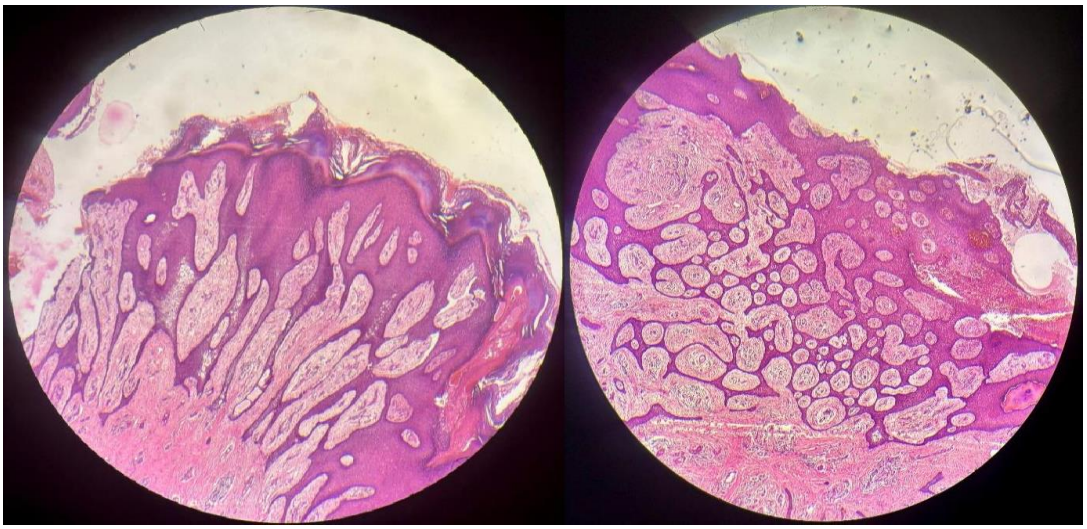

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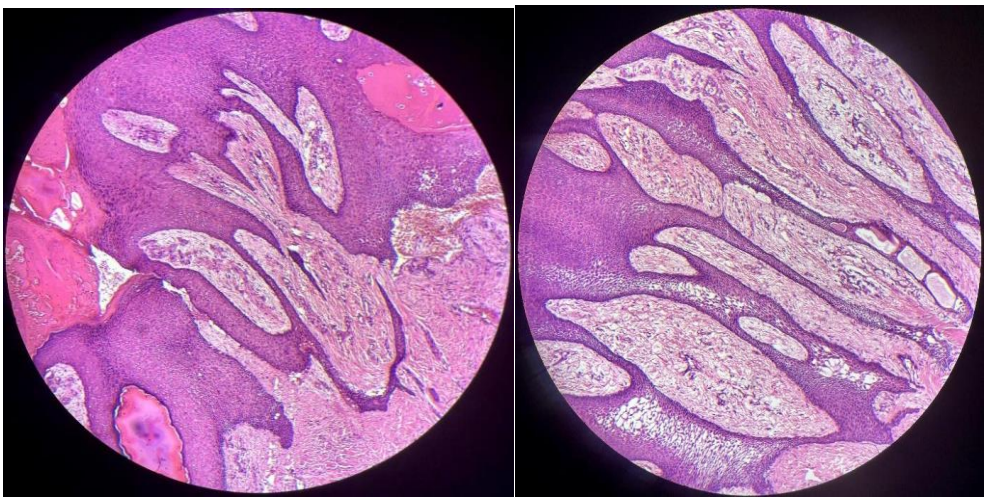

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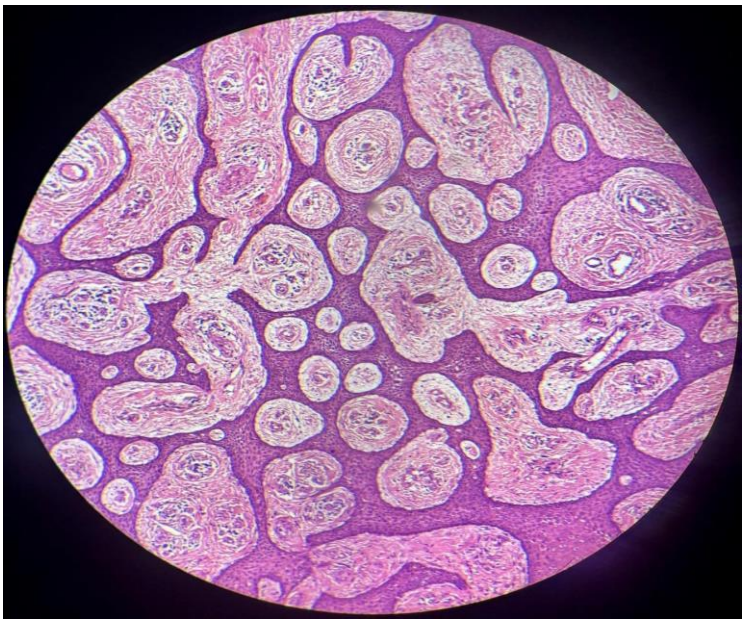

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DISCUSSION

Eccrine syringofibroadenoma (ESFA) (synonyms: syringofibroadenoma, eccrine syringofibroadenomatous hyperplasia[14], acrosyringeal adenomatosis[6]) is a rare benign adnexal tumor arising most often on the extremities of elderly individuals (between seventh and eighth decades of life) with characteristic histopathology. It was first described by Mascaro in 1963[15]. ESFA has a predilection for distal extremities but can affect other sites[16]. It has been reported in the heel, dorsum of foot, leg, face, back, trunk, rarely the nail pulp[3,17], eyelids[18] and perianal region[19].

On histological examination, thin epithelial cords made up of Monomorphous cuboidal cells are observed, anastomosing irregularly and infiltrating the superficial dermis in a reticular pattern with a peculiar appearance, similar to crab claws[20]. These cells are basaloid and smaller than the adjacent keratinocytes[8]. These cords are attached to the epidermis which is almost always hyperkeratotic and acanthotic, as in our case[21,22,18,23-25]. Rich fibrovascular myxoid stroma can be observed between the cords containing ductal structures[26]. There may or may not be lumen formation and discrete lymphocytic infiltrate[8]. Ductal differentiation is detected within the descending epithelium[27]. The tumor is usually limited to the papillary dermis[15]. Malignant transformation is possible, mostly involving the non-reactive form of ESFA[28].

The origin is not well defined, but it is associated with the proliferation of adnexal epithelial cells that form ducts[29], which arise from the excretory portion of the eccrine sweat glands[16]. These tumors are believed to arise from the acrosyringium, dermal sweat duct or both[30]. Human papillomavirus 10 (HPV-10) may also play a role in the development of eccrine syringofibroadenomas.

Clinical findings have been reported to be variable. It can appear as macules, papules, nodules or plaques[4,26,31,32]. The nodules and plaques can take on a budding and warty appearance or even a hyperkeratotic appearance[32,33]. Among the findings of the physical examination, a possible characteristic of the affected region is the "mossy leg" aspect[8].

ESFA is divided into five types by Starink[34] and French[35], according to morphology, number of lesions and associated factors[4]. 1. The solitary sub type is the most common[16], represented by the appearance of a verrucous mass or single non-hereditary nodule located on the lower limbs of elderly patients[26]. 2. Multiple ESFA without associated cutaneous findings (eccrine syringofibroadenomatosis) are non-familial palmoplantar lesions without significant associated cutaneous findings. 3. Multiple ESFA with hidrotic ectodermal dysplasia (HED) presents in two different variants, Schöpf-Schulz-Passarge syndrome and Clouston's syndrome caused by mutations in WNT10A and GJB6 respectively[36,27,37]. 4. Nevoid ESFA (non-familial unilateral linear ESFA) is a rare genetic mosaicism, producing diffuse plantar hyperkeratosis. It may be the result of a somatic mutation in the early embryonic stage. 5. Reactive ESFA associated with inflammatory or neoplastic dermatosis such as chronic eczema, infections (e.g., viral warts, tuberculosis verrucosa cutis, atypical mycobacterium, chromoblastomycosis), inflammatory psoriasis or persistent local infection, erosive palmoplantar lichen planus[38], bullous pemphigoid[14], burn scar[39], diabetes mellitus with polyneuropathy and chronic ulcers[40], hidrotic ectodermal dysplasia, squamous cell carcinoma[30], epidermolysis bullosa, lepromatous leprosy, lymphedema (elephantiasis), secondary to primary cutaneous amyloidosis, venous stasis, nevus sebaceous, Bowen's disease , clear cell acanthoma[27,37,41,28,42,43] etc. Apart from these five types, another clear cell variant was also reported by Hu et al. in 2005[44]. A specific type of eccrine remodelling or ductal repair, due to repeated damage to eccrine structures, is believed to be the pathogenesis[4,26]. Our case falls into the fifth group. Indeed, the ulcer is the site of constant tissue remodelling and repair which can lead to epithelial changes and eccrine ductal proliferation, thus resulting in ESFA. The clinical appearance in this case suggested the possibility of squamous cell carcinoma. The clinical course of ESFA is mostly benign. However, the co-existence of ESFA and squamous cell carcinoma has been reported[30,11]. It is unclear whether ESFA develops in response to squamous cell carcinoma or squamous cell carcinoma represents a malignant degeneration of ESFA. The malignant degeneration of ESFA has been termed as eccrine syringofibrocarcinoma (ESFAC)[11,45,12]. The mechanisms of underlying malignant transformation remain unclear. Recurrences and metastatic disease have not been reported[30].

Clinically, pyogenic granuloma and verruca vulgaris should be taken into

consideration for differential diagnosis. The former usually has a history of trauma, which is characterized by red or dark-red papules and nodules, and the latter has a rough or verrucous surface. Both of them can be distinguished easily according to the histopathological examination. The histopathological differential diagnosis may include acrosyringeal nevus, fibroepithelial tumor of Pinkus(variety of BCC), eccrine poroma, clear cell acanthoma, pseudoepitheliomatous hyperplasia, artifacts of histologic processing, reticulated seborrheic keratosis and syringofibroadenocarcinoma. Plasma cell infiltrates, and strong PAS positivity is characteristic of acrosyringeal nevi. Fibroepithelial tumor of Pinkus shows focal changes typical of basal cell carcinoma with peripheral palisading and clefting artefact and loose fibrous stroma[12]. Eccrine poromas show more uniform proliferation of small epithelial cells with vertical thick strands/coloumns stretching into the dermis. In syringofibroadenocarcinoma there is an area of transformation where a malignant phenotype emerges displaying cytological atypia.

Treatment of ESFA depends on the number, location and resectability of the lesions[2,17,46]. Treatment of choice preventing malignant degeneration 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[15,41,28]. Trauner et al reported a patient with ESFA successfully treated with a dual pulse width flashlamp pumped pulsed dye laser[47].

CONCLUSION

The aim of this paper was to report a characteristic presentation of reactive ESFA in the context of a chronic leg ulcer. It is an infrequent pathology with a complete benign course. The suggested pathogenesis of this tumor includes repeated eccrine duct trauma resulting in eccrine duct remodelling and repair. Knowledge about the disease and diagnostic possibilities, considering the clinical presentation, helps to guide investigation and treatment more appropriately. All dermatologists should be familiar with the histopathological pattern of ESFA, so that the diagnosis of ESFA is not missed. Solitary lesions are cured by complete excision, while the treatment of multiple lesions is dependent on the size and location of the tumor. Close observation and follow-up may be an alternative to early excision, especially when complete excision is difficult due to involvement of larger areas. Since ulcers are common cutaneous findings, physicians should consider ESFA and its malignant potential in the differential diagnosis of similar lesions developing from a skin ulcer.

Declaration of Patient Consent: The authors certify that they have obtained all appropriate patient consent wherein the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

Authors Contribution: The authors have contributed in conceptualization, drafting and editing of the manuscript; collection, analysis and interpretation of data; intellectual participation in the studied case; critical review of the literature and approval of the final version of the manuscript.

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