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Case Report

An unusual presentation and a new treatment of eccrine syringofibroadenoma

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Abstract Eccrine syringofibroadenoma (ES) is a rare disease that clinically ranges from nodule on extremity to diffuse forms. A case of ES was assessed and compared with classification of ES. Our case had diffuse verrucous lesions since infancy, hypotrichosis, and loss of teeth. She had coincidental bullous pemphigoid. The patient was successfully treated with etretinate.

Key words
Eccrine syringofibroadenoma, bullous pemphigoid, etretinate.

Introduction

Eccrine syringofibroadenoma (ES) was first described by Mascaro in 1963 as a nodular lesion on the extremity. Since its original description it has become apparent that there are several different variants of this process, some of which are extensive. These more diffuse forms have been referred to as eccrine syringofibroadenomatosis, acrosyringeal naevous, eccrine poromatosis and linear eccrine poroma.[1]

All forms of ES are relatively rare, with 41 reported cases so far. Although it may be present coincidentally, it may be associated with hidrotic ectodermal dysplasia, characteristic histology includes anastomosing strands and cords of basaloid cuboidal epithelial cells that contain ducts. These extend from the epidermis and are surrounded by a rich fibrovascular stroma. It may be associated with bullous pemphigoid (BP).[1] We present one case of ES and BP with diffuse lesions that had eluded diagnosis for many years.

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Case report

A 40-year-old woman presented with exudative lesions since infancy on both extremities that biopsy was done several times but without definite diagnosis. Due to recurrent gingivitis, she had lost all her teeth by the age of 20. There was no perspiration dysfunction. Since several months ago she presented with pruritus and urticarial lesions. On examination there were absent teeth, racquet nail in left thumb, hypotrichosis, multiple hypertrophic verrucous brownish papules, plaques, nodules on both extremities and some on trunk (**Figures 1** and **2**) and hypertrophic verrucous plaques on soles (**Figure 3**), there were eczematous and urticaial lesions on face , trunk and proximal parts of lower extremity with multiple tense bulla overlaying (**Figure 4**).

Histopathology revealed thin anastomosing epithelial cords and strands forming a lattice

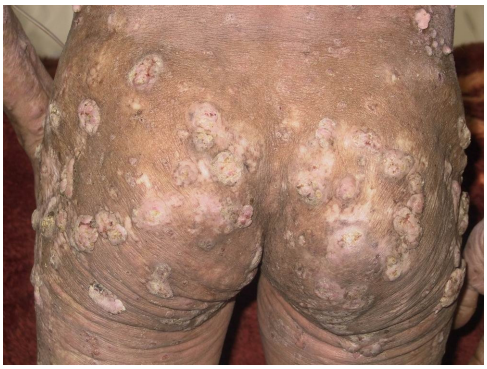

Figure 1 Verrucous plaques on buttock.

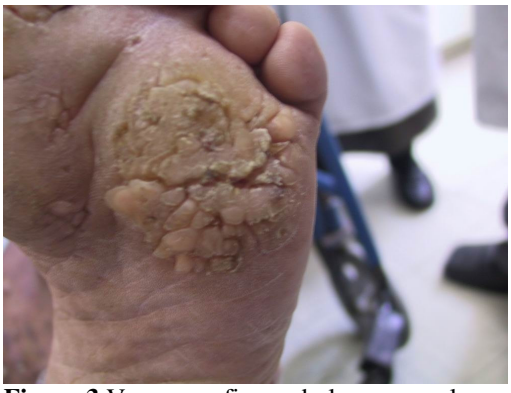

Figure 3 Verrucous fissured plaque on sole.

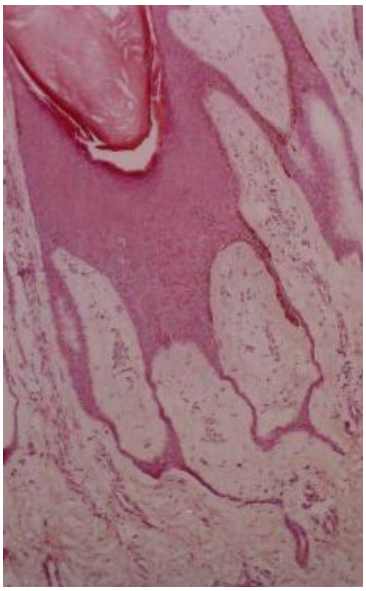

Figure 5:Anastomosing thin epithelial cords with a fibrovascular core connected to the epidermis (H&E, x100).

connected to the undersurface of epidermis (**Figure 5**). The cells were smaller and more basophilic than the epidermal keratinocytes.

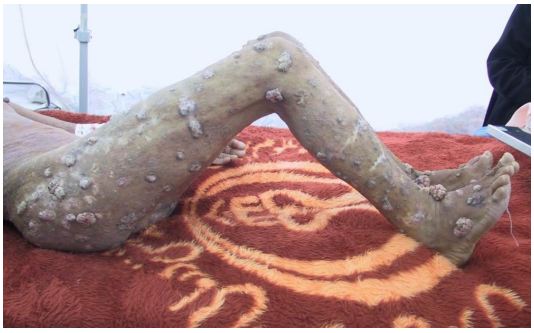

Figure 2 Lesions on lower extremity.

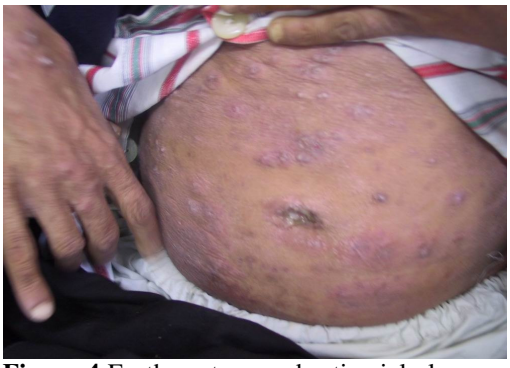

Figure 4 Erythematous and urticarial plaques on abdomen

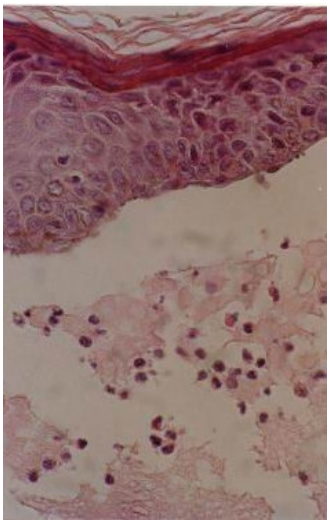

Figure 6 Subepidermal bulla containing some eosinophils, lymphocytes fibrin strands (H&E, x400).

Few ducts were present within the strands. Between strands, there was a rich fibrovasular stroma with mild lymphoplasma cell infiltrate. Eccrine glands were increased. The cells within strands were weakly PAS positive. There were lesions with histological features of

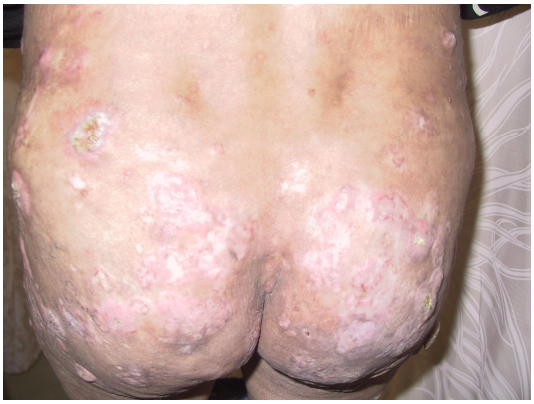

Figure 7

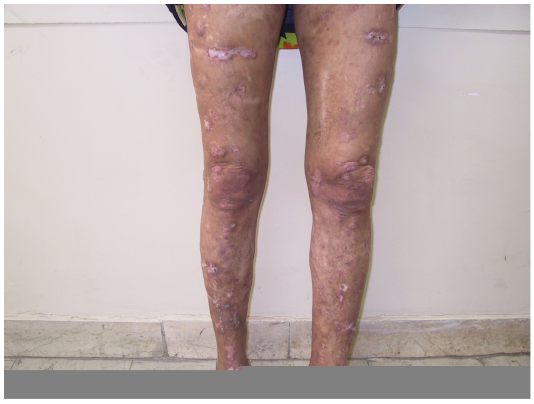

Figure 8 Substantial improvement 5 months after etretinate therapy.

epidermal nevus and keratoacanthoma. The microscopic findings were severe hyperkeratosis, papilomatosis, and acanthosis with elongation of rete ridges and mild basal hyperpigmentation resembling seborrheic keratosis. There was focal invaginating mass of kertinizing, well-differentiated squamous cells with eosinophilic glossy appearance. Islands of squamous cells had keratin at the center. There was subepidermal bulla containing eosinophils, lymphocytes and fibrin strands (**Figure 6**). Direct immunofluorescence showed IgG and C3 at basement membrane zone.

Considering the histopathological features suggestive of keratoacanthoma, etretinate 25 mg daily was started. After one month, the lesions became better therefore the treatment continued. After 5 months almost lesion healed except very verrocous ones (**Figures 7** and **8**).

Discussion

ES is a distinct neoplasm of acrosyringeal cells related to two well recognized and readily distinguished neoplasm of the intraepidermal sweat duct unit, eccrine poroma and hidracanthoma simplex. ES should be distinguished from fribroepithelioma of Pinkus with focal spread of tumor down enclosed eccrine sweat ducts and from tumors of follicular infandibulum that occasionally may show preexistent sweat duct that enter the lesion.[2] Immunohistological studies by Mehregan *et al*. suggested that it differentiates forwards the acrosyringium. Mehregan showed that ES was positive for antieccrine gland monoclonal antibody (EKH6),[3] but because EKH6 is not specific for the acrosyingium their immunohistochemical study dose not provide incontrovertible evidence that this tumor originates from the acrosyringium. Comparison between the staining pattern of the tumor and that of normal eccrine glands with regard to other antikeratin antibodies showed that the tumor expressed an immunophenotype similar to that of intradermal eccrine duct.[4]

ES, according to the clinical presentation, has been classified into different forms. 1) Solitary ES: is typically a nonhereditary verrucous growth of relatively trivial importance in middle aged and elderly patients. 2) Multiple ES in the Shöpf syndrome, the patients typically have multiple erythematous papules in a mosaic pattern and a rough surface on the palms and

soles. Lesions appear between age 15 and 25 years. Other cutaneous manifestation are hydrocystoma, hypotrichosis, hypodontia, hypoplasia of nails. 3) Multiple ES without associated cutaneous findings: most of these patients have familial palmoplantar lesion only and no significant associated cutaneous findings. 4) Non-familial unilateral linear ES: this rare form probably represents a genetic mosaic caused by a postsomatic mutation at an early embryonal stage.[2] Another subtype of ES is reactive ES associated with inflammatory or neoplastic dermatoses. This type was associated with BP, lichen planus, long standing leg ulcer and stasis dermatitis. It is noteworthy that more than 60% of reactive ES occurred on the lower legs or feet in association with skin ulcer or inflammation caused by impaired circulation blood.[5] One case report revealed 2 patients that developed palmoplantar erythema following resolution of BP. Clinical manifestations ranged from prominent well-demarcated erythematous areas with focal erosions and fissures to mild erythema. The authors speculated that these lesions resulted form the underlying inflammatory process in BP.[6]

In our case distinct clinical manifestation, earlier presentation and association with other cutaneous manifestation reveal that this case may be another presentation of ES that does not fall in any subtype of present classification. In this case association with BP may be coincidental and inflammatory process in BP did not prepare the ground for ES. We propose etretinate for treatment of ES, as our patient showed good response to this therapy. The speculative mechanism for this may be modulation of the growth of intradermal eccrine duct but more experience in therapy and laboratory findings is needed to define the precise mechanism of action of etretinate.

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