Case Report

Eccrine syringofibroadenoma: an unusual presentation of a rare disease

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Abstract

Eccrine syringofibroadenoma (ESFA) is a rare disorder with differentiation towards ductal eccrine apparatus. There are a variety of clinical manifestations and distribution of lesions but the histopathological features are remarkably similar with benign appearing anastomosing cords of eccrine ductal epithelial cells with or without lumina formation, are embedded in loose fibrovascular stroma. We report a case of 55-year-old Muslim man having large pale pink slightly verrucous plaque on right upper back since last 20 years with central ulceration for 8 years. Punch biopsy was done and showed features of ESFA.

Key words

Eccrine syringofibroadenoma.

Introduction

Eccrine syringofibroadenoma (ESFA) is a rare benign proliferation with differentiation towards ductal eccrine structure and was first reported by Mascaro in 1963. Affected patients are typically older adults with most presenting in their seventh and eighth decades. The clinical manifestations are variable and range from solitary lesion to multiple papules and nodules arranged in a linear fashion. 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. In contrast, the histological features are remarkably similar and are benign appearing anastomosing cords of eccrine epithelial cells with or without formation of lumina, embedded in loose fibrovascular stroma. Clinical course of ESFA is typically benign in nature. However, recent reports suggest the possibility of an association of ESFA with squamous cell carcinoma or malignant transformation of ESFA.

Case report

A 55-year-old Muslim man presented to dermatology OPD, Midnapore Medical College with a 14cmx12cm pale pink plaque almost annular in shape on right upper back since last 20 years. The central part of the plaque was ulcerated, slightly verrucous and exophytic with granulation tissue. The periphery of the lesion was pale to pinkish in color. There was induration and scarring in some places (Figure 1). The lesion was very small (peanut size) to begin with and increased slowly to attain the present size. Initially overlying skin was intact and subsequently became ulcerated with exophytic granulations for last 8 years. There were history of mild pain, itching and irritation. There was no history suggestive of reactive pattern of the disease manifestation. Patient did not suffer from any other disease.

Biopsies from the margin of the lesion and from central ulcerated growth were taken. Histopathological examination of the central lesion showed necrotic and granulation tissue.
The specimen from the margin of the lesion showed downgrowth of the eccrine epithelial cells arranged in interanastomosing cords embedded in loose fibrous stroma. Higher power microscopy examination found the benign appearing cuboidal eccrine epithelial cells and focal ductal differentiation (Figure 2, 3 and 4).

The patient was referred to surgeon for further management.

Discussion

ESFA is a rare cutaneous adnexal tumor which was first described by Mascaro 1963.1 Since that time numerous reports have detailed the entity’s clinical, histological, immunophenotypic and ultrastructural features.2,6,7,8 The consensus belief is that ESFA derives from or differentiates towards the acrosyringium or eccrine dermal duct.9 It is still unclear whether the lesion is neoplastic, hamartomatous or reactive in nature.10 The clinical appearance is diverse yet the histological features are remarkably similar. Until recently, ESFA was considered to have an entirely benign clinical course, however, recent reports have demonstrated an association of ESFA with squamous cell carcinoma or malignant transformation of ESFA.
The histological features of ESFA are diagnostic and include multiple thin anastomosing cords and strands of benign appearing epithelial cells which are connected to the undersurface of the epidermis and with or without formation of lumina which is embedded in a loose fibrovascular stroma.

Immunohistochemical studies show that the lesion cells are positive for keratin 6, keratin 19 and filaggrin. Electron microscopic studies show that the ducts have a keratinization pattern similar to acrosyringeal cells.

In the literature reviewed, most of case reports of ESFA are associated with reactive conditions like burn scar ulcer, bullous pemphigoid, erosive palmoplantar lichen planus and peristomal skin or associated with numerous conditions like nevus sebaceous, hidrotic ectodermal dysplasia, and diabetic polyneuropathy. But in our patient, there was no history of diabetes mellitus or any other reactive condition. Hence, ESFA may probably arise as a benign neoplastic proliferation of acrosyringeal cells.

The treatment of ESFA depends on the number, location and resectability of lesion. Solitary ESFA is cured complete by excision. In cases that are too large to excise in toto, generous sampling to rule out malignant transformation is suggested.

References