Eccrine syringo fibro adenoma: an unusual presentation of a rare tumour
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Abstract: Eccrine Syringo fibro adenoma (ESFA) is a benign adnexal tumour thought to be of eccrine origin also named syringo fibro adenoma of Mascaro, after the man who first described this tumour. It typically occurs as a solitary nodule on the extremities of an elderly person. Clinically, solitary ESFA is typically a nonhereditary verrucous growth. Histological findings typically show a proliferation of anastomosing strands, cords, and columns of monomorphous epithelial cells that harbour eccrine duct formations embedded in a fibro vascular stroma. We report a case of 19 year old male who presented with a cauliflower growth over left foot measuring 11x11x3cm with lymphoedema and past history of trauma and thorn pricks. Clinical diagnosis was kept as lymphoedema with chromoblastomycosis. Incisional biopsy report was inconclusive. So wide local excision of the mass was done and final diagnosis of ESFA with lymphoedema was offered. In the reviewed literature, only 80 cases have been reported till now. But cauliflower like growth has not been reported in the gross morphology in any of the cases. So considering cauliflower like growth, this case is a very rare presentation of ESFA. Thus this case is extremely rare in gross morphology as well as clinical presentation.

Keywords: Eccrine syringo fibro adenoma, benign adnexal tumour

INTRODUCTION
Eccrine Syringo fibro adenoma (ESFA) is a rare, benign tumor of eccrine sweat gland origin that usually presents as a nodule on the extremities. It can also present as an ulcerative plaque, verrucous lesion, popular or nodular lesion or as palmoplantar keratoderma. ESFA was first reported by Mascaro in 1963 [1]. Site of predilection is the extremity although it can affect the face, back, abdomen, buttocks and rarely the nails [2]. In contrast, the histologic features are remarkably similar and are characterized by multiple anastomosing strands, cords, and columns of monomorphous epithelial cells that harbour eccrine duct formations embedded in a fibro vascular stroma. The histogenesis of this entity is controversial. Many authors suggest an acrosyringeal differentiation, though eccrine duct origin has been suggested based on cytokeratin expression. The clinical course of ESFA is typically benign [3]. However, recent reports suggest the possibility of an association of ESFA with squamous cell carcinoma or malignant transformation of ESFA [4, 5].

CASE REPORT
A 19 year old male presented to the dermatology outpatient department with a cauliflower growth over left foot measuring 11x11x3cm since 2 years which was small to start with and gradually increased in size to attain the present size. He complained of swelling and pain over left leg and foot since 4 yrs. The patient also had history of itching off and on. He had past history of trauma to left leg in road traffic accident with fractures of femur and tibia and crush injury. He also gave history of multiple thorn pricks to left foot in the past. Clinical diagnosis was kept as lymphoedema with chromoblastomycosis. Radiological findings were suggestive of lymphoedema with decreased joint space of left foot. Incisional biopsy report was inconclusive. So wide local excision of the mass was done and final diagnosis of ESFA with lymphoedema was offered. The histopathological examination showed growth 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).
DISCUSSION

ESFA is an uncommon tumor of eccrine glands that was first described by Mascaro in 1963 [1]. ESFA usually manifests as a solitary nodule on the...
extremities of an elderly person. Other sites of occurrence include the face, trunk, and rarely the nails. Clinical findings are variable, ranging from solitary nodules to multiple papules, nodules, and plaques. Starink [6] classified ESFA into four clinical subtypes: 1) multiple ESFA associated with hidrotic ectodermal dysplasia, 2) multiple ESFA without associated cutaneous features, 3) unilateral linear ESFA, 4) solitary ESFA, and French subsequently proposed the fifth subtype, reactive ESFA. It is still unclear whether the lesion is neoplastic, hamartomatous or reactive in nature [7]. 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 fibro vascular stroma [2, 8, 9, 10]. Immunohistochemical studies show that the lesion cells are positive for keratin 6, keratin19 and filaggrin [7]. Electron microscopic studies show that the ducts have a keratinization pattern similar to acrosyringeal cells. The histologic differential diagnosis includes fibroepithelial tumor of Pinkus, tumor of the follicular infundibulum, pseudo epithelio matous hyperplasia, papillary eccrine adenoma, reticulated seborrheic keratosis, squamous cell carcinoma, and artifacts of histologic processing [11]. ESFA may occur as a reactive proliferation associated with other conditions like ulcer of skin, chronic lymphoedema, venous stasis, nail trauma, burn scar, nevus sebaceous, bullous pemphigoid, and epidermolysis bullosa [12, 13]. The suggested pathogenesis of reactive ESFA includes repeated eccrine duct trauma resulting in eccrine duct remodeling and repair. In the literature reviewed, most of the case reports of ESFA are associated with reactive conditions; as in our patient, there was history of thorn prick and lymphoedema. The treatment of ESFA depends on the number, location, and resectability of lesions. Solitary ESFA is cured by complete excision [2]. In cases that are too large to excise in toto, generous sampling to rule out malignant transformation is suggested.

REFERENCES:


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