

Case Report

Eccrine Syringofibroadenoma- A Very Rare Entity in Dermatopathology: Case Report

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Abstract: Eccrine syringofibroadenoma is a very rare tumor in dermatopathology. It was first described by Mascaro in 1963. Patients usually present with solitary, hyperkeratotic, nodular plaque, which may be several centimetres in diameter, on external surface of the body. Hereby we report a case of 41 years male patient presented with a scalp swelling of 9 months duration which was measuring about 2.5X1.5X1cm and a clinical diagnosis of 'Fibroma' was made and excision biopsy was performed, histopathologically diagnosed as 'Eccrine Syringofibroadenoma' based on the criteria proposed in Lever's histopathology of skin. It is a rare benign eccrine tumor with anastomosing strands and fibrovascular myxoid stroma, but rarity of the tumor favours documentation in literature.

Keywords: Eccrine syringofibroadenoma, Fibroma, Acrosyringal cells

INTRODUCTION

Eccrine syringofibroadenoma is a very rare tumor in dermatopathology [1]. It was first described by Mascaro in 1963 [2]. Patients usually presents with solitary, hyperkeratotic, nodular plaque [3], which may be several centimetres in diameter [4].

Multiple lesions of syringofibroadenoma may be referred as syringofibroadenomatosis [5]. Other name of this lesion is acrosyringal adenomatosis [6]. The most common clinical presentation is solitary, often verrucous papules or nodules. Rarely it may present as disseminated lesions. Eccrine syringofibroadenomatosis has been associated with some other entities like inflammatory and neoplastic conditions including bullous pemphigoid, lichen planus, ulcers, squamous cell carcinoma, sebaceous naevus, and chronic lymphoedema. On the basis of the presence of fibrous stroma, some authors consider syringofibroadenoma as a hyperplasia rather than a neoplasia. It may be associated with inherited palmo-plantar keratodermas including hidrotic ectodermal dysplasia known as Schöpf- Schultz-Passarge syndrome an autosomal dominant syndrome, whose genetic aberration has been localized to chromosome 13q [7].

CASE REPORT

We reported a case of 41 years male patient presented with a scalp swelling of 9 months duration which was measuring about 3X1.5X1cm and a clinical diagnosis of 'Fibroma' was made and excision biopsy

was performed, histopathologically diagnosed as 'Eccrine Syringofibroadenoma'. There is no other similar lesion elsewhere in the body or any of the family members. There is no history of systemic diseases like diabetes mellitus. Cutaneous examination revealed single, nodular, flesh-colored nodule associated with crusting. On gross examinations single greyish white firm tissue mass measuring 2.5 X 1.5 X 1 cm, cut section shows grey white irregular areas of tumor mass (Fig. 1).



Fig. 1: Grey white tumor mass with area of myxoid changes

Multiple sections studied show anastomosing epithelial cords of acrosyringal cells embedded in fibrovascular myxoid stroma. Cords of acrosyringal cells at places show formation of lumina and some clear

cell changes are observed in epithelial cords. Stroma shows mild to moderate infiltration of lymphocytes and plasma cells (Fig. 2, 3).

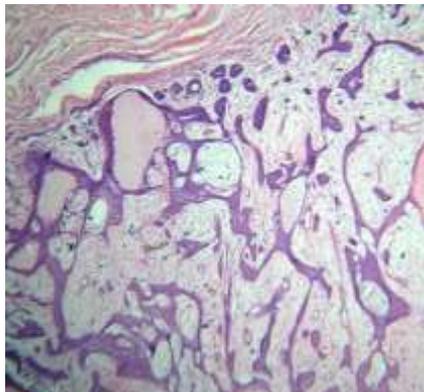


Fig. 2: Show anastomosing epithelial cords of acrosyringal cells embedded in fibrovascular myxoid stroma (H & E X 100)

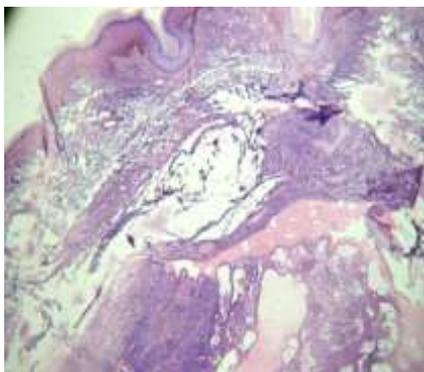


Fig. 3: Show epidermis, acrosyringal cells & stroma with lymphocyte, plasma cell infiltration (H & E X 40)

DISCUSSION

Eccrine syringofibroadenoma is an uncommon tumor of eccrine glands that was first described by Mascaro in 1963 [1, 2]. Eccrine syringofibroadenoma usually manifests as a solitary nodule on the extremities of elderly [3, 8]. Other sites include the face, trunk and rarely the nails. Clinical findings have been reported to be variable. It ranges from solitary nodules to multiple papules, nodules, and plaques [2, 8, 9].

Starink classified eccrine syringofibroadenoma into four clinical subtypes: (a) multiple eccrine syringofibroadenoma associated with hidrotic ectodermal dysplasia, (b) multiple eccrine syringofibroadenoma without associated cutaneous features, (c) unilateral linear eccrine syringofibroadenoma (d) solitary eccrine syringofibroadenoma, and French subsequently proposed the fifth subtype, reactive eccrine syringofibroadenoma [10].

The diagnosis is performed by its characteristic histopathological features. The histopathological

features typically exhibit proliferation of anastomosing strands and cords of monomorphous epithelial cells in reticular pattern with eccrine duct formations embedded in a fibrovascular myxoid stroma. The differential diagnosis includes eccrine poroma, acrosyringal nevus, syringofibroepithelial carcinoma, fibroepithelial tumor of Pinkus (Variety of BCC), tumor of the follicular infundibulum, papillary eccrine adenoma, pseudoepitheliomatous hyperplasia, squamous cell carcinoma, reticulated seborrheic keratosis and artifacts of histologic processing [8, 11].

In case of acrosyringal nevus strong PAS positivity and plasma cell infiltrate are the distinguishing features, whereas in eccrine poroma, tumour 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 typical of basal cell carcinoma with peripheral palisading and clefting artefact and loose fibrous stroma [12].

CONCLUSION

Eccrine syringofibroadenoma has a complete benign course and rarely associated with chronic ulcer. The suggested pathogenesis of this tumor includes repeated eccrine duct trauma resulting in eccrine duct remodelling and repair. Solitary lesions are cured by complete excision, while the treatment of multiple lesions is dependent on the size and location of the tumor. There is a focal area of squamous metaplasia or malignant transformation like squamous cell carcinoma is also reported. However, close observation and follow-up may be an alternative to early excision, especially when complete excision is difficult due to involvement of larger areas.

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