



Case Report

Syringocystadenoma papilliferum of eyelid – A deviant case report

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Abstract

Syringocystadenoma papilliferum, a distinct dermatologic entity, is an exceedingly rare benign skin neoplasm and seems to have arisen from apocrine or eccrine adnexal structures. The diagnosis is clinically suspected and histologically confirmed due to its non-distinct clinical findings and varied presentations. Since it usually appears at birth or during puberty and adolescence, it is called as Childhood tumor. Complete surgical excision is the treatment of choice followed by detailed histologic examination, to avoid the risk of malignant degeneration. Herein, we have reported a rare case of Syringocystadenoma papilliferum of eyelid in an adolescent female.

Key words

Syringocystadenoma papilliferum, Eyelid, Childhood tumor.

Introduction

Syringocystadenoma is a benign hamartomatous adnexal tumor, the histogenesis of which is still unclear [1]. It has been theorized that pluripotent cells of apocrine lineage give rise to Syringocystadenoma papilliferum [1, 2]. This rare tumor may occur de-novo or within a nevus sebaceous of Jadassohn and presents commonly as a solitary lesion. Clinically, most of the lesions have non-specific features and are first noted at birth in 50% of the patients, while the remainder of 15-30% present later during adolescence and adulthood [3].

Scalp, face and neck are the common sites although rarely these tumours are found on the trunk, chest, breast, axillae, genitalia, arm, vulva, vagina, thigh, ear canal and eyelid. Syringocystadenoma papilliferum can rarely affect the eyelid skin [4] with very few documented cases in the literature.

Case report

19 year old female was presented with a well defined growth in the left upper eyelid since four years, which was gradually progressive in size during the past one year. On local examination, it was a well defined,

erythematous nodular lesion measuring 0.5x0.5 cm and a clinical diagnosis of pyogenic granuloma was considered followed by excisional biopsy for histopathological examination. Ophthalmic examination and systemic examination of the subject was insignificant.

On gross examination, a solitary lesion with 1.0x0.5 cm dimensions was noted which was firm and fleshy. Histopathological examination demonstrated a well circumscribed lesion which showed a papillary projection covered with squamous epithelium and ductal invaginations into the deeper portions of the lesion was noted. **(Photo – 1)** These ductal structures were lined by an inner columnar epithelium demonstrating luminal apical decapitation secretions, and an outer layer of small cuboidal cells. Underlying the papillary epithelium, the fibrovascular dermal tissue exhibited prominent plasma cell rich infiltrate with occasional lymphocytes and eosinophils. **(Photo – 2)** With the above classic histologic findings, a diagnosis of Syringocystadenoma papilliferum was arrived at.

Photo - 1: Microphotograph showing a well circumscribed lesion with cystic spaces and papillary projections in the papillary dermis. (100X)

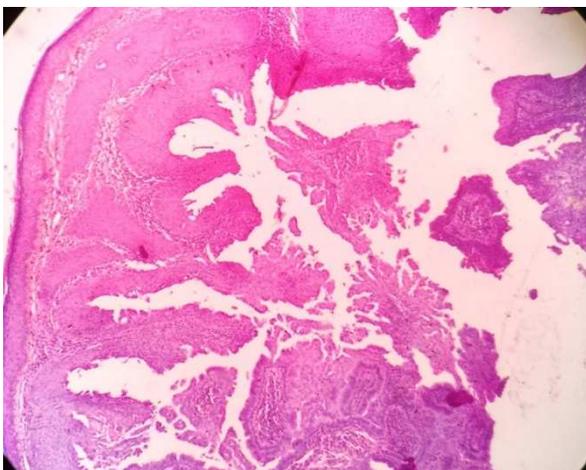
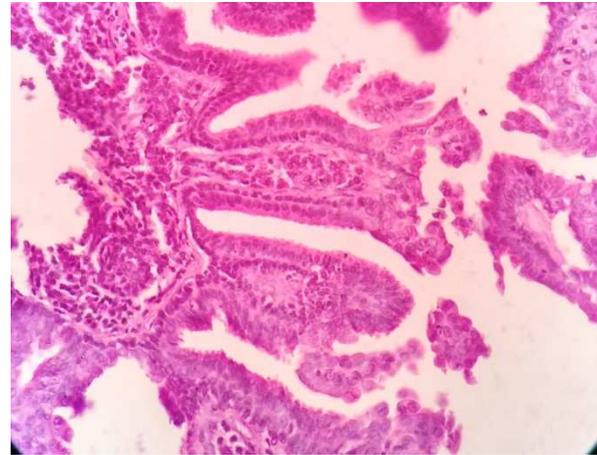


Photo - 2: Microphotograph showing papillary projections lined by outer cuboidal cells and inner columnar cells with luminal decapitation secretions. (400X)



Discussion

Skin, the largest organ of the body [5] is commonly affected by a multitude of neoplastic, congenital and developmental lesions and aberrations, some of which are so rare to be mentioned in aeons of medical literature. Syringocystadenoma papilliferum, an unusual skin tumour was first described by Stokes [6] in 1917 termed as “Nevus Syringocystadenoma papilliferum”.

Clinically, most of the cases of Syringocystadenoma papilliferum are first noted at birth and other cases develop in infancy, childhood and adolescence. The lesion presents as an erythematous or brownish papule, nodule or tumour of varying sizes, usually between 1 to 4 cm [7] with flat, smooth, verrucous or papillomatous surface and is more common in females [8]. Most patients of Syringocystadenoma papilliferum present with a solitary lesion, however multiple papules arranged in linear pattern are usually associated with nevus sebaceous.

Clinical differential diagnosis for syringocystadenoma papilliferum includes viral



warts, verrucous carcinoma, pyogenic granuloma, tuberculosis verrucous cutis, subcutaneous fungal infection and giant lymphangioma. The differential diagnosis for linear lesions includes nevus comedonicus, linear epidermal nevus, eccrine nevus, cylindroma and basaloid follicular hamartoma.

Fugita, et al. [1] noted that Syringocystadenoma papilliferum is most commonly associated with nevus sebaceous followed by basal cell epithelioma, sebaceous epithelioma, apocrine hydrocystoma, trichoepithelioma, and eccrine spiradenoma. However, association of Syringocystadenoma papilliferum with many other lesions has also been noted [1].

Eyelid is a very rare site for the occurrence of Syringocystadenoma papilliferum [9]. Yap FB, et al., in 2010 reviewed for cases of Syringocystadenoma papilliferum outside the head and neck region in the English literature, only 69 cases were with such characteristics and none was reported on the eyelid [1]. In a study by Barbarino et al in 2009, fourteen cases with Syringocystadenoma papilliferum of the eyelid were identified [9]. Helmi A, et al. in their study in 2011, reported three cases of arising in the eyelid [10]. To date, there have been only seventeen cases of Syringocystadenoma papilliferum reported on this rare location.

The histogenesis of Syringocystadenoma papilliferum is controversial. It was suggested by Harkey and Helming that, the lesion is either derived from the glandular ducts intermediate between apocrine and eccrine or an adenoma of eccrine ductal origin [1]. According to Lever, it is a hamartoma derived from pluripotent cells [2]. Pinkus postulated that papillomatous area may have been derived from apocrine or eccrine glands and hamartomatous proliferation of the involved skin would give rise to the adenomatous component of the lesion [1].

Rapid enlargement or ulceration over the growth is indicative of a malignant transformation and is termed Syringocystadenocarcinoma papilliferum. Up to 10% cases of Syringocystadenoma papilliferum show transformation into basal cell carcinoma [7]. However, no such malignant transformation or association with malignant neoplasm of eyelid has been reported [9].

Conclusion

In conclusion, we have added to the literature, another case of Syringocystadenoma papilliferum of upper eyelid, which is frequently misdiagnosed clinically and such a presentation may generate multitude of differential diagnoses, hence it must be sent for histopathological examination.

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