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Syringocystadenoma Papilliferum of the Upper Eyelid: A Rare Case Report

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Abstract

Syringocystadenoma papilliferum is a benign adnexal skin tumour of the apocrine or the eccrine type with characteristic histological features and varied and non-distinct clinical findings. It is relatively a rare neoplasm, which is called as a childhood tumour, since it usually appears at birth or during puberty. Most of the tumors are located in the scalp and face that presents as multiple warty papules. Our case is syringocystadenoma papilliferum of the upper lid and medial canthal region of left eye in an adult female, histologically confirmed.

Keywords: Adnexal tumor, nevus sebaceous, syringocystadenomapapilliferum

Introduction

Syringocystadenoma papilliferum is a rare benign hamartomatous adnexal tumour which originates from the apocrine or the eccrine sweat glands. It is relatively a rare neoplasm, predominantly a childhood tumour. In about 50% of those who are affected, present at birth, and further 15%-30%, the tumour develops before puberty [1]. The lesion of syringocystadenoma papilliferum usually measures <4cms in diameter. The tumour has varied clinical presentations. The plaque type which presents as a hairless area on the scalp, is commonly associated with a sebaceous nevus of Jadassohn. Appearance of the lesion in the face and neck region is seen in the linear type and the solitary nodular type shows predilection for the trunk [2].

A presentation with multiple lesions is rare, and in those lesions which arise outside the head and neck region, it is even more uncommon. Syringocystadenoma papilliferum may occur de-novo or within a nevus sebaceous. It occasionally co-exists with other tumours such as basal cell carcinomas and verrucous carcinomas [3]. Histologically, there are characteristic duct-like structures covered by two layers of glandular epithelial cells,located in the upper dermis, with a varying degree of papillomatosis. An infiltrate of plasma cells is seen in the stroma, especially in the papillary projections[4]. Histopathology is confirmatory and immunohistochemistry further differentiates between apocrine or eccrine origin.

Case Report

A 65-year-old female presented with complaint of lesion on the upper lid and medial canthal region of the left eye since childhood and increasing in size for the past 4 months. There was history of itching present. Examination revealed a 1 x 1.5 cm lobulated swelling in the upper lid and medial canthal region of left eye, cystic in consistency and translucent in one area with skin involvement at the summit of the swelling about 0.5cm.The clinical diagnosis of skin adnexal tumor was made and was advised excision biopsy.

Fig. 1a - 1x 1.5 cm lobulated swelling in medial canthal region of left eye

Fig. 1b - Demonstration of translucency

Patient underwent excision Biopsy and histopathological examination revealed SyringocystadenomaPapilliferum.

Patient underwent excision Biopsy and histopathological examination revealed Syringocystadenoma Papilliferum.

Figure

Fig. 2a - Intraoperative picture

Fig. 2b – Specimen after excision

Gross histopathological examination showed single fragment of skin with underlying cyst measuring 1.2x1x0.7 cm ,filled with clear fluid.

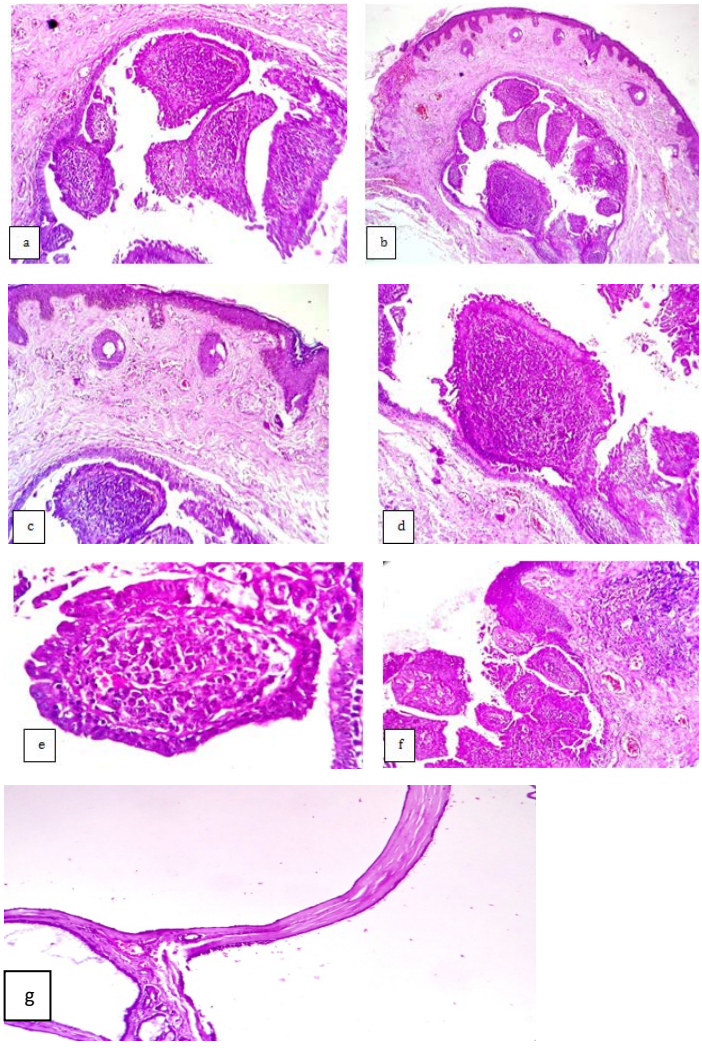

Fig. 3a - Keratinised stratified squamous epithelium of skin with adnexal structures and underlying cystic lesion. (H and E 40x)

Fig. 3b - Keratinised stratified squamous epithelium of skin with adnexal structures and underlying cystic lesion. (H and E 100x)

Fig. 3c - The underlying cystic lesion showing lining by apocrine cells and focally by stratified squamous epithelium, numerous papillary projections extending into the lumina lined by two layers of cells and the core of the papillae showing dense plasma cell infiltrates. (H and E 100x)

Fig. 3d and 3e - The papillary projections are lined by two layers of cells with oval nuclei and dense eosinophilic cytoplasm and the core of the papillae showing dense plasma cell infiltrates. (H and E 400x)

Fig. 3f - Areas showing the cystic lesion extending from the epidermis. (H and E 100x)

Fig. 3g - Adjacent areas showing cyst lined by a single layer of apocrine epithelium. (H and E 100x)

Fig. 3h - Adjacent areas showing cyst lined by a single layer of apocrine epithelium. (H and E 400x)

Discussion

Syringocystadenoma papilliferum is rare benign adnexal sweat gland neoplasm characterized by asymptomatic, skin colored to pink papules, or plaques of highly variable appearance. Most common sites are head and neck region; however, tumor in other areas, such as vulva, external ear, lower leg, and scrotum, have also been reported. [5] In our case, the lesion @ medial canthal region of left eye. It usually appears at birth or during infancy and around the time of puberty. In about one-third of cases, syringocystadenoma papilliferum is reportedly associated with nevus sebaceous. Multiple tumors of adnexal origin, such as trichoblastoma, apocrine adenoma, hidradenoma papilliferum, and trichilemmoma, are being reported to arise along with nevus sebaceous. [6] In this case, the tumor was neither present since birth nor associated with other tumors.

Three clinical types have been described

1. Plaque type: It presents as alopecic patch on the scalp that enlarges during puberty to become verrucous, nodular, or crusted plaques.
2. Linear type: It consists of multiple reddish, firm papules, or umbilicated nodules 1-10 mm in size.
3. Solitary nodular type: These are domed pedunculated nodules 5-10 mm in size and located near trunk shoulder and axilla. [5]

It reportedly evolves within three stages:

1. Infantile stage: It appears as alopecic orange yellow plaque.
2. Adolescent stage: Under androgenic influence the plaques undergoes various changes such as hyperkeratosis, hyperpigmentation, and sebaceous gland formation.
3. Adult stage: It is characterized by the presence of large sebaceous glands, ectopic apocrine glands, and epidermal hyperplasia. [7]

During the period of adult stage, variety of benign or malignant lesions can develop. [10] The malignant tumors reported in association with syringocystadenoma papilliferum are squamous cell carcinoma, basal cell carcinoma, and ductal carcinoma. Basal cell carcinoma is the most common symptom and have been reported in 10% of the cases while only two cases of squamous carcinoma have been reported. [9]

Till now, only two cases of verrucous carcinoma in conjunction with syringocystadenoma papilliferum, have been published [10]. Ductal carcinomas which arise from Syringocystadenoma papilliferum have been reported as well [11]. An ulceration or a rapid enlargement is indicative of a malignant transformation.

Syringocystadenocarcinoma papilliferum is a malignant counterpart of syringocystadenoma papilliferum [9]. The diagnosis is clinically suspected and histologically confirmed. Due to the risk of a malignant change, a prophylactic surgical excision, followed by a detailed histological examination, is the treatment of choice.

Approximately, one-third of the cases are said to arise in precursor lesions such as organoid nevi. Yamamoto *et al*. reported the origin of pluripotent cells on immunohistochemistry and ultrastructural studies by investigating the immunohistological patterns of 12 different anti-cytokeratin (CK) antibodies, and several other markers in five cases of this tumor concluded that tumor epithelium was composed of several cell types demonstrating various stages from the primitive clear cells to the basal cells that support the concept that SCAP is a hamartomatous tumor that arises from pluripotent cells. Boni *et al*. reported mutation of PTCH and p16. [5] Histopathology typically shows varying degrees of papillomatosis along with cystic invaginations and malformed sebaceous glands. Immunohistochemistry helps in differentiating the origin of the tumor, i.e. either eccrine or apocrine, but is of no clinical significance. Positive immunoreactivity for proteins 15 and 24 and zinc-2 glycoprotein demonstrates evidence of apocrine differentiation, while positivity for CKs demonstrates eccrine differentiation. [5]Immunohistochemistry was not done in the present case.

Smooth muscle actin (SMA) positivity is normally the feature of myoepithelial cells but syringocystadenoma papilliferum do not contain myoepithelial cells and hence the positivity indicates immaturity of the tumor. [8] The only treatment for syringocystadenoma papilliferum is excision biopsy that also helps in confirming the diagnosis. Other modalities rarely used for treatment are CO_2 laser therapy while excision and grafting are unfavorable. [5]

Conclusion

Syringocystadenoma papilliferum is an uncommon sweat gland tumor with a widely variable clinical appearance. The presentation of tumor may generate multiple differential diagnosis, thus it must be histologically confirmed. Surgical excision with reconstruction is the treatment of choice.

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