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Case Series

Pathology Section

Syringocystadenoma Papilliferum- A Case Series

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ABSTRACT

Syringocystadenoma papilliferum is also known as papillary syringadenoma. This is a warty tumour of the scalp, neck and face that occurs at any age. These lesions usually have an adjacent nevus sebaceous or adjacent basal cell carcinoma. Here, the authors present case series of eight cases, four male and four female patients, with varied presentations at different sites: scalp,upper eyelid, left arm and back, aged 11 years to 68 years, mean age being 45 years. Most of the cases had a clinical diagnosis of sebaceous horn and one was diagnosed as squamous cell carcinoma. All of them had a histopathological diagnosis of Syringocystadenoma Papilliferum.

Keywords: Apocrine, Eccrine, Nevus sebaceous, Contiguous squamous proliferations, Hidradenoma, Tubular apocrine adenoma

INTRODUCTION

Syringocystadenoma Papilliferum (SCAP) is a benign rare neoplasm that is present at birth or develops in childhood and is more common in the females [1,2,3]. It usually occurs on the head and neck, but it can also involve the chest, breast, axilla, genitalia, arm, thigh and eyelid [3]. The cell of origin for this tumour is apocrine sweat glands and rarely from eccrine glands [2]. The size of the lesion measures from 1 to 3 cm in diameter [4]. This tumour usually occurs as a solitary lesion and multiple lesions are rare. It may occur denovo or within a nevus sebaceous. The importance of this lesion is to rule out existing basal cell carcinoma and verrucous carcinoma [4]. This is a series of eight cases which was histologically diagnosed as syringocystadenoma papilliferum.

CASE SERIES

Case 1

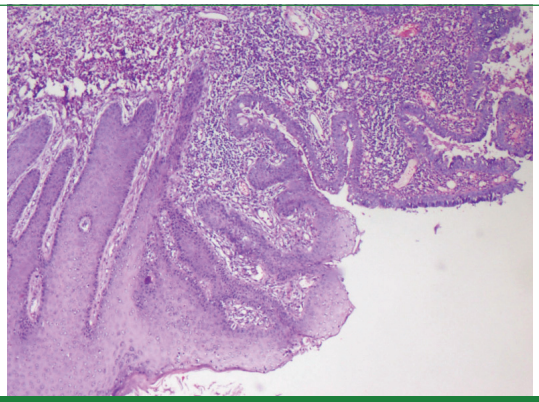
A 53-year-old male patient, presented with a 3-year duration of painless nodular growth measuring 2.1×1.8 cm over the mid-scalp associated with itching. Clinically, the case was diagnosed as sebaceous horn and excision was done. Histopathology showed hyperkeratotic squamous epithelium arranged in papillae [Table/Fig-1]. The underlying dermis showed adnexal structures with infiltrating cystically dilated apocrine glands lined by bilayered epithelium with amphophilic secretions. Background dense plasma cell infiltrate is seen. The final diagnosis was SCAP.

Case numbers	Age	Sex	Site	Histopathological diagnosis
1	53	M	Scalp	Syringocystadenoma papilliferum with nevus sebaceous
2	38	M	Scalp	Syringocystadenoma papilliferum
3	62	F	Upper eyelid	Syringocystadenoma papilliferum
4	50	M	Scalp	Syringocystadenoma papilliferum
5	53	F	Left arm	Syringocystadenoma papilliferum
6	68	M	Scalp	Syringocystadenoma papilliferum
7	22	F	Back	Syringocystadenoma papilliferum
8	11	F	Scalp	Syringocystadenoma papilliferum

[Table/Fig-1]: Clinical presentation and histopathological diagnosis.

Case 2

A 38-year-old male patient, presented with a 4-year duration of an erythematous papule with a smooth surface, measuring 1.8×1.3 cm over the scalp [Table/Fig-1]. Clinical diagnosis was given as sebaceous horn and excision was done. Histopathology showed cystic invaginations lined by outer cuboidal epithelium and inner columnar epithelium with decapitating secretions [Table/Fig-2]. Background plasma cell infiltrate is noted. The final diagnosis was SCAP.



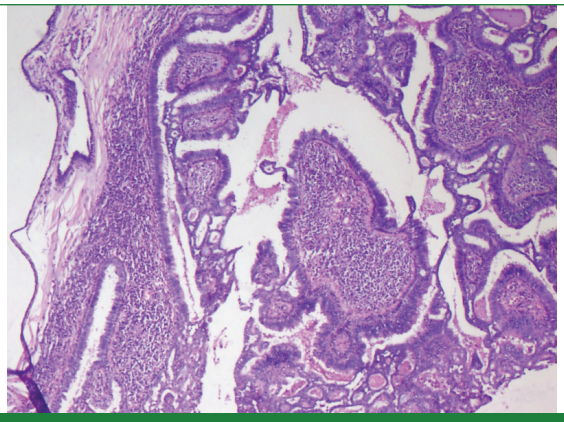
[Table/Fig-2]: 4x Haematoxylin and Eosin (H&E): revealed epidermis showing papillomatosis and was thrown up into papillary projections.

Case 3

A 62-year-old female patient, presented with a 2-year duration of an erythematous plaque with central crustation, measuring 1.9×1.7 cm over the upper eyelid [Table/Fig-1]. Clinically, the case was diagnosed as sebaceous horn and excision was done. Histopathology showed epidermis with cystic invaginations and lined by squamous epithelium gradually transitioning into a bilayered epithelium with inner columnar and outer cuboidal cells with a decapitation reaction and background plasma cell infiltrate. The final diagnosis was SCAP.

Case 4

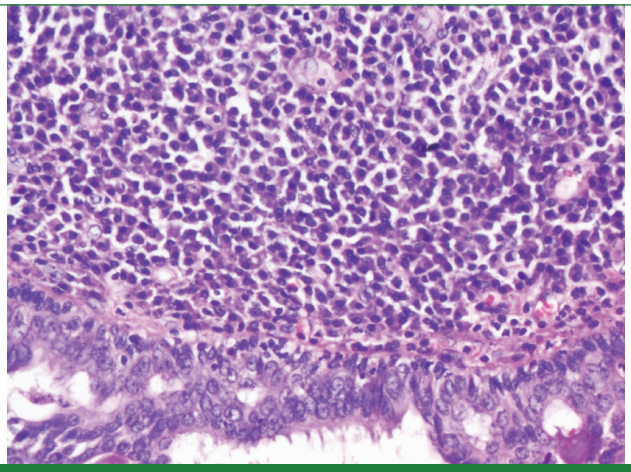
A 50-year-old male patient, presented with a 3-year duration of a verrucous ulcerative growth measuring 3.1×2.5 cm over the scalp [Table/Fig-1]. Clinically, it was diagnosed as squamous cell carcinoma. Wide local excision was done. Histopathology showed verrucous hyperplasia with hyperkeratosis and granulosis with cystic invaginations from the epidermis with luminal papillae lined by bilayered epithelium with a background plasma cell infiltrate [Table/Fig-3]. No dysplasia was seen in the sections studied. The final diagnosis was SCAP.



[Table/Fig-3]: 10x [H&E]: revealed a bilayered epithelium composed of luminal columnar cells with vesicular oval nuclei, faint eosinophilic cytoplasm with active decapitation reaction, outer row of small cuboidal cells.

Case 5

A 53-year-old female patient, presented with a 5-year duration of a slow-growing erythematous nodule measuring 2.8x2.4 cm over the left arm [Table/Fig-1]. Clinically was diagnosed as sebaceous horn and excision was done. Histopathology showed epidermis thrown into papillary projections with cystic invaginations lined by bilayered epithelium. Also associated sebaceous hyperplasia was seen in the superficial dermis with absent hair follicles [Table/Fig-4]. The final diagnosis was SCAP with associated sebaceous naevus.



[Table/Fig-4]: 40x [H&E]: revealed surrounding stroma with a diffuse plasma cell infiltrate.

Case 6

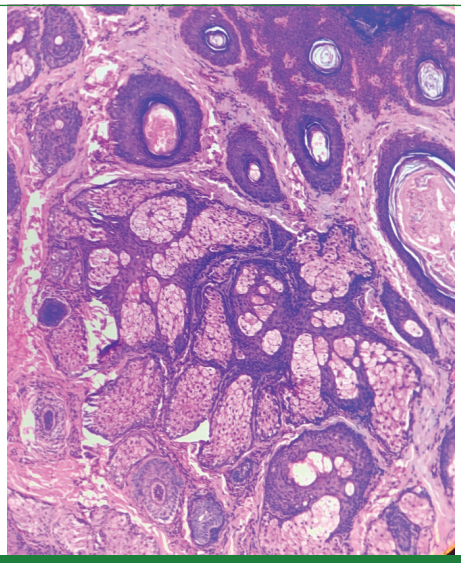
A 68-year-old male patient presented with a 3-year duration of a single, nodular skin-coloured, mass measuring 3.8x3.2 cm over the scalp [Table/Fig-1]. Clinical diagnosis was given as tubular apocrine adenoma and excision was done. Histopathology showed epidermal hyperplasia thrown into papillary projections with cystic invaginations lined by a bilayered epithelium. Background plasma cell infiltrate was seen. Based on this, the final diagnosis was given as SCAP. Tubular apocrine adenoma was ruled out since it is a dermal tumour with no epidermal connection and absent plasma cells.

Case 7

A 22-year-old female patient, presented with a 2-year duration of the solitary nodule with central umbilication and fistula with discharging fluid measuring 3×2.2 cm over the back [Table/Fig-1]. Clinical diagnosis was given as Hidradenoma and excision was done. Histopathology showed Cystic invaginations of the surface epithelium projecting into the dermis, covered by a double layer of outer cuboidal cells and inner columnar cells with decapitation secretions. The final diagnosis was given as SCAP. Hidradenoma was ruled out since it presents as a dermal nodule with no epidermal connection and usually arises in the vulva and anogenital region.

Case 8

An 11-year-old female patient, presented with a 2-year duration of the slow-growing erythematous nodule with central crustations measuring 3.6×3.1 cm, over the scalp. Clinical diagnosis was given as Hidradenoma and excision was done. Histopathology showed an exoendophytic configuration with the transition from squamous epithelium at the surface to a bilayered ductal epithelium, with decapitation secretion and plasma cells [Table/Fig-5]. The final diagnosis was given as SCAP. Hidradenoma was ruled out since it presents as a dermal nodule with no epidermal connection and usually arises in the vulva and anogenital region.



[Table/Fig-5]: 10x [H&E]: revealed dermis with large closely packed sebaceous glands, with immature hair follicles, suggestive of nevus sebaceous component.

DISCUSSION

Syringocystadenoma papilliferum was originally described by Werther in 1913 as an unusual tumor that was termed “naevus syringadenomatosus papilliferus [3]. It is postulated that SCAP arises from the pluripotent cells with the potential to exhibit either apocrine or eccrine lineage of which apocrine differentiation is more common [5]. It is usually seen at birth or during puberty and is hence called a childhood tumour [4]. It is more common in women [3]. In this case series, the tumour is diagnosed at the median age of 45 years with equal sex predilection. SCAP is known to be associated with nevus sebaceous and there is a risk of development of basal cell carcinoma [2], squamous cell carcinoma, ductal carcinoma [4] and very rarely with verrucous carcinoma [3]. Generally, three clinical types have been described. The plaque type, linear and solitary nodular forms with plaque-type being the most common [2].

Histologically, the epidermis showed irregular epidermal hyperplasia contiguous with endophytic cystic change that occupied all levels of the dermis and superficial subcutaneous fat [2,3]. Lining epithelium consists of two rows of cells, an outer layer of small cuboidal cells and an inner layer of columnar cells with demonstrable “decapitation” secretions.

Apocrine glands are deep inside the dermis [2]. The stroma contains a dense mononuclear cell infiltrate, which is comprised predominantly of plasma cells [4]. Immunohistochemistry (IHC) staining showed epithelial membrane antigen positivity in columnar cells [5]. Focal positivity of CD (Cluster of Differentiation)- 56 is seen in columnar cells. The Cytokeratin (CK)- 19 is positive in both columnar and basal cells. CK-5 and p63 are positive in basal cells. Smooth muscle actin is focally positive in basal cells [5].

The differential diagnosis of SCAP includes hidradenoma papilliferum, papillary eccrine adenomas, warty dyskeratoma and tubular apocrine adenoma [6]. Hidradenoma papilliferum presents as papillary and cystic lesions lined by bilayered epithelium similar

to SCAP. However, hidradenoma papilliferum is limited to the dermis with epidermal sparring. Plasma cell infiltrate is absent [7].

Papillary accrine adenoma also shows papillary projections into cystic invaginations. However in SCAP, papillae are thicker and lined by glandular epithelium with decapitation secretion and stroma contains plasma cells, all of these being absent in Papillary accrine adenoma [8]. Warty dyskeratoma presents as verrucous epidermal hyperplasia but lacks ductal epithelium with suprabasal acantholysis of keratinocytes, some of which are dyskeratotic [9].

Tubular apocrine adenoma has a lobular arrangement of apocrine tubular structures in the dermis surrounded by hyalinised stroma. Pseudopapillae is more common whereas SCAP is associated with true papillae. The epidermis is spared [10]. Rarely SCAP may be associated with small growths occurring alongside the tumour. They are called contiguous squamous proliferations, the pathogenesis of which is unclear [11].

Aggarwal D et al., have subtyped the contiguous squamous proliferations into two groups, Group 1: being reactive or hyperplastic conditions which include verrucous hyperplasia, papillomatosis and Group 2: including preneoplastic and neoplastic conditions namely Bowen's disease and squamous cell carcinoma. In their study, they found out that CK-7 and GATA-3 was positive in both syringocystadenoma and neoplastic squamous proliferation component, whereas non-neoplastic squamous proliferations and adjacent normal epidermis were negative for CK-7 and GATA-3 [11]. Based on these, the authors concluded that neoplastic contiguous squamous proliferations likely arise from the adenomatous component while hyperplastic contiguous squamous proliferations possibly originate from adjacent squamous epithelium/epidermis [11]. None of the cases in this study had associated contiguous squamous proliferations.

Syringocystadenocarcinoma papilliferum is the rare malignant counterpart of syringocystadenoma papilliferum. Ulceration or rapid enlargement is indicative of malignant transformation [2]. The diagnosis is clinically suspected and histologically confirmed by the presence of asymmetry, poor circumscription, marked cytological atypia and extension into dermis [2].

CONCLUSION(S)

In conclusion, syringocystadenoma papilliferum is a rare neoplasm, and even if it is called a childhood tumour, it rarely appears in adults. In the present case series, the clinical diagnosis was sebaceous horn in most cases with one case suspected as squamous cell carcinoma. Histopathological examination is the gold standard for the diagnosis of SCAP. Histopathological examination is also important to rule out co-existing malignancies. Due to the risk of the development of malignancy, complete surgical excision along with the histopathological examination is the treatment of choice.

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