

Syringocystadenoma Papilliferum in an Unusual Location – A Rare Presentation

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Case Report

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Abstract: Syringocystadenoma papilliferum is an uncommon benign tumor of disputed histogenesis, with a predilection for the scalp and forehead. Less common sites of involvement are the chest, upper arms, male breast, eyelids, scrotum, and thighs. There is an associated organoid nevus in approximately one-third of cases¹. We reported a case of Syringocystadenoma papilliferum with sessile nodular lesion located on the vulva of an 49 yr old female. This case illustrates the atypical location of this adnexal tumor and adds to the differential diagnosis of lesions on the vulva. Only few cases reported in the literature.

Keywords: Syringocystadenoma papilliferum, skin adnexal tumor, organoid nevus.

INTRODUCTION

Syringocystadenoma papilliferum is an uncommon skin adnexal tumor. It occurs most commonly on the scalp or the face; however, in about one-fourth of the cases, it is seen elsewhere. It is usually first noted at birth or in early childhood and presents as a papule or several papules in a linear arrangement, or as a plaque. The lesion increases in size at puberty, becoming papillomatous and often crusted. On the scalp, syringocystadenoma papilliferum frequently arises around puberty within a nevus sebaceus that has been present since birth [1]. Syringocystadenoma papilliferum has also been reported in other locations, including thigh, breast, and eyelid. However, it has rarely been reported in the female genitalia.

CASE REPORT

A 49-year-old woman presented with a single nodular lesion in vulval region of 8 months duration.

It was sessile nodule (2cm) with eroded surface on left side of introitus in labia majora (Figure 1). The patient reported increase in redness around the nodule and increase in size which she noticed over the last few days.

Gross examination showed single skin covered soft tissue measuring 2x1x0.5 cm. Cut section showed a thick walled cyst with irregular inner surface showing multiple papillary projections. Microscopic examination

revealed stratified squamous epithelium of skin with the underlying stroma showing a fibrocollagenous cyst wall lined by stratified squamous epithelium thrown into multiple papillary folds lined by a double layered to multilayered epithelium with focal areas showing decapitation secretion and the core of the papillae containing plasma cells and few lymphocytes. Deeper areas show chronic inflammatory cell infiltration and fibrous stroma (Figure-2, 3 & 4).



Fig-1: Sessile nodule (2cm) with eroded surface on left side of introitus in labia majora

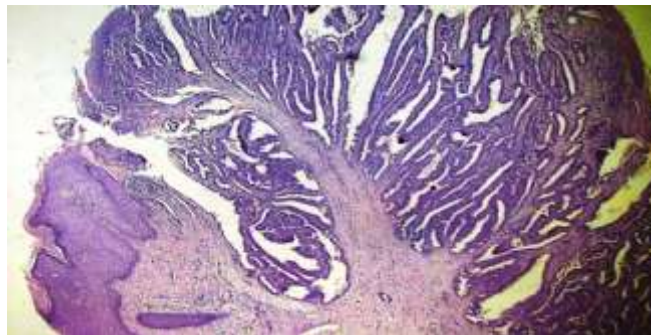


Fig-2: Stratified squamous epithelium of skin with the underlying stroma showing multiple papillary folds lined by a double layered to multilayered epithelium.

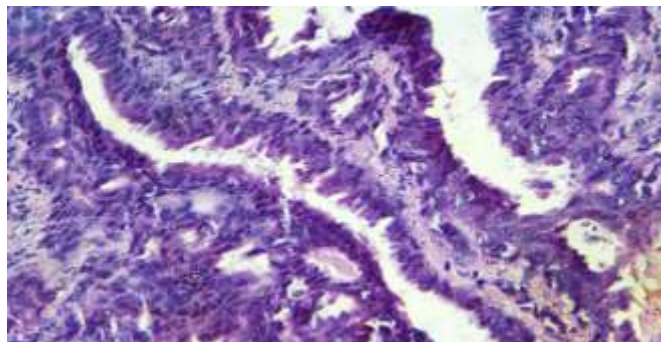


Fig-3: Focal areas showing decapitation secretion

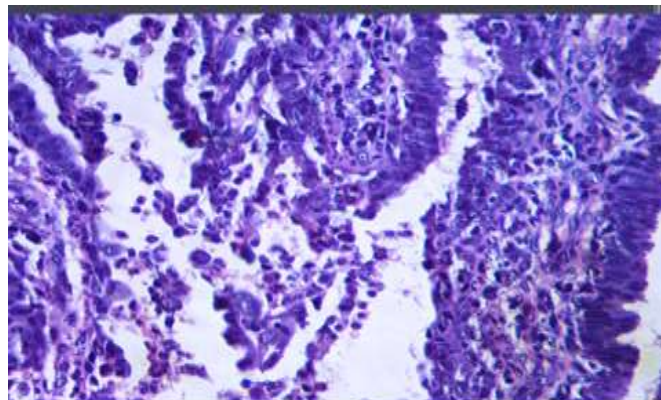


Fig-4: Core of the papillae containing plasma cells and few lymphocytes.

DISCUSSION

Syringocystadenoma papilliferum is an uncommon benign tumor of disputed histogenesis, with a predilection for the scalp and forehead. Less common sites of involvement are the chest, upper arms, male breast, eyelids, scrotum, and thighs. The tumor has a varied clinical appearance, most often presenting as a raised warty plaque or as an irregular, flat, gray or reddened area. Linear papules and nodules are occasionally present. The lesions measure from 1 to 3 cm in diameter and are usually solitary. Alopecia accompanies those on the scalp. There is increasing evidence for an apocrine histogenesis, but the possibility of an eccrine origin for a few cases seems likely. Another theory suggests the apoecrine glands as the origin of this tumor. It may be derived from pluripotent cells. Allelic deletions have been reported at 9p21 (p16) and 9q22 (the patched gene) [2].

Investigations using special stains, immunohistochemistry, and electron microscopy have produced variable results; therefore, different authors have postulated differentiation toward eccrine, apocrine, and apoecrine gland [3]. Syringocystadenoma papilliferum is an uncommon skin adnexal tumor. The majority of these tumors occur in the head and neck region and are occasionally associated with nevus sebaceous and basal cell carcinoma.

Helwig and Hackney [4] suggested that 10% of Syringocystadenoma papilliferum (SP) cases develop into basal cell carcinoma. This association is attributed to similar allelic deletions of human homologue of the *Drosophila* patched gene (*PTCH*) [5]. Syringocystadenoma papilliferum has also been reported in other locations, including thigh, breast, and eyelid. However, it has rarely been reported in the female genitalia [6, 7]. Syringocystadenoma papilliferum sometimes occurs in association with other lesions, including condyloma acuminatum [8] and verrucous carcinoma [9].

One differential diagnosis in the female genitalia is hidradenoma papilliferum, which is a benign tumor of apocrine sweat gland that usually presents as a dome-shaped tumor arising in the interlabial sulci and is usually asymptomatic. Histologically, hidradenoma papilliferum is characterized by a complex delicate fibrovascular branching stalk. Unlike SP, it is usually not connected to surface epithelium. Two cell linings cover the stalks: outer epithelial cells and inner myoepithelial cells. Another distinguishing feature is the absence of plasma cells in the papillary stalks [10].

Although benign, SP can be associated with malignant tumours such as basal cell carcinoma, metaplastic adenocarcinoma and ductal carcinoma [11, 12]. There has been report in the literature of pure intraductal carcinoma of the breast and invasive

carcinoma of ectopic breast tissue of the vulva detected simultaneously in the same patient [13]. SP may also be linked to its malignant counterpart syringocystadenoma papilliferum. It is difficult to determine what the estimated risk of malignant transformation of the SP is due to its rarity.

Carcinoembryonic antigen is usually present in the epithelial cells. Gross cystic disease fluid protein-15 (GCDFP-15) is variably positive in the tumor cells. The luminal columnar cells express CK7 and more than 70% are positive for CK19. The basal cuboidal cells usually express CK7 also, but the expression of CK19 by these cells is variable. IgA and secretory component have also been demonstrated in these cells, and it has been suggested that the cells attract plasma cells by a similar mechanism to that utilized by glands of the secretory immune system. Further evidence for the existence of an eccrine variant comes from a study of the eccrine-specific marker IKH-4 which has labeled one case.

CONCLUSION

Syringocystadenoma papilliferum is a benign cutaneous adnexal tumor that rarely occurs in the female genitalia. Because of its association with malignant lesions, it should not be confused with hidradenoma papilliferum, which commonly occurs in this region. We report this case because of its rare location. In future we should not miss this tumour as differential diagnosis in vulval region.

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