SYRINGOCYSTADENOMA PAPILLIFERUM PRESENTING AS VULVAL CYST - A RARE CASE REPORT

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ABSTRACT

Syringocystadenoma papilliferum is a benign skin adnexal tumour with verrucous appearance that is commonly located in the scalp, neck and face. It usually presents at birth as a solitary papule or as multiple papules and increases in size after puberty. It occurs either de novo or within some lesions such as nevus sebaceous. We report a case of a 75 year old female with a cystic lesion in the labia majora, which on histopathological examination revealed syringocystadenoma papilliferum. This case is presented for its unusual location in the vulva as only a few cases were reported in this location in the literature.

Keywords: *Syringocystadenoma papilliferum, Verrucous, Papule, Skin adnexal tumour & Nevus sebaceous*

INTRODUCTION

Syringocystadenoma papilliferum is a benign skin adnexal tumour commonly located in the head and neck region, especially the scalp or the face. [1] In one fourth of the cases, it is seen in other sites like thigh and breast. It is very rarely located in the female genitalia. It is usually presents since birth as a birth mark or in early childhood. [2]

CASE REPORT

A 75 Years female presented with a cystic swelling in the labia majora for one month duration. With a clinical diagnosis of Bartholin's cyst, the swelling was excised and sent for histopathological examination.

Macroscopic examination showed single skin covered soft tissue measuring 3x2x1 cm. [Figure1] Cut section showed a thick walled cyst with irregular inner surface showing multiple papillary projections. [Figure2]

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. [Figures 3 and 4]

An impression of Syringocystadenoma papilliferum was then made. On follow up, the patient was asymptomatic and the wound was found to be healed well.

Fig 1: Gross appearance - External surface

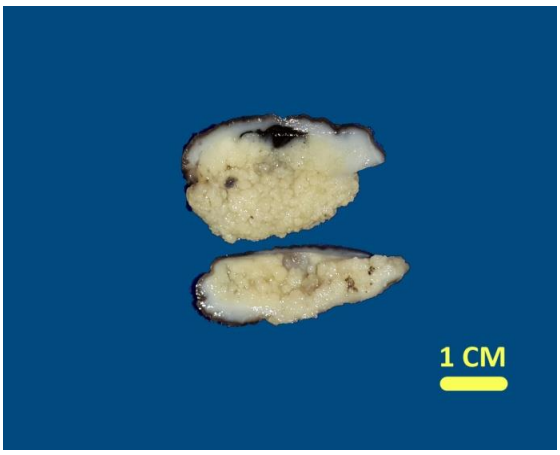

Fig 2: Cut section showing a cyst with inner surface showing multiple papillary projections

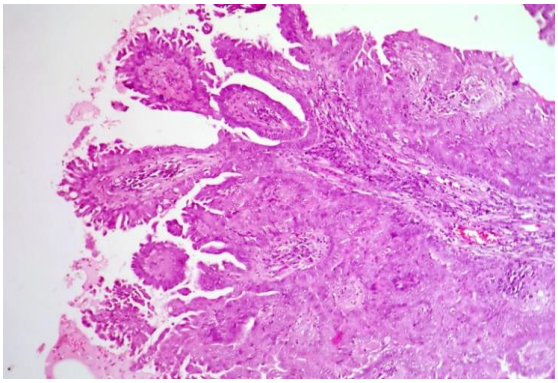

Fig 3: Cyst wall lined by stratified squamous epithelium thrown into multiple papillary folds. (H and E x40)

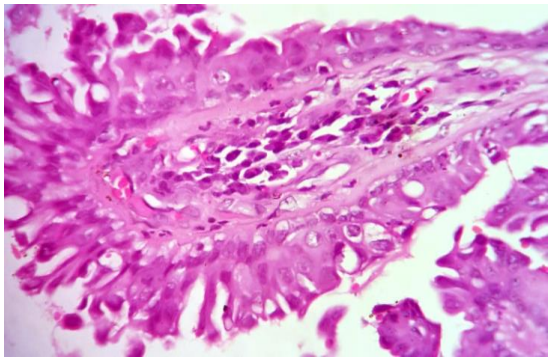

Fig 4: The papillae are lined by a double layered epithelium with decapitation secretion and the core showing numerous plasma cells and lymphocytes. (H and E x100)

DISCUSSION

Syringocystadenoma papilliferum is a rare benign skin adnexal tumour with verrucous appearance, commonly located in the head and neck region, especially the scalp, neck and face. [1], [3] Uncommon sites of occurrence include trunk, breast, arms, axilla, lower limbs and genital region.[4], [5], [6], [2] Clinically it presents as a papule or plaque and increases in size after puberty to form papillomatous and crusted lesion. On the scalp, the lesion is usually associated with alopecia. Usually it is less than 4 cm in size. Rarely, it can present as multiple lesions. [7]

Histologically, the epidermis shows papillomatosis, with cystic invaginations extending downward. Upper portion of the cyst shows keratinised squamous lining whereas the lower portion shows two-cell lining of glandular epithelium. The inner row is composed of tall columnar cells with oval nuclei and faintly eosinophilic cytoplasm, some with decapitation secretion and outer cuboidal cells with scanty cytoplasm and round nucleus, which are the myoepithelial cells. Dense inflammatory cell infiltration with plasma cells is seen in the stroma, especially in the core of the papillary projections. [1], [3] Eccrine syringofibradenoma is a particular variant with prominent fibrous component. [1]

The malignant counterpart of this tumour is syringocystadenocarcinoma papilliferum. In one third of the cases, there is association with nevus sebaceous. In one tenth of the cases, it is associated with basal cell carcinoma. [1] In a study by Felix Boon-Bin Yap et al, review of the literature for cases of syringocystadenoma papilliferum outside the head and neck region yielded 69 cases with such characteristics. Out of which, 5 cases were reported in the vulva and 12 cases were associated with viral warts, nevus sebaceous, linear hidradenoma, hidradenoma papilliferum, verrucous carcinoma, syringocystadenocarcinoma and papillary eccrine adenoma. [2]

Syringocystadenocarcinoma papilliferum is a tumour exhibiting both apocrine and eccrine differentiation which has been proved with special stains (alcian blue, colloidal iron, and periodic acid-Schiff (PAS)), Immunohistochemistry and electron microscopy in various studies. [1], [8] Syringocystadenoma papilliferum expresses AE1/AE3, CAM 5.2, epithelial membrane antigen (EMA), and carcinoembryonic antigen (CEA). The inner layer is positive for smooth muscle antigen (SMA). The results of markers of apocrine differentiation are variable. Some authors have found gross cystic disease fluid protein 15 (GCDFP-15) positive. [1]

Syringocystadenocarcinoma papilliferum has to be differentiated from hidradenoma papilliferum. The latter is commonly seen in the perineal region presenting as nodules with more complex papillary patterns. It has the same two cell epithelial lining, but lacks the plasma cells in the stroma. [9]

It should also be differentiated from adenocarcinoma, in which there is marked nuclear atypia, infiltrative growth pattern and lack of myoepithelial layer. [1], [10]

Syringocystadenoma papilliferum is found to be associated with malignant tumours like verrucous carcinoma, basal cell carcinoma, sebaceous carcinoma and ductal carcinoma. [2]

CONCLUSION

Syringocystadenoma papilliferum is a relatively rare tumor outside the head and neck region. Solitary tumors in such unusual locations leads to multiple differential diagnoses and must be examined histopathologically for confirmation.

This case is presented for its rare unusual location in the vulval region.

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