

Case Report

SYRINGOCYSTADENOMA PAPILLIFERUM AS A CUTANEOUS MASS IN AN UNUSUAL LOCATION: A CASE REPORT

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SUMMARY

Syringocystadenoma papilliferum (SP) is an uncommon adnexal adenoma, most often located on head and neck. We described an unusual appendage tumour, most importantly appearing at later age with vulvar location adjacent to the labio-femoral sulcus situated in the cutaneous tissue and having rapid growing pattern. Different prominent Ig class was found in the secretory component of the tumour epithelial cells and in the plasma cells underlying the tumour epithelium. A diagnosis of SP was made by characterizing histopathologic features of the biopsy specimen.

KEY WORDS: Syringocystadenoma papilliferum, unusual location, immune globulin distribution.

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INTRODUCTION

Syringocystadenoma papilliferum is a slow-growing, benign skin appendage tumor that most frequently occurs on the scalp or face, but one fourth of the cases can be seen elsewhere¹⁻³. It is generally noted at birth or in early childhood and consists of either one papule or several papules in a linear arrangement of a solitary plaque⁴. The lesion increases in size at puberty, becoming papillomatous

and often crusted⁵.

The histogenesis of this neoplasm is likewise uncertain^{2,6}. Although most cases originate from the apocrine sweat gland apparatus^{5,7} some reports point to an eccrine origin^{5,7} or pluripotential epidermal origin⁸, but most authorities favor an apocrine derivation^{2,6}. The present case was indented to report SP, which exclusively appears at later age with rapid growing pattern and in an unusual location.

CASE HISTORY

A forty-five years old woman had a rapidly growing mass on the vulvar location near to right labio-femoral sulcus. The mass had been seen to be present for two months when she was admitted in. It had recently increased in size accompanied by pain and pruritis. The examination revealed a skin colour, oval and solitary 2x2 cm tumour-like mass situated at Cutaneous tissue without any features in the overlying skin. The site and appearance of the tumour resembled a vulvar Cutaneous tumour or skin appendage tumour.

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The entire tumour was totally excised and submitted for microscopic examination. In the histopathologic examination, it was circumscribed tumour surrounded by a fibro-connective tissue. It was a totally large cystic endophytic tumour and situated at Cutaneous tissue of the vulva. The tumour had no communication with the surface and not associated with naevus sebaceous. There was a small focus of ectopic eccrine glandular element surrounding the tumour. The most recognizable feature was the presence of papillary processes at various sizes lined with double epithelial cell layers. The layer nearest the dermis consists of flattened or cuboidal small cells with scanty cytoplasm and large round or oval nuclei. The layer adjacent to the lumen is composed of columnar cells with decapitation secretion, centrally located oval

nuclei and pink cytoplasm. The degree of papillation was variable ranging from tubuloglandular structures to a cribriform or lace-like pattern to epithelial cells modestly projecting into the lumen. The fibrous connective tissue of the papillary stalk contained abundant plasma cells together with a few lymphocytes and histiocytes. Thus, the diagnosis was Syringocystadenoma papilliferum since these features were typical for SP (Fig. 1-2).

In addition to routine hematoxylen-eosin (H.E) stain, the tissue sections were stained immunohistochemically for the immunoglobulins; Ig A and Ig G (Fig. 3-4). Plasma cells located in the fibrovascular papillary stalk and the epithelial cells covering papillary surface were stained densely for the Ig A and faintly for Ig G (Fig. 3-4).

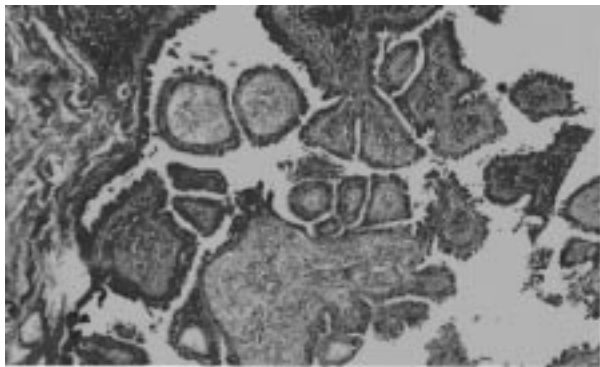


Fig. 1: A central invagination lined by two cell layers with villous formation. Stromal infiltrate contains mainly plasma cells (Hematoxylin-eosin, x 120).

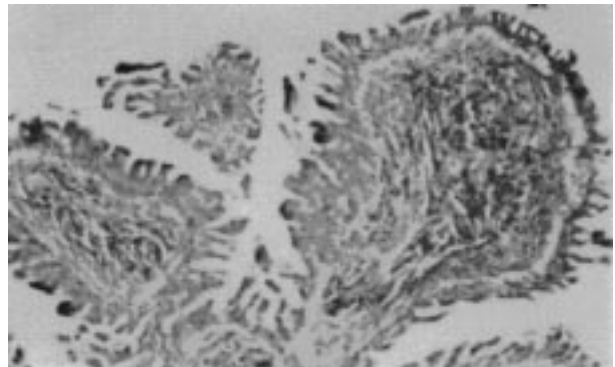


Fig. 3: Most of epithelials and plasma cells showed positive staining with IgA (Ig A, x240).

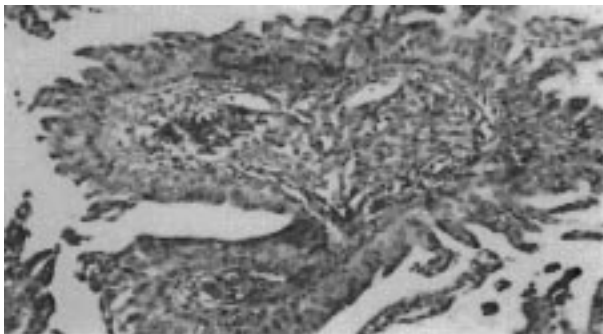


Fig. 2: Note two distinct epithelial cell layers with small cuboidal cells near the dermis and columnar cells adjacent to the lumen (Hematoxylin-eosin, x 240)

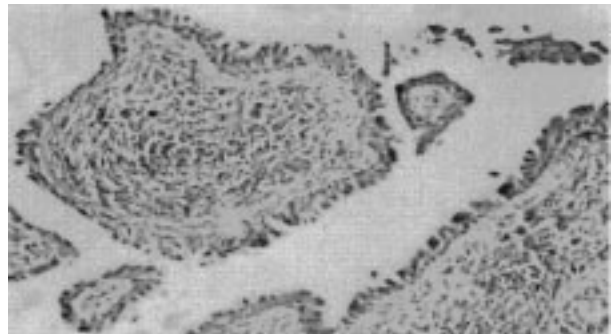


Fig. 4: Some of epithelials and plasma cells showed positive staining with IgG (Ig G, x240)

DISCUSSION

We believe that the present case is unusual for its location, rapid growth and clinical presentation with pain and pruritis. Syringocystadenoma papilliferum is most commonly located on the head and neck^{1,5,9-12} and one fourth of the cases were seen elsewhere apart from head and neck^{1,2,13}. It has been reported that in the literature there were 145 cases, with 75% on the head and neck, 20% on the trunk and 5% on the extremities. Seven of the eight extremity lesions occurred on the lower limbs^{1,5,9-11}.

In our case, the tumor was situated at the Cutaneous tissue of the vulva. The primary occurrence of Syringocystadenoma papilliferum on the vulva has not been seen in the literature. It is most likely that this is a first-ever report in the literature, emphasizing an unusual location of Syringocystadenoma papilliferum: at Cutaneous tissue of the vulva.

Syringocystadenoma papilliferum usually asymptomatic, although pain¹² and pruritis¹⁴ were reported. Patients with scalp lesions may complain of mild irritation or bleeding when combing their hair¹⁰. In our case, the patient experienced pruritis and pain in association with an increase in the size of her lesion.

Syringocystadenoma papilliferum is a slow-growing, benign skin appendage tumor¹⁻³ which is often noted at birth or in the early childhood, with rapid growth at puberty^{4,5}. The present case was 45 years old women and the tumor has been present for two months and progressively increased in size.

The usual morphologic type of Syringocystadenoma papilliferum is a solitary plaque^{1,2,5} and one to several papules^{4,5}. The plaques are usually less than 4 cm in diameter and range in colour from skin colour to dark brown. They may be flat and smooth or raised with a papillomatosis or verrucous surface⁵. The less common papular lesions are skin colour to pink and less than 1 cm in diameter. When there are multiple papules, they are usually arranged in a linear configuration^{4,14}. This clinical course differed from the case presented here

because our patient had the lesion in subcutaneous tissue of the vulva. Syringocystadenoma papilliferum has been associated with several neoplasms and hamartomas. Its occurs most often with a nevus sebaceous². In our patient there was no clear-cut nevus sebaceous in the biopsy specimen.

Microscopically, Syringocystadenoma papilliferum displays a variety of surface features ranging from smooth to papillomatous to a cup-like depression or small papillations. The most recognizable feature of Syringocystadenoma papilliferum is the presence of papillary processes of various sizes lined with two epithelial layers¹. The degree of papillation is variable ranging from tubuloglandular structures to a cribriform pattern or epithelial cells modestly projecting into the lumen.

The luminal layer is columnar with decapitation secretion, whereas the outer layer consists of smaller cuboidal cells with larger nuclei and comparatively meager cytoplasm. The connective tissue stroma that surrounds the duct-like structures projects into the papillations. It is usually rich in inflammatory cells of various types, most notably plasma cells. The tumor itself is free of pilosebaceous units. In contrast, many dilated sweat glands frequently exist beneath the lesion¹.

Jakobiec *et al.* defined three histopathologic features of Syringocystadenoma papilliferum. Firstly, the cystic spaces within the dermis were lined by nonkeratinizing duct like epithelium which was continued with the keratinized surface opening. Secondly, the papillary projections protruded at the surface and within the dermal cystic invaginations. Finally, a prominent plasmacytic dermal infiltration surrounded these papillary projections and invagination¹⁵. The final feature was also seen with our case. Thus, histologic evidence in this case supports the diagnosis of Syringocystadenoma papilliferum.

The histogenesis of Syringocystadenoma papilliferum is still an ongoing debate whether it is apocrine or eccrine derived^{1-3,5,7,12}. The results of light microscopic studies^{1,16} electron

microscopic studies^{7,8} and histochemical analysis^{17,18} have been contradictory in these matters. Syringocystadenoma papilliferum was first reported as a tumor of sweat gland origin by Peterson in 1892¹⁹. Since Shiefferdecker's differentiation of the sweat glands into apocrine and eccrine in 1917²⁰ the attempts to prove the apocrine or eccrine origin of the lesion have led to considerable controversy.

The decapitation secretion observed in Syringocystadenoma papilliferum is usually thought to imply an apocrine origin^{1,2,5} but the decapitation secretion demonstrated in some cases of eccrine spiradenoma and other eccrine tumors¹⁶. In addition, light microscopic studies of Syringocystadenoma papilliferum tended to favour an apocrine²¹ rather than an eccrine origin⁵.

From histochemical and electron microscopic findings, Hashimoto et al.^{8,17} reported that Syringocystadenoma papilliferum is a tumor differentiating towards eccrine structures and therefore classify Syringocystadenoma papilliferum in their monograph^{8,17} as a tumor of eccrine sweat gland origin. Lever insisted on the primary epithelial germ theory²². Pinkus has reviewed the dynamics of Syringocystadenoma papilliferum on the basis of light microscopical observations²³ and concluded that the histogenesis of SP is multi-form. The most cases originate from proliferated mature apocrine sweat gland apparatus and some are associated with the eccrine sweat gland or originate from undifferentiated pluripotential cells² in the adult epidermis stimulated by trauma or by unknown factors. These conflicting evidence led to the suggestion of a mixed eccrine and apocrine origin for these lesions, originating from an apocrine gland or from a pluripotential glandular cell^{3,24}.

An alternate theory suggests that it is originated from the recently described apocrine gland^{25,26}. This hybrid type of gland exhibits microscopic, immunohistochemical and ultra-structural features of eccrine and apocrine elements, including decapitation secretion.¹⁶

The secretory immune system is a branch of

the humoral immune system concerned with the production and secretion of secretory IgA. This process requires the participation of both plasma and epithelial cells. Bone-marrow-derived B lymphocytes within Peyer's patches migrate after antigen exposure to mesenteric lymph nodes where they proliferate and differentiate into IgA-producing cells. These cells enter the bloodstream via the thoracic duct and return selectively to the intestinal lamina propria where they undergo further maturation into IgA-secreting plasma cell²⁷.

Dimeric IgA secreted by these cells is released as secretory IgA at the luminal epithelial surface^{28,29}. Circulating IgA plasma cells or their precursors might alternatively localize to other glands capable of secretory IgA production. In this manner, secretory IgA is present in a variety of secretions, including those of the salivary glands³⁰ and breast^{31,32}. An alternative method for secretory IgA secretion is uptake of circulating dimeric IgA from the blood and subsequent secretion by epithelial cells. This has been demonstrated in the resting mouse mammary gland³³.

Certain neoplasms possess the ability not only to synthesize secretory component but also to attract IgA plasma cells and take up IgA into the cytoplasm. This inference has been derived from the information similar to the report for colonic adenoma,^{34,35} medullary carcinoma of the breast³⁶. Thus, Syringocystadenoma papilliferum joins the list of plasmacytotropic neoplasms. The intracellular handling of IgA by plasmacytotropic neoplasms appears to recapitulate the pathway utilized by normal epithelial cells³⁷.

A highly diagnostic feature is the almost invariable presence of fairly dense cellular infiltrate entirely composed of plasma cells in the stroma of this tumor, especially in the papillary projections. These are predominantly of the IgG and IgA classes^{11,38}.

In our case, the plasma cells and superficial epithelial cell layer showed IgA and IgG positivity. Examination of the plasmacytic infiltration in Syringocystadenoma papilliferum demonstrated preponderant Ig A and Ig G

immunostaining within the infiltrating cells along with intraepithelial IgA positivity. It was proposed that the plasmacytotropic infiltration of this entity was similar to that of the glands of the secretory immune system.

We have concluded that because the breast is a modified apocrine sweat gland and is a component of the secretory immune system, it can be tempted to apply these findings toward an apocrine histogenesis for Syringocystadenoma papilliferum. In this manner, neoplastic transformation of the apocrine glandular epithelial cells would be accompanied by expression of the ability of these cells to attract plasma cells.

To our knowledge, this case represents the first report in the literature that located subcutaneous tissue of the vulva with rapid growing pattern, pruritis and appearing at later age.

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