

VULVAR SYRINGOCYSTADENOMA PAPILLIFERUM: A RARE GYNAECOLOGIC TUMOUR - Case report and Literature review

ABSTRACT: Syringocystadenoma papilliferum (SCAP) is a benign skin adnexal tumor that arises from apocrine sweat glands. SCAP is commonly seen in the head and neck area of the body but rarely occurs in the vulva-perianal skin.

We present the case of a 25-year-old primipara who presented with painless right vulva swelling of 9 months duration which was not increasing in size. Examination revealed a 2cm long mass situated in the inferio-medial part of the lower half of the right labium majus which readily slipped under the skin. There was no associated skin discoloration, ulceration, tenderness, or differential warmth. An excision biopsy was performed under general anaesthesia and the specimen was subjected to histological examination. Histopathology revealed Syringocystadenoma papilliferum. The patient was asymptomatic and the wound had healed well on a follow-up visit.

INTRODUCTION Syringocystadenoma papilliferum is a slow-growing, benign skin adnexal tumor that most frequently occurs in the head and neck region in 7 out of 10 cases^{1,2}. Other sites have been reported but vulva lesion is very rare³⁻⁷. It is generally noted at birth or during early childhood up until puberty and usually consists of either a papule or several papules in a linear arrangement⁸. The lesion increases in size at puberty, becoming papillomatous and often crusted. The aetiopathogenesis of SCAP is unclear⁹. Although most cases originate from the apocrine or eccrine glands. Some reports of pluripotential epidermal origin have also been documented⁹. The present case was noted after puberty in a rare location. The importance of this lesion lies in its possible co-existence with other benign tumors and the possibility of malignant transformation⁶.

CASE PRESENTATION: A 25-year-old primipara was referred to the gynaecological clinic on account of a right vulva swelling noticed nine months prior to presentation. There was no associated pain, discharge, ulcer or swelling elsewhere in the body. The swelling had not increased in size over the said period. She had no relevant past gynaecologic, obstetrics, medical, family, or social history. She was clinically well and her vital signs were normal. Chest, cardiovascular and abdominal examination were essentially normal. The mass was located in the inferio-medial part of the lower half of the right labium majus measuring 2cm in diameter. It was rubbery with no discoloration, ulceration, tenderness or differential warmth. It was not attached to the overlying structures or skin. The left labium majus, vagina, cervix and uterus appeared normal. She had no palpably enlarged groin lymph nodes. Her blood samples tested negative for human immunodeficiency virus, hepatitis B virus and hepatitis C virus. Her packed cell volume was 38%. She was counselled on the need for excision which she consented to. This was done under general anaesthesia on December 21, 2021 and the specimen was subjected to histological examination which revealed Syringocystadenoma papilliferum. On follow-up, the patient was asymptomatic and the wound had healed well.

DISCUSSION: Syringocystadenoma papilliferum (SCAP), also known as papillary syringadenoma is a benign, hamartomatous, adnexal skin tumor. The origin is uncertain but it is thought to be derived from the apocrine glands eccrine sweat glands or pluripotent stem cells. It has been reported in different parts of the body but it most commonly occurs in the head and neck region. Other reported sites include the trunk, extremities, genitalia, abdomen, auditory canal, buttock, vulva and scrotum, pinna, eyelid, outer ear canal, postoperative scar, scalp, nipple, thigh, axilla, back, and right lower abdomen. It is an extremely rare condition in gynaecological practice with a few cases identified in the literature.⁷⁻⁹ These cases were reported in women between 27 and 64 years old. There is no gender predilection. Most cases (about 50%) are present at birth or in early childhood¹ and increased in size, becoming crusted and papillomatous in texture at puberty. However, 30% of cases developed for the first time during puberty¹.

Although benign, co-occurrence of basal cell carcinoma, verrucous carcinoma, metastatic carcinoma, and ductal carcinoma have been documented⁸. It may be observed in association with other benign adnexal neoplasms such as nevus sebaceous (8 - 19% of patients), apocrine nevus, tubular apocrine adenoma, apocrine hidrocystoma, apocrine cystadenoma, and clear cell syringoma⁹. Differential diagnosis of SCAP includes molluscum contagiosum, syringoma, hydradenoma Papilliferum, and basal cell carcinoma⁸.

The clinical presentation is variable but usually presents as a solitary papule or several papules arranged in a linear fashion. It could also present as warty plaques, often in the setting of nevus sebaceous⁹. Nodular or verrucous transformation may occur at puberty.

Diagnosis is typically confirmed on histology. It frequently shows as in our case... (See histology report)

Immunohistochemistry, albeit, was not done.

In view of possible malignant transformation, a complete excisional biopsy with a 3-5mm healthy skin margin is recommended¹³. The depth of excision is not clearly documented in the literature. We encourage the inclusion of SCAP in the differential diagnosis of vulvar lesions. Excisional biopsy appears to be an effective treatment.

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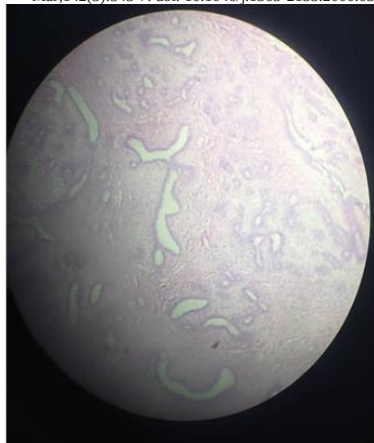
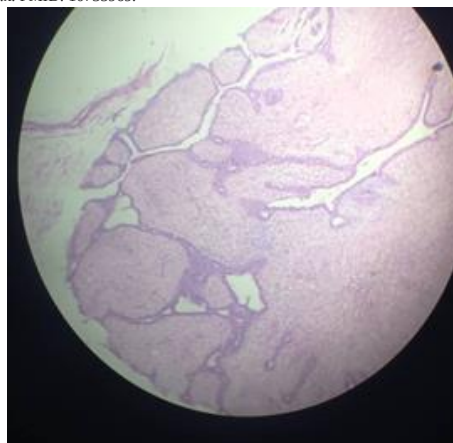


Figure 1: Photomicrograph of Syringocystadenoma papilliferum showing cystic component, irregular tubules with intervening fibrocollagenous tissue .Figure 2: Photomicrograph of Syringocystadenoma papilliferum showing papillary architecture of epithelial lining.