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Case presentation

Linear syringocystadenoma papilliferum on female breast: a rare appendageal tumour on an uncommon location

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Abstract

Syringocystadenoma papilliferum is a rare benign adnexal tumor commonly located on the head and neck region and is usually associated with a nevus sebaceous. Linear lesions are uncommon and lesions on the breast are extremely rare. We report here a case of linear SCAP occurring *de novo* on the left breast of a 35-year-old healthy woman. Histopathology examination showed the characteristic papillary projections lined by a double layer of cells inside epidermal invaginations.

Keywords: syringocystadenoma papilliferum, linear, female, breast

Introduction

Syringocystadenoma papilliferum (SCAP) is a benign adnexal tumor most commonly located on the head and neck region often in association with a nevus sebaceous. SCAP may also arise *de novo* without any pre existing skin lesion [1]. Sometimes present at birth, the lesions usually develop during childhood. The usual morphology is a brownish or erythematous papule, nodule, or plaque. Multiple lesions in a linear arrangement are a rare occurrence [2]. About one fourth of the cases occur outside the head and neck region [1]. We hereby report an adult-onset case of SCAP occurring *de novo*, representing the third report in the English literature of SCAP occurring on the female breast.

Case synopsis

A 35-year-old woman presented with bright red cauliflower-like growths on her left breast. The condition started as a small dusky red plaque on normal skin about eight months previously. Additional small lesions appeared over the vicinity, coalesced, grew in size, and developed surface erosion. Besides mild irritation, the condition was asymptomatic. The patient was treated previously with hot compresses, various topical medications, and systemic antibiotics without any benefit. There was no significant past or family history. The patient had otherwise enjoyed excellent health. Cutaneous examination revealed two erythematous, multilobular exuberant plaques over the left breast. The lateral one was globular and about 1.5 cm in diameter and the medially situated larger one was linear, leaf shaped, and 3-4 cm in length.



Figure 1. Erythematous, lobulated growths on the left breast.

The surface of the tumor was moist but free from any kind of discharge. The adjacent skin showed mild hyperpigmentation and telangiectasia. Examination of the breasts and regional lymph nodes was noncontributory. The rest of the cutaneous and systemic examination was normal.

The differential diagnosis included squamous cell carcinoma, SCAP, and giant granuloma pyogenicum. A lesional biopsy specimen showed cystic invaginations of the epidermis with numerous papillary folds in the lower part of the invagination. The invaginations and the papillary folds were lined by two layers of cells; the inner columnar and the outer cuboidal. A moderately dense cellular infiltrate rich in plasma cells was also visible. There was no cellular atypia, keratin pearls, or vascular proliferation.

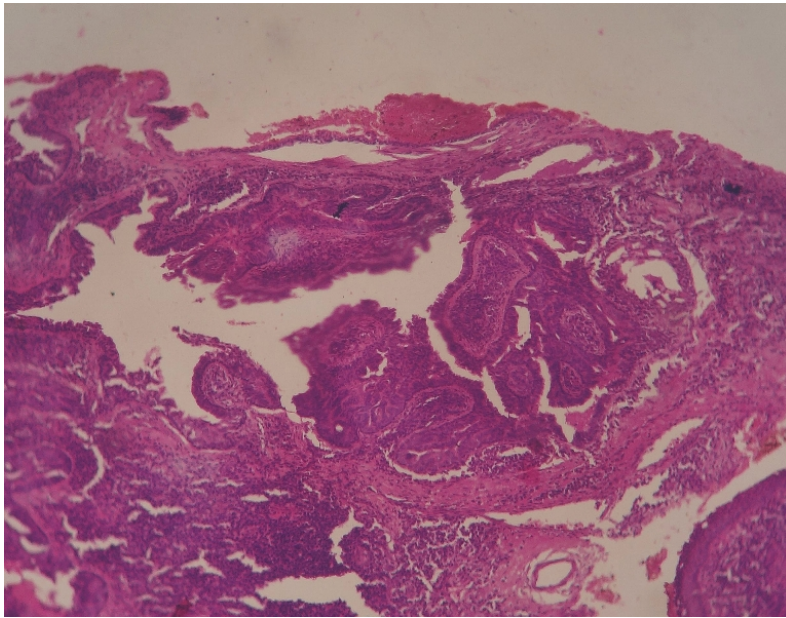


Figure 2. Papillary projections inside epidermal invagination lined by double layer of cells. There is a rich infiltrate in the core of the projections. H & E. X 100

On the basis of the clinical findings and characteristic histopathology, a final diagnosis of syringocystadenoma papilliferum was made and surgical excision was advised for the patient.

Discussion

Syringocystadenoma papilliferum is an uncommon appendageal tumor of uncertain histogenesis. Most evidence support an apocrine origin, but eccrine or apoecrine gland origin has also been suggested [3]. The lesions may be derived from pluripotent stem cells.

SCAP usually is a solitary lesion with varied appearance, most commonly presenting as a verrucous papule or a warty plaque or nodule. Linear lesions occur very rarely. Associated with nevus sebaceous in about one-third of cases, SCAP may be associated with a wide variety of other tumors such as basal cell epithelioma, sebaceous epithelioma, apocrine hidrocystoma, trichoepithelioma, eccrine spiradenoma, and eccrine poroma.

Yap et al [4] reviewed 69 cases of SCAP outside the head and neck region and found most lesions occurred on the trunk (53.5%), followed by extremities (33.8%), and genitalia (12.5). We could locate only five reports of syringocystadenoma involving the breast [5-9]. Of the five cases, two occurred on female breasts, one presented as a cauliflower-like growth mimicking carcinoma [8], and the third was associated with nevus sebaceous and tubular apocrine adenoma [9].

The histology of SCAP is distinctive. Cystic invaginations extend from the epidermis and from the lower portions of the invaginations; numerous papillary projections extend into the lumina of the invaginations. The papillary projections and the lower portion of the invaginations are lined by glandular epithelium consisting of two rows of cells: an inner luminal row of columnar cells some of which may show active decapitation secretion and an outer row of small cuboidal cells with round nuclei [10]. An infiltrate rich in plasma cells is found in the core of the lesion.

Although SCAP is known to be associated with malignant tumors such as basal cell carcinoma, sebaceous carcinoma, verrucous carcinoma and ductal carcinoma, cases of the linear type turning into malignancy is yet to be reported [2]. The malignant counterpart of SCAP, syringocystadenocarcinoma papilliferum, is an extremely rare neoplasm that occurs in association with SCAP/nevus sebaceous. It typically presents as a long-standing exophytic nodule, often associated with ulceration, secretion, or pain and occurs on the head and neck regions of elderly individuals [11]. Histologically, syringocystadenocarcinoma generally shows features of invasive or in situ adenocarcinoma. However, wide morphological variation may occur, including an invasive component of squamous cell carcinoma. Pagetoid migration of the neoplastic cells, mucinous ductal metaplasia, and ductal

changes analogous to those seen in the adenocarcinoma of breast have been described [12]. The tumor has low-grade malignant potential and recurs rarely. There have been no reports of distant metastases but locoregional metastases can occur [13].

The treatment of choice for SCAP is excision, which is necessary to confirm the diagnosis and to rule out the possibility of malignancy. The CO2 laser excision of SCAP is a treatment option in areas of the head and neck unfavorable for excision and grafting [14].

To conclude, we have reported an unusual case of adult-onset linear SCAP, which occurred de novo on the female breast. SCAP must be considered in the differential diagnosis of lobulated linear plaques on the breast of either sex.

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