



## CASE REPORT

# Sebaceous Carcinoma of the Vulva: Case Report and Review of Literature

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## Abstract

Extraocular sebaceous carcinoma is an uncommon malignant tumor that usually affects the head and neck. Despite being rich in sebaceous glands, vulvar sebaceous carcinoma is extremely rare. We report a case of vulvar sebaceous carcinoma in a 64-year-old woman that presented as an asymptomatic nodule and was successfully treated by wide local excision. The number of reported cases of vulvar sebaceous carcinoma is very small, more cases need to be collected in order to identify prognostic factors and appropriate management modalities.

## Keywords

Sebaceous carcinoma, Vulva, Local excision

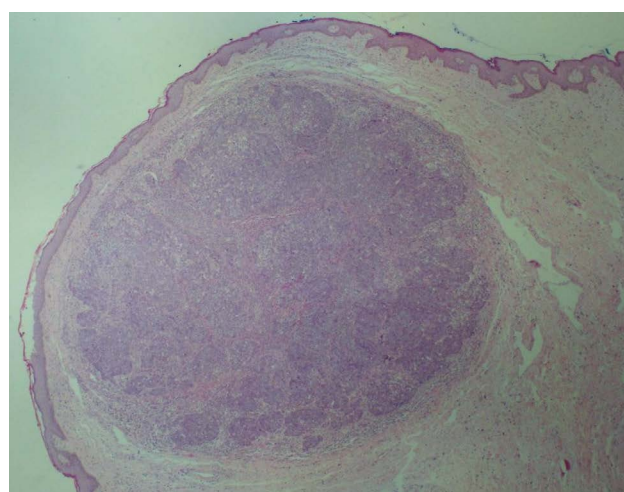
## Introduction

Sebaceous carcinoma (SC) is a well described type of malignant adenocarcinoma with sebaceous differentiation. However, this type of neoplasm is very rarely seen in the vulvar area. The most common malignant neoplasms of vulva are squamous carcinomas (87%) and malignant melanomas (6%) [1]. Today, there are only a few reported cases of sebaceous carcinoma of the vulva and they have varying clinical courses. Here, we report a case of vulvar sebaceous carcinoma that presented as a slow growing lump without any associated vulvar skin lesions or metastases that was successfully treated with local excision.

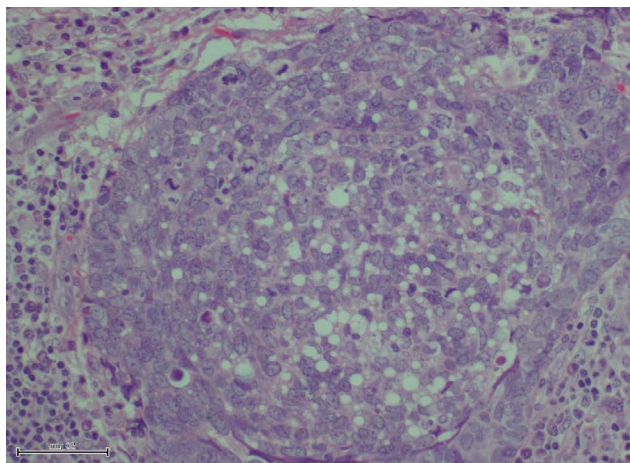
## Case Report

A 64-year-old woman was referred to our Vulvar clinic by her primary care physician for a vulvar lump that developed over 12 months. The lump was rela-

tively asymptomatic; there was no associated pain, pruritus, bleeding, or discharge. She had other comorbidities of diabetes, chronic renal impairment, and osteoporosis with a BMI of 18 kg/m<sup>2</sup>. Her other personal and family medical was unremarkable. Physical examination revealed a 2 × 1.5 cm erythematous nodule on the right labium minus (Figure 1). The surrounding area was clear. No enlarged inguinal nodes were detected. Examination of vagina and cervix were within age-appropriate normal limits. A Keyes punch biopsy was performed and was suggestive of adnexal neoplasm. Computed tomography did not reveal any abnormality in pelvis and groin nodes. In view of the patient's comor-



**Figure 1:** Microscopy-Nests and lobules of basaloid cells accompanied by a dense lymphoplasmacytic infiltrate low power. 2 × 1.5 cm erythematous nodule on the right labium minus (please locate the nodule).



**Figure 2:** Microscopy-Nests and lobules of basaloid cells accompanied by a dense lymphoplasmacytic infiltrate high power.

bidities, the lump was excised under local anesthesia. Histologically, the specimen showed skin with a dermal tumor composed of nests and lobules of basaloid cells accompanied by a dense lymphoplasmacytic infiltrate (Figure 2). The tumor cells showed a high nuclear to cytoplasmic ratio with pleomorphic, irregular nuclear membranes, vesicular nuclei and prominent nucleoli. The cytoplasm had a foamy and micro vesicular appearance, which was more prominent towards the center of the lobules (Figure 3). Occasional large intracytoplasmic vacuoles were present. There was brisk mitotic activity. Some areas showed comedo necrosis. The surface epithelium was focally ulcerated. Pagetoid spread of tumor cells to the overlying epidermis was noted, but no definite lymphovascular invasion was seen. These tumor cells were strongly and diffusely immunoreactive to p53. Combined with clinical presentation, these findings lead to the diagnosis of vulvar sebaceous carcinoma. All resection margins were uninvolved. At the time of reporting, the patient has now been recurrence-free for 22 months.

## Discussion

Extraocular sebaceous carcinoma most often occurs on the head and neck region. Although sebaceous glands are abundant and may be prominent on the vulva, only 10 cases of vulvar sebaceous carcinoma have been reported in the literature, to our knowledge. The presentation is often nonspecific with ulcer, plaque, nodules or exophytic tumors. The duration of symptoms ranges from 4 months to 2 years and the age at presentation from 31 to 89 years. Due to their nonspecific presentation, they are often recognized late. Vulvar sebaceous carcinoma was reported to be associated with Bowen's disease [2,3] and colonic adenocarcinoma, likely in the setting of Muir-Torre syndrome [4], although most cases were isolated [5].

Very little is known about the aggressiveness and natural history of vulvar SC. The rarity of the condition



**Figure 3:** Sebaceous carcinoma vulva-presented as yellowish nodule in the right inter labial sulcus.

limits the possibility of reliably estimating remission, recurrence, and survival rates. It is, hence, difficult to formulate treatment recommendations. The few cases in the literature have very differing clinical courses (Table 1). Khan, et al. reported a SC in a 49 years old woman with disease spread to the inguinal lymph nodes who underwent local excision and bilateral groin node dissection but recurred within 7 months [6]. In contrast to this, Kawamoto, et al. described a case that also had inguinal metastases at presentation; she was offered surgery with adjuvant radiotherapy and was recurrence free for the rest of the follow-up period [2]. Similarly to ours, a number of reported cases had not spread at presentation and did not recur after initial treatment [7-10]. To date, only one case was documented to have distant metastasis [11]. Commonly used treatment modalities include surgical excision (wide local excision or hemi vulvectomy with or without inguinal lymphadenectomy), chemotherapy and/or radiotherapy as adjuvant treatment or for recurrence.

It is difficult to identify clear prognostic factors due to the extremely small number of cases. Nonetheless, we note that only the case with local recurrence had local-regional lymphatic spread at presentation and was only treated surgically. The other case with lymphatic spread was successfully treated with surgery and adjuvant radiotherapy; no recurrence was noted by the end of follow up. This indicates a possibility of lymphatic spread being a predictor of recurrence and adjuvant radiotherapy an adequate treatment modality in such cases. Even though, extraocular SC is thought to be more aggressive than its periocular counterpart, these reports suggest that with appropriate management most case can be successfully cured.

## Conflict of Interest

I have no conflict of interest. This project was conducted at the KK Women's and Children's Hospital, 100 Bukit Timah Rd, Singapore. This project received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

Table 1: Literature with differing clinical courses.

Author	Year published	Age	Site	Symptoms duration	Lesion description	Size	Stage at presentation	Treatment	Follow-up period	Outcome
Current study	2018	64	Labium minorum	12 months	Exophytic lesion	2 × 1.5 cm	Stage I	Wide local excision	25 months	No recurrence
Thakur, et al. [5]	2017	55	Right vulva	NR	Multiple nodulo-ulcerative lesions	2.5 × 2 cm, 1 × 1.5 cm, 1 × 1.5 cm	Metastasis to right inguinal LN	Radiotherapy	NR	NR
Sullivan, et al. [8]	2016	76	NR	NR	Asymptomatic papule	5 × 3 mm	Stage I	Local excision with left ILND	10 months	No recurrence
Pusiol, et al. [7]	2011	51	Labium majorum	6 months	Exophytic lesion	2.5 × 1.5 cm	Stage I	Tumor excision	18 months	No recurrence
Khan, et al. [6]	2003	49	Labium majorum	NR	Papilloma-like	5 mm	Metastasis to inguinal LN	Wide local excision with bilateral ILND Laparotomy with PLND	7 months	Recurrence
Escalonilla, et al. [3]	1999	76	Labium majorum	4 months	Exophytic red and white nodule Associated with Bowen's disease	4 × 3 cm	NR	Adjuvant radiotherapy Excision biopsy Radical vulvectomy with bilateral ILND	12 months	No recurrence
Carlson, et al. [10]	1996	46	Labium majorum	NR	Sebaceous cyst-like Pruritic	NR	Stage I	Left radical hemivulvectomy with left ILND	31 months	No recurrence
Kawamoto, et al. [2]	1995	78	Labium minorum	6 months	Yellowish white nodule	2.5 × 1.5 cm	Metastasis to left inguinal LN	Simple vulvectomy with left ILND. Adjuvant radiotherapy	17 months	No recurrence
Jacobs, et al. [4]	1986	89	Vulva	12 months	Pink white plaques					
Ikuse, et al. [11]	1976		Labium majorum	NR	Red ulcerated tumor	4 × 4 cm	Metastasis to the lungs	NR	No recurrence	Demise
Rulon, et al. [9]	1974	31	Labium minorum	6 months	Raw, yellow, slightly indurated plaque	2 × 1.1 × 0.3 cm	NR	Excision	13 years 7 months	No recurrence

NR: Not Reported; LN: Lymph Node; ILND: Inguinal Lymph Nodes Dissection, PLND: Pelvic Lymph Nodes Dissection.

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