

## Case report

# Sebaceous carcinoma of the vulva: A case report and review of the literature☆☆☆☆☆



Stephanie A Sullivan, MD<sup>a,\*</sup>, Arthur-Quan Tran, MD<sup>a</sup>, Siobhan O'Connor, MD<sup>b</sup>, Paola A. Gehrig, MD<sup>a,c</sup>

<sup>a</sup> University of North Carolina at Chapel Hill, Gynecologic Oncology, United States

<sup>b</sup> University of North Carolina at Chapel Hill, Pathology, United States

<sup>c</sup> University of North Carolina at Chapel Hill, Lineberger Comprehensive Cancer Center, United States

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## 1. Introduction

Sebaceous carcinoma (SC) is a rare malignancy of the sebaceous glands that is most commonly found in the periocular area. While there are many sebaceous glands in the vulva, extraocular vulvar SC is especially rare with only eight previous cases described in the literature. Due to limited data, little is known about the appropriate management and prognosis of vulvar SC. Muir-Torre syndrome is an autosomal dominant disorder, a subgroup of hereditary nonpolyposis colorectal cancer, that has been found to be associated with some SC (Ponti et al., 2005). Lesions are frequently described as yellow-tan nodules that are larger and ulcerated. We present a case of vulvar SC in a patient who presented with a small, discrete papule treated with surgical intervention alone.

## 2. Case report

The patient, a 76-year-old Caucasian female, presented to her local provider for a routine exam and noted some mild discomfort in her groin area. She denied other symptoms such as pruritus, pain, bleeding,

ulceration, discharge, vaginal bleeding, or dysuria. Her provider biopsied a visible papule which returned initially has a squamous cell carcinoma. The patient was referred to the University of North Carolina for Gynecologic Oncology care. On histologic review of her biopsy, nodules of tumor were present in the dermis with extension into the epidermis. The tumor was composed of atypical basaloid cells with enlarged nuclei, scattered mitotic figures, with some cells demonstrating finely vacuolated, foamy, or clear cytoplasm, consistent with sebaceous differentiation (Fig. 1). Based on these findings, a diagnosis of vulvar sebaceous carcinoma was favored. The tumor measured 5 mm in horizontal extent and depth of invasion was 3 mm. Margins were negative, but close, with tumor <1 mm from one epithelial margin, 1.4 mm from the other epithelial margin, and 1.5 mm from the deep margin.

The patient's past medical history is remarkable for depression, hypothyroidism, gastroesophageal reflux, arthritis, and restless leg syndrome. She had a previous hysterectomy for benign leiomyoma and no other gynecologic history. She has no known family history of malignancies. General physical exam at the patient's consultation visit was unremarkable with a healing biopsy site, no lymphadenopathy, and no other lesions noted in and around the vulva. No residual tumor was palpable. Options for management, including surgical resection, were discussed. Due to close margins on the original pathology specimen, the patient opted for a re-resection around the original vulvar biopsy site with left inguinal lymphadenectomy. Pathologic results were negative for any residual tumor with no lymph node metastasis present.

The patient received no adjuvant therapy. Six months after her surgical intervention, she complained of left lower extremity edema and underwent lower extremity Doppler testing which returned positive for a deep venous thrombosis. Her symptoms resolved and her anticoagulation was stopped after 12 weeks of therapy. After 10 months of follow-up, she has no evidence of recurrence and her lower extremity symptoms have resolved.

## 3. Discussion

This case of vulvar SC is the ninth to be published to date in the literature and was similarly treated with only surgical excision. Extraocular SC was once thought to be highly aggressive; however, recent reports suggest similar prognosis for ocular and extraocular disease (Moreno et al., 2001; Dasgupta et al., 2009). Previous cases of vulvar SC vary in

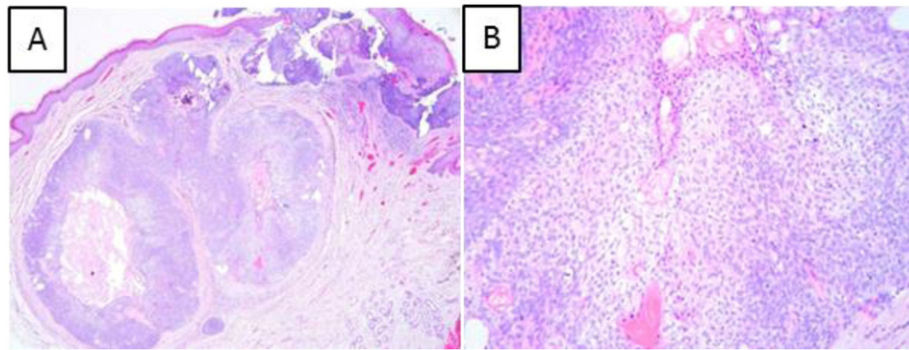
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\* Corresponding author at: University of North Carolina, Department of Obstetrics and Gynecology, Division of Gynecologic Oncology, Campus Box 7572, Chapel Hill, NC 27599, United States.

E-mail address: [sasulliv@unch.unc.edu](mailto:sasulliv@unch.unc.edu) (S.A. Sullivan).



**Fig. 1.** A. Nodules of tumor in the dermis with extension into the epidermis (HE,  $\times 40$ ). B. Basaloid cells and vacuolated/clear cells with scattered mitotic figures (HE,  $\times 200$ ).

clinical characteristics and treatment with overall prognosis being favorable with only one patient deceased and one with disease recurrence at the time of publication (Khan et al., 2003; Ikuse et al., 1976). The majority of reports include cases with follow-up intervals well within a 5-year time frame, with only one case reporting follow up at 13 years (Rulon and Helwig, 1974). Thus, the true disease free interval and 5-year overall survival is unknown.

The pathogenesis of extraocular SC remains poorly understood. There are no cases reported that appear to be associated with HPV infection, a common finding in squamous cell carcinoma. Additionally, only two cases appear to be associated with Bowen's disease and two with a strong family history suggesting possible Muir-Torre syndrome (Escalonilla et al., 1999; Jacobs et al., 1986; Carlson et al., 1996). If patients are diagnosed with a SC, a detailed family history should be taken, and consideration should be made for genetic counseling due to the known association of Muir-Torre syndrome and any SC. Care should be taken to detail pathologic characteristics for future cases to better understand the pathogenesis.

Unlike ocular SC, little is known about prognostic factors in vulvar SC. Older age, higher grade tumors, and distant metastasis have been described as poor prognostic factors. Importantly, lymph node metastasis have not been described as an independent prognostic factor for sebaceous carcinoma of the head and neck (Thomas et al., 2013). Of the eight cases previously described, two of the five women who had inguinal lymphadenectomy, had positive lymph node involvement (Khan et al., 2003; Kawamoto et al., 1995). The size of tumor in relationship to positive node status is variable (0.5 cm and  $2.5 \times 1.5 \times 1.0$  cm tumors with positive nodes and similarly large and small tumors with negative nodes) (Khan et al., 2003; Carlson et al., 1996; Kawamoto et al., 1995; Pusiol et al., 2011). Little information is available in the literature regarding lymph space invasion and depth of invasion, therefore no associations can be made. Only one previous case in the literature reports distant metastasis with metastatic disease to the lung. This patient presented with a  $4 \times 4$  cm red ulcerated tumor on the labia major and had died at the time of the publication with no additional information regarding her diagnostic workup or treatment course (Ikuse et al., 1976). Additional cases need to be collected to establish defined prognostic factors for vulvar SC as the prognostic factors appear to be distinct as compared to both squamous cell carcinoma of the vulva and ocular SC.

Due to the rarity of this disease, optimal treatment is unknown; however, it is reasonable to consider a similar approach to squamous

cell carcinoma of the vulva. Both of the patients who had lymph node metastasis received adjuvant radiotherapy (RT) with only one having a recurrence of disease within the time frame of reporting (Khan et al., 2003; Kawamoto et al., 1995). There were no other adjuvant therapies provided to previously reported cases. Although there is limited data, surgery appears to be the appropriate first intervention with a goal of complete excision of disease. Little is known about RT of sebaceous carcinoma (ocular and extraocular) with only 5.3% of patients receiving RT in the largest collection to date (Dasgupta et al., 2009). It is reasonable to consider adjuvant therapy with positive lymph node metastasis such as RT, however there is little information guiding this practice.

Vulvar SC is a rare disease that appears to have many clinicopathologic differences as compared to classic squamous cell carcinoma of the vulva. Care should be taken to report in detail any future cases to gain a better understanding of SC tumor behavior and response to therapy.

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