Basal cell carcinoma of the vulva: A rare case in the postmenopausal woman*

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ABSTRACT

Basal cell carcinoma (BCC) is a rare tumor of the vulva because BCC is frequently seen in skin sites exposed to sun. Vulvar basal cell carcinoma accounts for < 1% of all BCCs and 2-5% of all vulvar carcinomas. The standard treatment for vulvar BCC is surgical excision and it produces good survival outcomes.

This is a case report of an 83 year-old female who presented with a vulvar nodule associated with pruritus and bleeding. Initial vulvar biopsy revealed squamous cell carcinoma (SCCA). Patient underwent concurrent chemoradiation therapy for a Stage IIIB disease. Radical vulvectomy was subsequently done for tumor persistence. Surprisingly, the histopathology report of the persistent vulvar nodule revealed pigmented nodular basal cell carcinoma with lymphovascular invasion (LVSI). Patient was advised postoperative systemic chemotherapy but patient and relatives did not consent for the systemic treatment. Patient had no evidence of disease at 8 months post-surgery.

Keywords: basal cell carcinoma, vulvar carcinoma

INTRODUCTION

Basal cell carcinoma (BCC) is frequently seen in skin sites exposed to sun thus making the vulva a rare site of occurrence. Vulvar BCC accounts for < 1% of all BCCs and 2-5% of all vulvar carcinomas. BCC of the vulva is commonly seen in postmenopausal women with a mean age of 70 years. The standard treatment for vulvar BCC is surgical excision with favorable outcomes.[1]

This is a case report of an 83 year-old female who presented with a vulvar nodule associated with pruritus and bleeding. Vulvar biopsy revealed squamous cell carcinoma (SCCA) and patient underwent concurrent chemoradiation therapy for a Stage IIIB disease. Radical vulvectomy was subsequently done for tumor persistence and final biopsy revealed a rare case of pigmented nodular basal cell carcinoma.

CASE

An 83-year old widow from Cavite presented with a 9-month history of a right vulvar nodule associated with vulvar pruritus and bleeding. There was no history of radiation therapy and sun exposure to the vulva nor a family history of skin cancer. She previously worked as a seamstress at a local shop. Initial consult was done with a private physician and was advised observation and monitoring. Three months after the initial consult, there was a note of gradual enlargement of the labial mass. She then consulted a gynecologic oncologist. Physical examination revealed an 8.0 x 5.0 cm nodular mass involving the entire labia majora extending to the clitoris, external urethral meatus and lower 3rd of the vagina with perianal sparing. There were palpable inguinal lymph nodes measuring 3.0 x 3.0 cm on the right and 1.0 x 2.0 cm on the left. The rest of the genital and pelvic exam was essentially normal. Biopsy of the vulvar mass showed keratinizing squamous cell carcinoma (SCCA) (Figure 1). The mass was considered unresectable and was managed as a case of vulvar squamous cell carcinoma stage IIIB. Patient subsequently underwent chemoradiation: pelvic external beam radiation therapy (pEBRT) 5040 cGy plus vulvar EBRT 6540 cGy vulva with concurrent weekly cycles of Cisplatin (4 cycles) followed by vulvar EBRT boost 1800 cGy. One month after completion of therapy, there was still note of a 3.0 x 2.0 x 1.0 cm pink, flat lesion on the right labia majora near the clitoral fold with no gross urethral involvement and no inguinal lymphadenopathies (Figure 2). Assessment then was tumor persistence and patient subsequently underwent radical vulvectomy (Figures 3A & 3B). Histopathology of the persistent tumor showed pigmented nodular basal cell carcinoma with a 3.5 cm greatest tumor dimension at the right vulva (Figure 4).

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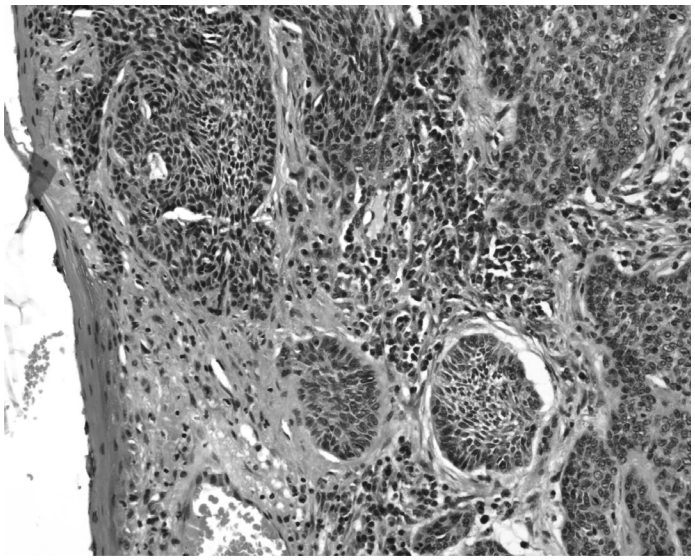

Figure 1. Histologic sections of the vulvar biopsy prior to chemoradiation showed nests of neoplastic basaloid cells with scant cytoplasm and round to elongated hyperchromatic nuclei showing nodular pattern with prominent palisading and are surrounded by a typical loose stroma and cleft like retraction spaces.

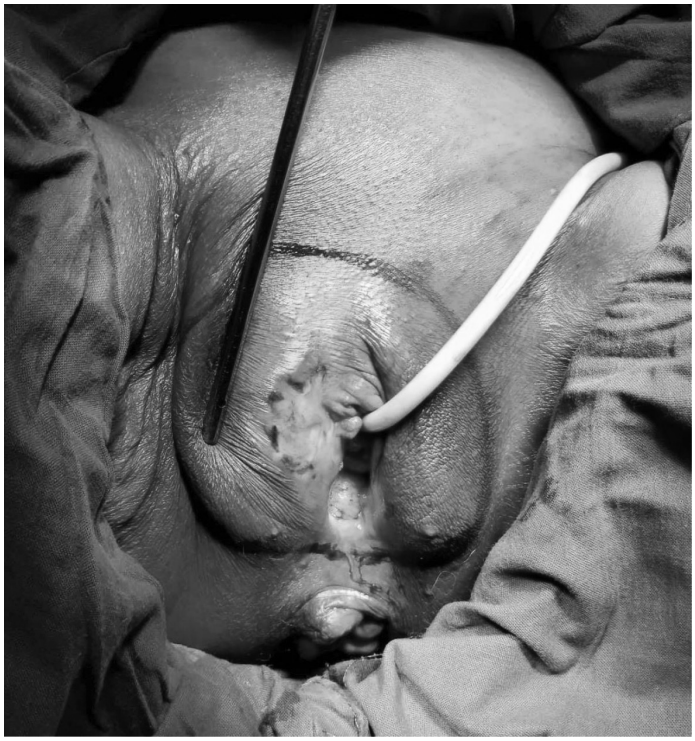

Figure 2. Vulvar lesion after chemoradiation

There was also note of lymphovascular space invasion (LVSI) with no extension to the perineural, deep soft tissue and peripheral margins. Postoperative evaluation after one month revealed a well-healed surgical site (Figure 5). Due to the presence of LVSI, adjuvant systemic chemotherapy was advised but patient and family opted for supportive care. Patient had no clinical evidence of disease at 8 months post-surgery.

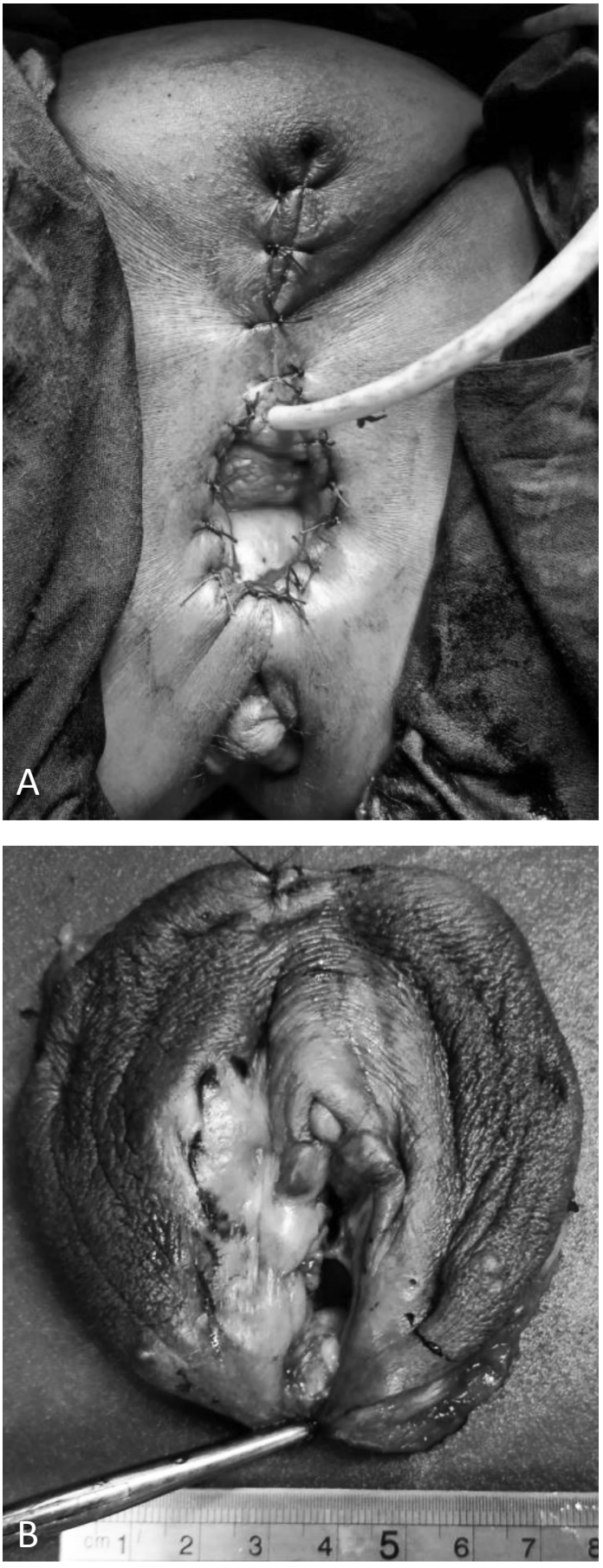

Figure 3. Vulva (A) and specimen post vulvectomy (B)

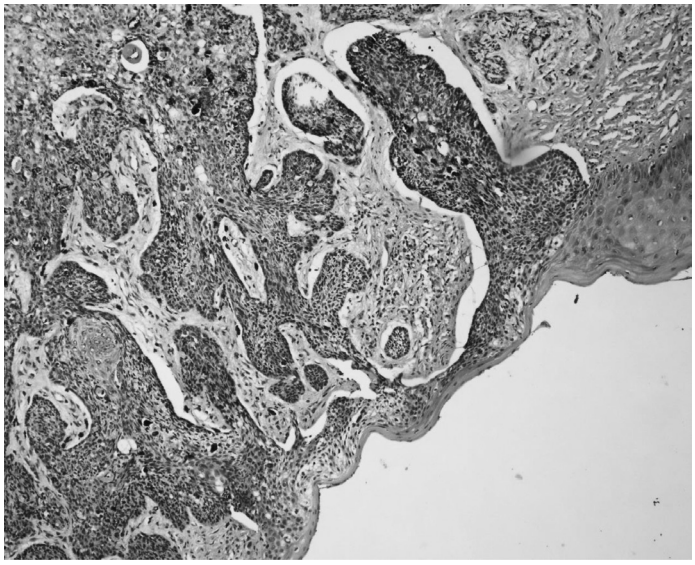

Figure 4. Histopathology of the vulvectomy specimen. Epidermal attachment is present which is typical of the tumor. The pigmented appearance is due to accumulation of melanin by dermal macrophages. Cleft like-retraction spaces present due to accumulation of stromal mucin.

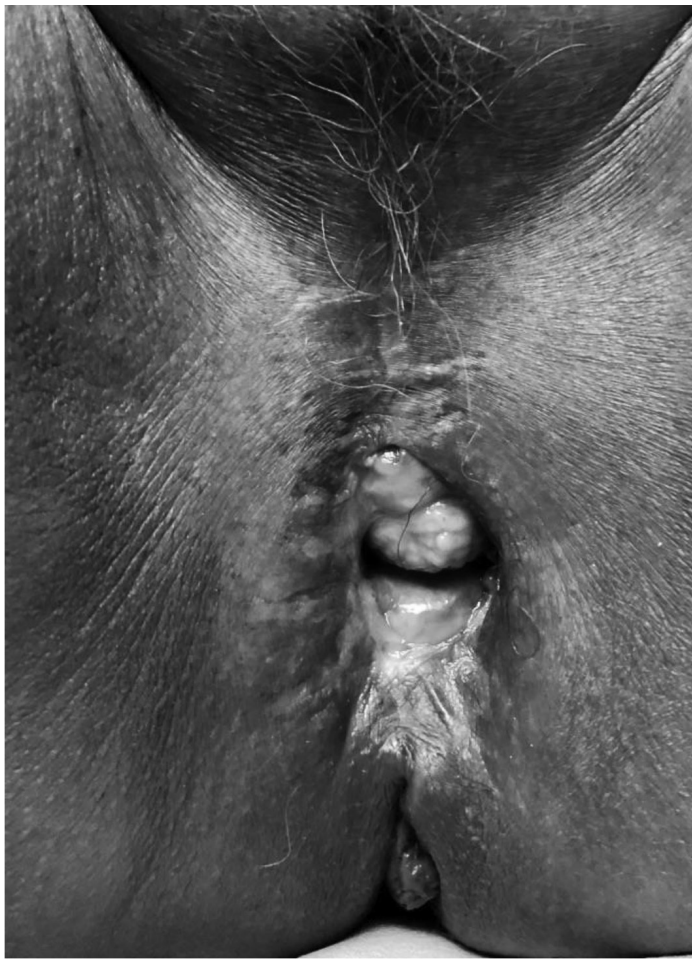

Figure 5. Post-operative examination of the vulva after one month showed no evidence of tumor recurrence with good wound healing.

DISCUSSION

Vulvar basal cell carcinoma (BCC) is a rare tumor with approximately 250 publications, mostly as case reports, worldwide. Vulvar BCC represents 2-3% of all vulvar cancers.1 In the Philippines, there is no published case report of vulvar BCC in the Herdin database.

The etiology of vulvar BCC is not clear. Risk factors identified are the same as those for cutaneous BCC: radiation exposure, arsenic ingestion, immunodeficiency, chronic irritation, hereditary skin conditions and nevoid basal cell syndrome.[1-5] The index case has no known predisposing factors, except for her postmenopausal age which has been reported to have a strong association with BCC development. In two large series of vulvar BCC, the age range of presentation is 20 to 96 years-old with a mean age of 70 years.[1-6] The evidence for viral association is less convincing for BCC than squamous cell carcinoma of the vulva.

The presenting signs and symptoms of vulvar BCC are non-specific causing a delay in diagnosis. The onset of symptoms to diagnosis ranged from a few months to as long as 9 years. The most common presenting complaints are vulvar pruritus (52%) and ulceration (50%).[2] Bloody vaginal or vulvar discharge is also common. Others present with vulvar irritation, soreness or a palpable nodule. The vulvar lesions may mimic inflammatory dermatosis such as eczema or psoriasis. Other differential diagnoses include chronic folliculitis, melanocytic lesions and candidiasis. In patients with dark complexion, vulvar BCC may present as hyperpigmentation thus mimicking melanoma or seborrheic keratosis.[3-4,7]

Diagnosis of vulvar BCC is confirmed thru biopsy. Histologic appearances are the same with BCCs of the skin. There are four histologic variants described in the literature. The nodular variant which is present in the index case is the conventional or prototypical type. The histologic appearance has a fibromyxoid stroma which is an integral part of the tumor. The stromal dependent neoplastic cells may explain the absence of metastasis in majority of BCC cases described in the literature.[9] The number of mitosis and melanin pigmentation reflects the rate of apoptosis while trauma and inflammation are responsible for the degree of atypia. The keratotic variant is the same as the nodular type but shows central squamous differentiation with keratinization and scanty stroma with no follicular differentiation. The superficial variant is often difficult to interpret due to tissue fragmentation, tangential sectioning, crush artefacts and cell smearing. In these cases, differentials include an inverted follicular keratosis, seborrheic keratosis, basaloid follicular hamartoma, pseudo-bowenoid condyloma acuminatum

or vulvar intraepithelial neoplasia (VIN) of basaloid type. The last histologic variant is the diffuse infiltrative variant which has an increased likelihood of local recurrence. [8-10]

BCCs are generally slow-growing but they are locally aggressive and confined to the dermis. Metastasis is rare occurring only in 0.1% of reported cases.[1-6] The most common site of metastasis is the inguinal lymph node. The tumor has a tendency to grow along the paths of least resistance which is the presumed reason for invasion of bones, cartilage and muscle in its late manifestation.[6]

Wide local excision with biopsy-proven negative margins of at least 1 cm remains the standard of care for primary and recurrent local vulvar BCC.[1,5,6] There is a 20% chance of local recurrence if there is positive resection margins.[1-6] Mohs micrographic surgery must be considered for recurrent disease and for cases where anatomic preservation is critical such as clitoral involvement. Routine inguinal lymphadenectomy is not warranted since tumors do not seem to readily metastasize, except for large tumors with deep invasion. Post-operative radiotherapy does not appear to have any effect on the recurrence or survival rate. Chemotherapy may be employed in cases with metastatic disease.[6]

SUMMARY

Vulvar basal cell carcinoma is a rare type of vulvar carcinoma. Awareness of its presentation in the genitalia is important as early detection is critical to allow complete surgical resection. Health care providers must have a high index of suspicion for such lesions in the vulva and perform a biopsy on any lesion on the vulva for postmenopausal woman. Wide local excision is the preferred treatment for primary

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