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Case report

The tell-tale heart: A case of recurrent vulvar carcinoma with cardiac metastasis and review of literature

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ABSTRACT

A 50-year-old female was diagnosed with vulvar cancer treated with left partial vulvectomy and bilateral lymphadenectomy. Ten months after her surgery, she presented with increased labial swelling, pain and discharge. Biopsy confirmed recurrence of squamous cell vulvar carcinoma. Incidentally, on restaging radiographic scans, she was found to have a large right ventricular mass which, after surgical debulking, was shown to be a squamous cell cancer of vulvar origin. She was commenced on chemotherapy with carboplatin and paclitaxel along with concurrent radiation therapy. Restaging PET scan showed persistent metastatic disease. She was switched to Cisplatin/Taxol after having hypersensitivity reaction to Carboplatin. She received 5 cycles with progression of disease in the follow up scans. She then received Nivolumab for 2 cycles. The patient then opted for comfort directed care given worsening functional status and progression of disease on repeat imaging. Secondary cardiac tumors are very rare and not extensively studied in oncology. Therefore, optimal management is not entirely clear. It is extremely rare for vulvar cancer to metastasize to the heart and only two cases have been reported in the literature. However, vulvar cancer metastasizing to the right ventricular cavity and endocardium has not been described before. We believe that this is the first ever such reported case.

1. Introduction

Vulvar carcinoma is a rare gynecological malignancy with a propensity to recur locally in most cases. However, distant recurrences can occur. We describe a case of 50-year-old Caucasian female who had intra-cardiac and pulmonary recurrences of a surgically resected FIGO Stage 1 squamous cell carcinoma. This case is unique due to its exceedingly rare presentation and challenging management.

2. Case

The patient is a 50-year-old nulliparous female with history of well controlled asthma and cigarette smoking who initially presented to the oncologist with recently diagnosed squamous cell cancer of the vulva. At the time of diagnosis, her symptoms included redness, itching and burning around the vulva unrelieved by the use of various antifungal and steroid creams. She was then seen by her gynecologist and a left vulvar biopsy was performed which showed keratinizing moderately differentiated infiltrating squamous cell carcinoma. Staging computed tomography (CT) and magnetic resonance imaging (MRI) scans showed localized disease without pelvic lymphadenopathy and no distant metastases. She underwent left partial vulvectomy and bilateral inguinal lymphadenectomy since intraoperative sentinel lymph node could not be identified with isosulfan blue injection. Microscopic examination of the resected specimens revealed 9 mm deep, 2.1 cm moderately differentiated, squamous cell cancer and a focus of positive cancerous margin adjacent to the urethral meatus. All the seven resected lymph nodes were negative for any cancer. Given the positive margin, she underwent distal urethrectomy three months after the initial diagnosis which failed to reveal any tumor.

The patient, six months after initial diagnosis of FIGO (International Federation of Gynecology and Obstetrics) stage 1b, T1b N0 M0, vulvar carcinoma, developed swelling of the labia and increased drainage around the genital area. She attributed the swelling to postoperative vulvar lymphedema and declined further evaluation including biopsy. However, over the next 4 months, her symptoms worsened with increasing swelling and pain in the genital area. An examination under anesthesia demonstrated bilateral labial swelling, erythema, ulcerated lesions and serosanguinous discharge. Biopsy showed recurrence of invasive vulvar squamous cell cancer (Fig. 1a).

A computed tomography (CT) scan of chest, abdomen & pelvis for restaging interestingly demonstrated a 6.8 × 4.9 × 6.2 cm mass in the

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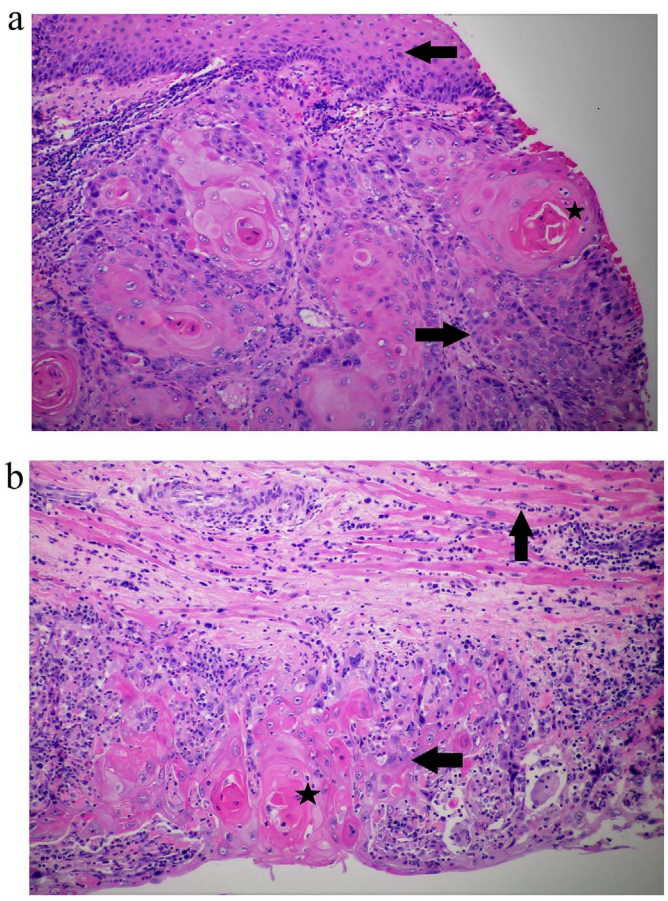

Fig. 1. a Microscopic view of vulvar biopsy demonstrating squamous cell carcinoma (rightward black arrow) and several keratin pearls (black star). Normal vulvar squamous epithelium is indicated by the leftward black arrow. b Microscopic view of RV mass biopsy showing infiltration by squamous cell cancer (leftward black arrow) along with keratin pearls (black star). Normal myocardium is indicated by black upward arrow.

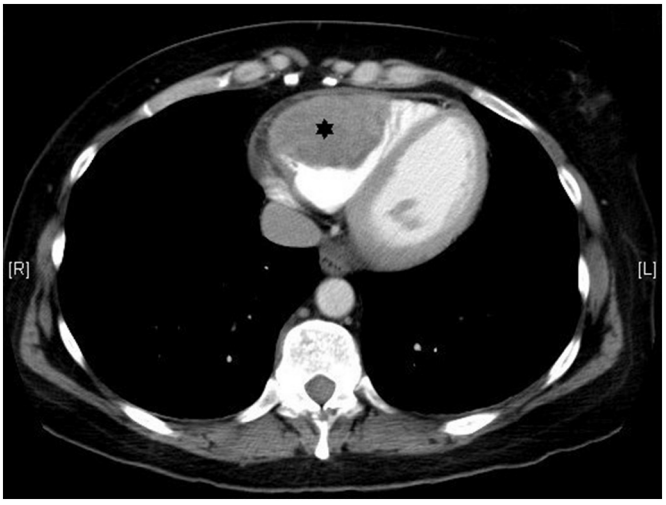

Fig. 2. CT scan of the chest showing dilated RV with a large hetergenously attenuated intracavitary mass with lobulated contours (black star).

right ventricle (RV) (Fig. 2). Echocardiogram (Fig. 3) confirmed the presence of a large RV mass adherent to the free wall extending from the base to the apex with a 2 × 1.8 cm mobile component. In addition, CT scan showed multiple pulmonary emboli and multiple sub-centimeter and one 1.6 cm cavitary pulmonary nodules concerning for metastatic disease. CT scan of the pelvis showed bilateral inguinal adenopathy and left vulvar thickening, consistent with recurrent

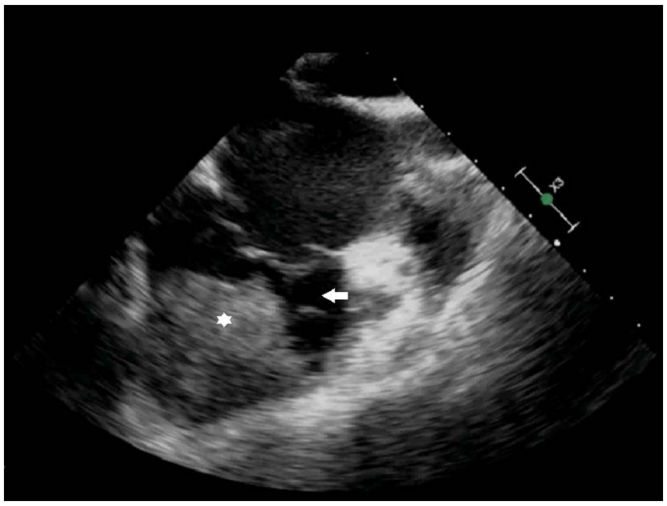

Fig. 3. Echocardiogram showing the same mass (white star) and right ventricular cavity (white arrow).

disease.

A differential diagnosis of intra-cardiac thrombus, primary cardiac tumor such as myxoma or sarcoma and metastatic cardiac tumor were considered. Particularly interesting was the lack of cardiac or pulmonary symptoms despite a large intra-cardiac mass. Therapeutic anticoagulation with intravenous unfractionated heparin was commenced. Immediate cardiothoracic surgery evaluation was undertaken given the size and location of the RV mass and high risk of embolization. A decision to surgically resect the mass was made, however, to the surprise of the surgeon, the mass was found to be densely adherent to the RV muscular wall without associated thrombus. Therefore, it was not amenable to surgical resection and only debulking was performed which showed well differentiated squamous cell cancer consistent with vulvar origin (Fig. 1b) confirming recurrent vulvar cancer with cardiac and pulmonary metastases.

Postoperative course was complicated by anemia treated with red cell transfusion and urinary tract infection treated with antibiotics. Towards the end of her two-week hospitalization, she developed new-onset atrial fibrillation controlled with beta blockers. In regards to her vulvar cancer, chemotherapy and radiotherapy were contemplated. She received her first dose of weekly chemotherapy regimen, consisting of low dose carboplatin and paclitaxel as inpatient which was continued after discharge for a total of 5 doses. Furthermore, concurrent pelvic radiotherapy was given with chemotherapy. Since most of her symptoms were related to local recurrence in the form of erythema, induration and extreme pain of the external gentalia, therefore, even though the patient had metastatic vulvar carcinoma, the rationale for radiotherapy was primarily palliative or pseudo-curative (radiation dose was 180 cGy).

After completing chemo-radiotherapy, a positron emission tomography (PET) scan showed residual vulvar disease with inguinal lymphadenopathy, persistent RV mass and increased pulmonary nodule size with peak standardized uptake values (SUV) of 8.1, 6.4 and 2.7, respectively. Full dose carboplatin and paclitaxel was then initiated but after her first full dose carboplatin, she developed an immediate hypersensitivity reaction. Carboplatin was switched to cisplatin. Her disease remained stable after 3 cycles of Cisplatin and Taxol. Bevacizumab was contemplated but in light of her recent open heart surgery and sternotomy, and risks of non-healing and bleeding associated with it, decision was made to defer it. She then developed peripheral neuropathy which became so severe that Chemotherapy had to be stopped after 5 cycles of cisplatin/Taxol. Repeat imaging studies at this point revealed progression of metastatic disease in lungs. The decision was made to stop chemotherapy. The next generation sequencing of her