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Case report

Rare phalanges soft tissue and bony metastasis in vulvar squamous cell carcinoma: Case report

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ABSTRACT

Vulvar cancer accounts for 0.3 % of new cancer cases within the Unites States. Metastatic vulvar cancer with disease beyond the pelvis is rare and has a poor prognosis. Data on primary treatment including systemic treatments for distant metastatic vulvar disease is limited due to rarity and lack of clinical trials. The purpose of this article is to present an atypical presentation of recurrent vulvar squamous cell carcinoma with metastasis to phalanges soft tissue and bone, clavicle and to the lungs and intracranial space.

1. Introduction

Vulvar cancer accounts for about 5–8 % of all gynecologic malignancies (Abu-Rustum, 2024). Risk factors for vulvar cancer include increasing age, HPV status, smoking, inflammatory and immunodeficiency conditions. Clinical presentation usually reveals vulvar mass, symptoms of pruritis, bleeding, or pain. Vulvar cancer initial diagnosis is made by tissue biopsy with squamous cell carcinoma being the most common histologic type. Initial vulvar cancer treatment typically involves radical local excision and surgical staging including groin node lymphadenectomy (Abu-Rustum, 2024). Metastatic vulvar cancer with disease beyond the pelvis is rare and is accompanied with a poor prognosis. The main routes of metastasis are by local spread, lymphatics, or hematogenous spread (Tolia and Tsoukalas, 2012). Typically, vulvar cancer spreads through the lymphatic system to the superficial inguinal lymph nodes followed by the deep femoral inguinal lymph nodes, making lymph node status the greatest prognostic factor for survival. Distant disease, which lies outside the normal spread, has a very poor prognosis. Primary treatment for distant metastatic disease can include radiation therapy, palliative treatment, and/or systemic chemotherapy however data on systemic treatments is limited due to rarity of disease (Prieske et al., 2016). Similarly, data on treatment options for recurrent vulvar cancer is limited resulting in no standard of care due to low incidence and lack of reported cases (Adhikari et al., 2021; Bizzarri et al., 2017; Kazdan and Dunn, 2019). Evidence has been extrapolated from cervical, anal and other squamous cell cancer treatments (Abu-Rustum, 2024). Treatment highly depends on site and extent of recurrence. Here we present an unusual case of recurrent vulvar squamous cell carcinoma with metastasis to phalanges soft tissue and bone along with lungs and intracranial space metastasis sites.

2. Case presentation

A 52 year old postmenopausal woman presented to clinic for a painful vulvar mass noted for the past four months which had been progressively worsening and uncomfortable, accompanied by a left groin mass for the previous two weeks. She was seen in the oncology office and expeditiously set up for surgical intervention. Initial surgery included partial radical vulvectomy with selective left inguinal lymphadenectomy. Gross surgical findings included left superficial inguinal lymphadenopathy measuring 4.5 × 4 cm, a 5 cm left sided vulvo-vaginal mass extending across the midline and posteriorly about 2.5 cm from the rectum and 5 cm into the vagina. The lesion was excised with 1 cm borders. Then, the 5 cm left inguinal lymph node was removed followed by two smaller lymph nodes, each about 1 cm in size. The remaining adipose tissue near these lymph nodes was also removed which ultimately contained three smaller lymph nodes.

Pathology showed HPV (p16) associated squamous cell carcinoma of the left vulva with negative margins and a depth of invasion of 12 mm. Closest margin to invasion was along the left edge with a 2 mm negative margin. All margins were negative for HSIL or VIN. The 5 cm lymph node revealed squamous cell carcinoma with extracapsular extension. Each of the 1 cm lymph nodes showed squamous cell carcinoma without extracapsular invasion. The remaining three lymph nodes extracted from the adipose tissue demonstrated no cancer.

She then completed primary radiotherapy with cisplatin chemo

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sensitization. Tumor was negative for PDL-1 immunostaining, so immunotherapy was not indicated. Her radiation therapy was intermittently held due to vulvar desquamation and pain and she also had a delayed chemotherapy treatment for thrombocytopenia. She completed radiation, targeted to the vulva, in May of 2022 and underwent 28 fractions, receiving a total of 6020 centigray (cGy). She completed chemotherapy and was then being seen for surveillance every two to three months for disease recurrence.

After initial surgery, radiation and chemotherapy completion, she then presented to the emergency department 14 months later despite negative surveillance visits, for dizziness and vomiting and imaging revealed lesions suspicious for brain and pulmonary metastasis. She underwent right suboccipital craniotomy, resection of intradural metastatic disease, microscopic dissection, duraplasty, cranioplasty and recovered without complication. The pathological analysis of the metastatic brain lesions were consistent with those of the primary vulvar tumor. She received stereotactic radiation therapy in the outpatient setting for brain metastasis. Post treatment CT imaging revealed progression of brain metastases as well as development of lung and bony metastases of the hip. The plan was then to initiate chemotherapy with carboplatin, paclitaxel, bevacizumab. She was then admitted for intractable pain due to widely metastatic vulvar cancer complications. During this hospitalization her left ring finger was noted to be dusky in appearance, necrotic, and painful. An X-ray of the hand revealed suspicious metastatic lesion to 4th digit. The finger subsequently began to bleed while the patient was hospitalized and the decision was made to amputate for symptomatic control of bleeding (Figs. 1 and 2). Pathology revealed metastatic vulvar squamous cell carcinoma, HPV associated. Tumor mass was in soft tissue and ulcerated the epithelium, invading into the bone. To our knowledge, this is the first case of soft tissue metastasis from vulvar cancer to the phalanges requiring amputation. Shortly after this hospital stay, the patient represented with coughing that resulted in a clavicular fracture. Imaging revealed a new 4 cm lytic mass on her left clavicle (Fig. 3). She was then placed on a cisplatin, paclitaxel, bevacizumab regimen with outpatient palliative care services.

3. Discussion

Vulvar carcinoma constitutes 3–5 % of all female genital tract malignancies peaking around 70 years of age (He and Xiao, 2024). There are two pathways in which vulvar squamous cell carcinoma occurs, those being HPV mediated and HPV independent. It has been shown that about 60 % of squamous cell carcinoma of the vulva arises from the independent pathway (Bigby et al., 2016). Prognosis depends on surgical pathologic stage, histologic differentiation and metastasis to lymph nodes with most frequent distant metastasis being to the liver and lungs (He and Xiao, 2024). The incidence of vulvar cancer with distant metastatic disease beyond the pelvis appears to be an uncommon entity based upon the small volume of cases reported in the literature (Maggino et al., 2000). Brain metastasis in gynecologic malignancy is limited with highest likelihood occurring in choriocarcinomas (Vázquez et al., 2007). Prevalence of brain metastasis in vulvar cancers appears to be unknown with few reported cases (Vázquez et al., 2007; Jaeger et al., 2020). Treatment when distant metastasis is involved usually consists of a platinum-based combination chemotherapy (How et al., 2021; Cormio et al., 2009). Treatment options are often palliative, experimental and or extrapolated from other more common cancers and focuses on improving quality of life. Prognosis is poor for distant metastatic vulvar cancer cases or in cases of recurrence. It has been reported that the two-year overall survival rate is only 11 % for these patients (Adhikari et al., 2021). Metastasis of vulvar squamous cell carcinoma primarily occurs through lymphatics with spread to bone being very rare and based on limited reported cases (Fischer et al., 2005).

Fig. 1. Vulvar squamous cell carcinoma with metastasis to left ring finger after amputation.

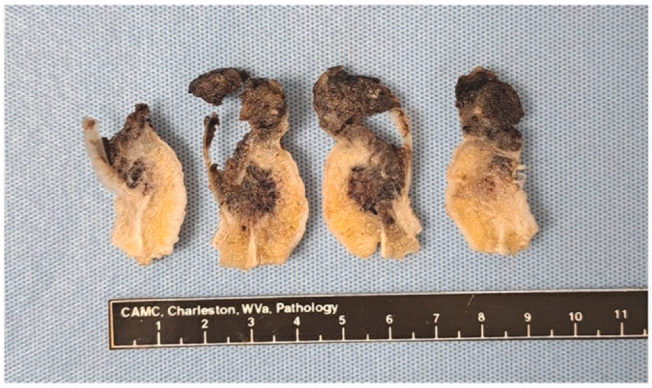

Fig. 2. Cross section of vulvar squamous cell metastasis to left ring finger showing bone invasion.

In our report, there was 14 months of remission following initial radiation and chemotherapy after excision and lymphadenectomy. She underwent craniotomy with dural metastasis resection and was then started on platinum based therapy with poor tolerance. It was then that bony metastasis was first seen in the phalanges and clavicle. Amputation was performed confirming metastasis. Prognosis was poor and she subsequently opted for palliative care.

Although this recurrence is atypical, it highlights the importance of monitoring metastasis in vulvar cancer treatment and importance of a thorough physical exam. There is need for additional studies to determine best treatment options for metastatic and recurrent vulvar cancer especially to rare sites of metastasis as presented in this case.

4. Conclusion

All cases of distant vulvar cancer metastasis should be reported in the literature until clinical studies can be completed to establish standard of care treatments specific to vulvar cancer and improve patient outcomes. From this case, we recommend soft tissue biopsy for any suspicious lesions in the setting of vulvar cancer, no matter how uncommon or underreported the metastatic site is. To the best of our knowledge from literature review, this is the first reported case of recurrent vulvar carcinoma metastasis to phalanges soft tissue and bone along with clavicle, intracranial space and lung metastasis.

Ethics approval

Our institution does not require ethical approval for reporting

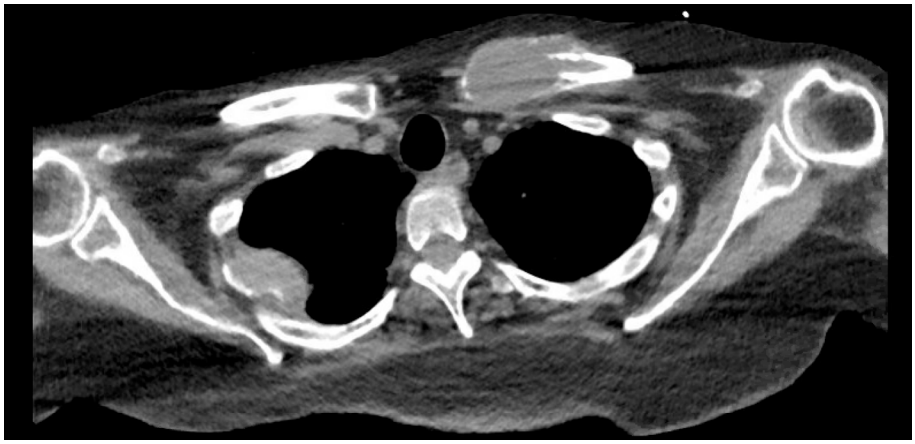

Fig. 3. Fracture and metastasis of left clavicle on CT.

individual cases or case series.

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Informed consent

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CRediT authorship contribution statement

Landon Foulger: Writing – review & editing, Writing – original draft, Conceptualization. **Kelly Simmons:** Writing – original draft. **Michael Schiano:** Writing – review & editing, Supervision, Conceptualization. **Stephen Bush:** Writing – review & editing, Supervision.

Declaration of competing interest

The Authors have no know competing financial interests or personal relationships that could have influenced the work reported in this paper.

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