SKIN

BRIEF ARTICLE

Vulvar Porocarcinoma with Inguinal Nodal Recurrence: A Case Report

Hannah J Porter, MD, MBA, MS[1], Nadeem Marghoob, DO[1], Hayden Christensen, BA[2], Matthew Dinehart, MD[3], Conor O'Neill, MD[4], Mark Wick, MD[5], Laura Greene, MD[3], Christine H Weinberger, MD[1]

[1] Division of Dermatology, University of Vermont Medical Center, Burlington, VT, USA
[2] University of Vermont Robert Larner College of Medicine, Burlington, VT, USA
[3] Department of Pathology and Laboratory Medicine, University of Vermont Medical Center, Burlington, VT, USA
[4] Division of Surgical Oncology, University of Vermont Medical Center, Burlington, VT, USA
[5] PRW Laboratories, PLC

ABSTRACT

Introduction: Porocarcinoma (PC) is an uncommon, aggressive malignancy of the sweat gland apparatus characterized by diverse clinical presentation and challenging histopathology. Patients diagnosed with PC face a grim prognosis due to high rates of recurrence and metastasis.
Case Report: We present a case of a vulvar porocarcinoma initially managed by surgical excisions, achieving clear margins. Subsequent surveillance revealed regional lymph node metastasis, prompting lymph node dissection.
Discussion: Here, we highlight the difficulty in recognizing and managing PC. While surgical excision remains the mainstay of treatment, there exists no consensus on adjuvant therapies and the role of sentinel lymph node biopsy at diagnosis. Given its rarity, diagnostic complexities, and aggressive behavior, PC warrants a collaborative approach to maximize patient outcomes.

INTRODUCTION

Porocarcinoma (PC) is an uncommon and aggressive skin cancer originating from the intraepidermal component of the sweat gland apparatus. It presents as a firm, erythematous, violaceous or skin colored papule, plaque or nodule.[1] Other designations include epidermotropic eccrine carcinoma, eccrine porocarcinoma, malignant eccrine poroma, and malignant hidroacanthoma simplex.

PC accounts for approximately 0.005–0.01% of all malignant cutaneous neoplasms, evenly distributed between males and females.[2,3] It predominantly affects the elderly, with incidence increasing with age, usually appearing in individuals in their 7th or 8th decade of life. Common anatomic locations affected are the head and neck, trunk, and extremities.

CASE REPORT

A 68-year-old female with no significant past medical history presented with a rapidly

growing and painful lesion of the right labia majora. Physical exam revealed a 1.3cm x 1.0cm flesh colored to slightly pink plaque with some central erosion and lack of a visible punctum (**Figure 1**). Given the diagnostic uncertainty and rapid growth, surgical excision was recommended and performed with 0.2cm margins the following day. Pathology revealed an irregular, invasive tumor composed of basaloid poroid cells displaying ductal differentiation and marked cytologic atypia. Although the tumor had squamous differentiation, given the well-developed ductal differentiation, a PC was favored (**Figure 2**). Given the unusual site, the tumor was sent for expert consultation and diagnosis was confirmed.

Re-excision with Mohs micrographic surgery (MMS) was recommended; however, the patient ultimately elected for standard re-excision, which was performed with 5mm margins. Follow-up examinations with clinical lymph node exams were performed every 6 months without evidence of recurrence. However, approximately 2 years after initial diagnosis, enlarged lymph nodes of the right inguinal basin were incidentally detected on a CT scan, raising concerns for recurrent malignancy. Ultrasound-guided lymph node biopsy was consistent with metastatic PC. Subsequent staging PET-CT was performed and revealed hypermetabolic lymphadenopathy in the right external iliac chain and inguinal region, consistent with regional metastasis. The patient subsequently underwent bilateral pelvic lymph node dissection.

DISCUSSION

Vulvar PC, as observed in our case, is highly uncommon but has been documented several times in the literature.[4-6] The precise pathogenesis underlying PC remains unclear, although it is favored to arise from the acrosyringium, the intraepidermal spiral duct of the sweat gland apparatus.[7] PC may occur de novo or may develop from a pre-existing poroma in up to 50% of cases.[8] Some studies propose that UV exposure is the culprit behind PC tumorigenesis due to PC's predilection for sun-exposed areas. Chronic immunosuppression is also noted as a risk factor, and there are reports of chemical exposures being linked to PC development. More recently, genetic studies have revealed recurrent gene translocations in PC, including YAP1-MAML2 and YAP1-NUTM1, opening new avenues of study for both prognostic markers and potential treatment options.[9]

The histopathologic findings of PC are varied and can prove a challenge to arrive at the correct diagnosis. They tend to be irregular, invasive tumors composed of basaloid poroid cells displaying ductal differentiation and marked cytologic atypia. However, they can appear bland with monomorphic cuboidal poroid cells.[10] They often have an increased mitotic index and necrosis is common. Squamous differentiation is also a frequent finding and can be difficult to differentiate from squamous cell carcinoma as seen in our case. Given the diagnostic difficulty PC poses on H&E, immunohistochemistry (IHC) is a useful tool that can aid in the diagnosis of PC. Carcinoembryonic antigen (CEA) and epithelial membrane antigen (EMA) IHC are both useful in identifying ductal differentiation present in PC. CD117 (CKIT) IHC has been documented as a more specific marker for poroid tumors and one which, when positive in the neoplastic cells, can help favor PC over SCC.

Wide local excision (WLE) remains the mainstay of PC treatment with recurrence rates of approximately 20%, regional lymph node metastasis rate of 20%, distant

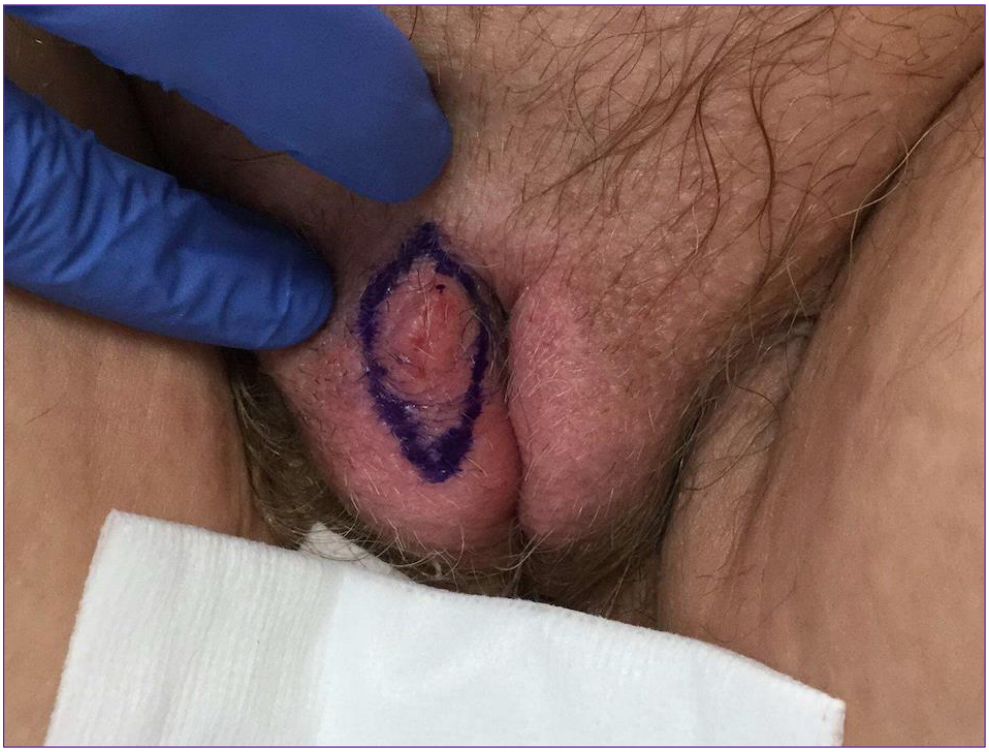

Figure 1. 1.3cm x 1.0cm flesh colored to slightly pink plaque with some central erosion and lack of a visible punctum.

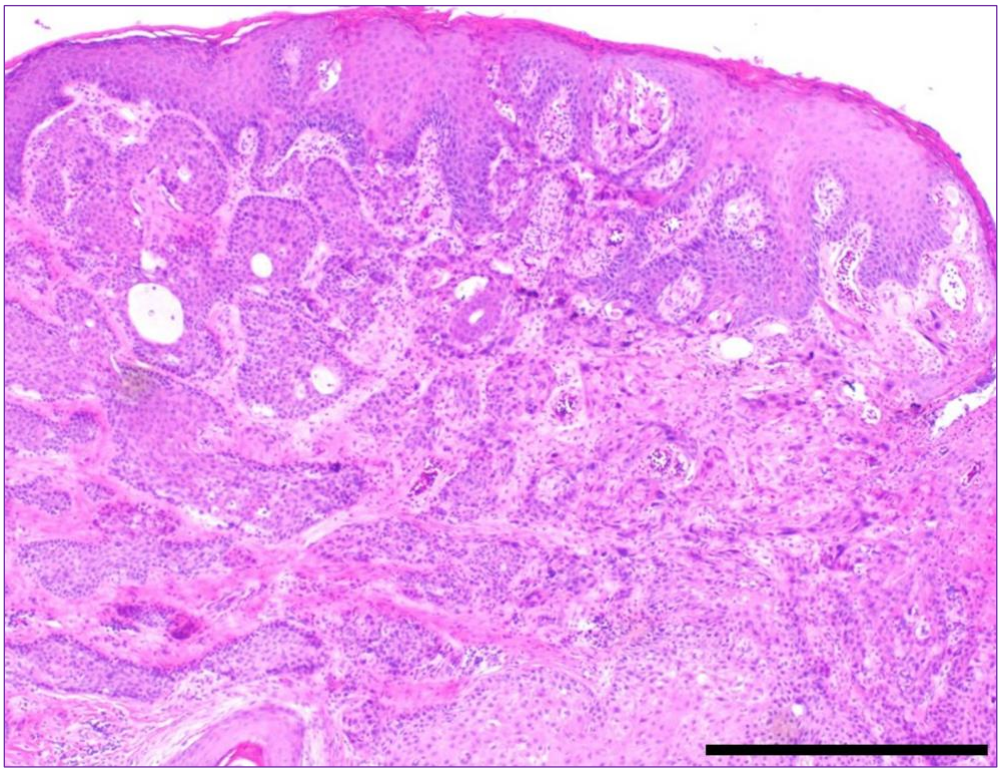

Figure 2. Basaloid poroid cells displaying ductal differentiation and marked cytologic atypia (H&E, original magnification x10, Scale bar=500µm).

metastasis rates of 12%, and overall mortality rates of approximately 7%.[11] Other treatment options include chemotherapy, interferon alpha, radiation therapy, and mohs micrographic surgery (MMS). Though data is limited with roughly 47 cases treated by MMS, recurrence rates appear to be lower than with WLE with only 2 reported cases of recurrence to regional lymph nodes after MMS.[11] There is no consensus regarding the role of sentinel lymph node biopsy (SLNB) in PC nor are there consensus statements on the role of adjunct radiotherapy, chemotherapy, or immunotherapy in the setting of recurrent or metastatic disease.

The majority of studies categorize PC as having a poor prognosis with high rates of recurrence and metastasis. For this reason, some authors suggest SLNB for some or all patients with PC, though the prognostic and therapeutic role of SLNB remain unclear. In one of the larger case series evaluating 13 patients with eccrine PC, 3 of 8 patients who underwent SLNB had a positive lymph node, and 4 of the 13 patients had obvious clinical or imaging findings consistent with nodal positivity or metastasis and proceeded directly to regional lymph node dissection. Although the sample size is small the authors suggest that early discovery of lymph node metastasis via SLNB may improve survival rate and recommend SLNB for all patients with PC who do not have obvious clinical or imaging findings suggestive of metastasis at time of presentation.[12]

CONCLUSION

Metastatic PC is associated with high rates of mortality and morbidity, highlighting the aggressive clinical behavior and the need for accurate and timely diagnosis when first biopsied. Given its rarity, non-specific clinical presentation, diagnostically challenging histology, and lack of defined treatment protocols, PC represents a challenging entity for clinicians and diagnosticians alike, thus warranting a collaborative approach to maximize patient outcomes.

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Corresponding Author:
Hayden Christensen
89 Beaumont Ave Given Box 70
Burlington, Vermont 05405
Phone: (435) 535-6301
Email: hayden.christensen@med.uvm.edu

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