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Pruritic, indurated vulvar plaques

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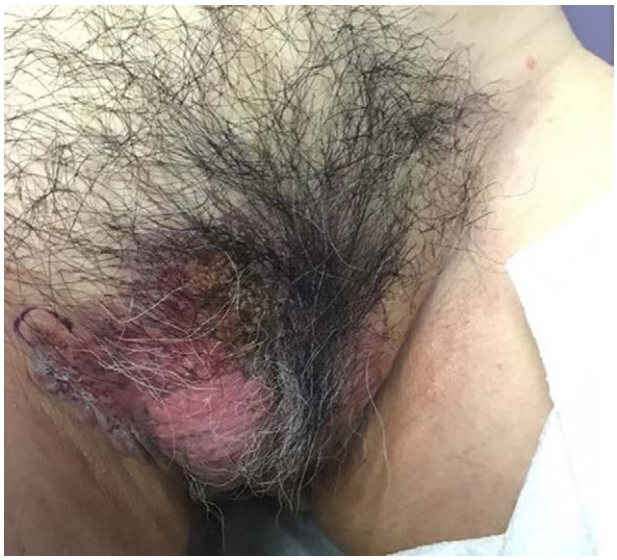

Fig. 1. Right vulvar area with erythematous, scaly, indurated, lichenified plaque.

Question 1

What is the most likely diagnosis?

A. Inverse psoriasis
B. Lichen simplex chronicus
C. Extramammary paget disease
D. Amelanotic melanoma
E. Hailey-Hailey disease

Correct answer: C. Extramammary Paget disease—history and exam as well as punch biopsy results were consistent with extramammary Paget disease (EMPD).

Case

A 77-year-old female presented in an outpatient dermatology clinic with a 13-year history of rash in the genital area. The patient reported pruritus, bleeding, irritation, and redness. The area of concern had been previously treated with topical corticosteroids and topical antibiotics, which improved pruritus but did not improve rash. On examination, an erythematous, scaly, indurated, lichenified plaque of the right suprapubic skin and bilateral labia majora was noted (Fig. 1). Lymphadenopathy was also noted of the right inguinal lymph node chain. There was no tenderness on palpation. The remainder of her exam was normal. Two punch biopsies were performed of two different locations within the affected area. Results demonstrated hyperkeratosis and parakeratosis with epidermal hyperplasia with epidermis showing a proliferation of atypical epithelioid cells showing clear cytoplasm (Fig. 2). Special stains were positive for CK 7 (Fig. 3), CEA, EMA, and HER-2/neu, negative for S100, CK 20, CDX2, and Melan-A leading to the final diagnosis of EMPD.

Discussion

EMPD is a rare intraepithelial adenocarcinoma most often presenting as a slowly growing erythematous, pruritic plaque on the genitals. It most often affects white, female patients 60–80 years in age. Patients with EMPD typically have a good prognosis with a 5-year survival rate ranging from 75% to 95%.[1,2] In the case of our patient, diagnosis was delayed for many years due to cultural beliefs as well as multiple misdiagnoses with the patient then being lost to follow up within that time span. The patient was originally diagnosed in 2007 with "vulvar itching" and "lichen simplex chronicus" and treated off and on with topical steroids and topical antibiotics for months at a time with no resolution. The patient was reluctant to go to gynecology and to allow examination of the

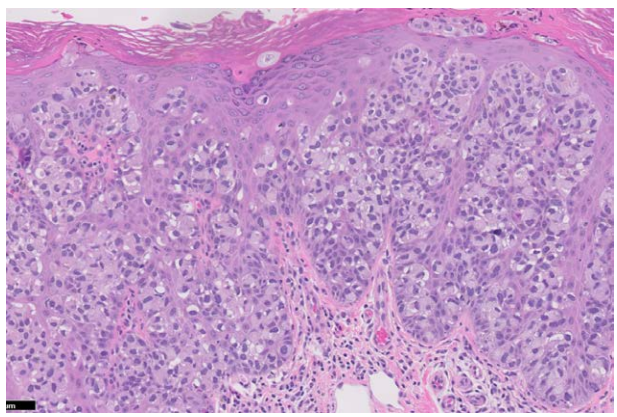

Fig. 2. Skin biopsy showing an intraepidermal proliferation of atypical epithelioid cells with enlarged, irregulsar nuclei and abundant amphophilic cytoplasm. These cells are distributed in aggregates and as individual cells in a pagetoid fashion (hematoxylin-eosin, 200×).

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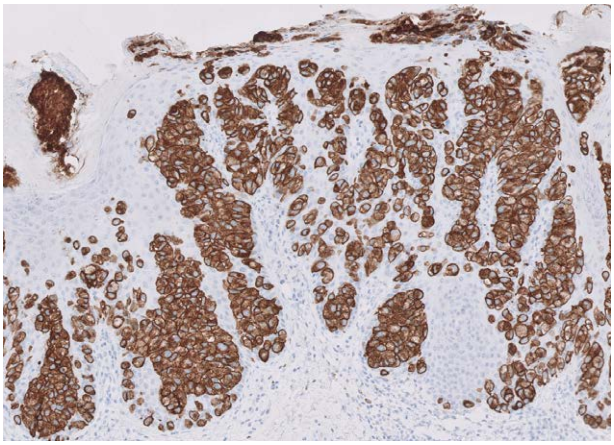

Fig. 3. Skin biopsy showing cytokeratin 7 immunostain (cytokeratin 7, 200×).

vulvar area due to modesty, a cultural barrier seen in Asian women and other minority female groups when compared to white female patients in Western healthcare practices.[3] A biopsy of the suspicious area would have been recommended, if not immediately, within 2 to 4 weeks of the patient presenting with an inadequate response to topical steroid therapy. Standard treatment of EMPD is surgery including Mohs surgery for the appropriate patient. Other treatment alternatives also include photodynamic therapy and topical Imiquimod in the correct clinical setting.[1]

Our patient was managed by gynecology oncology where the patient underwent a partial radical vulvectomy, resection of her right inguinofemoral lymph nodes (IR-guided node biopsy previously performed confirmed node involvement), negative cystoscopy, and vulvar reconstruction by plastic surgery. Patient was staged IIIB, invasive EMPD of the vulva. Cisplastin weekly and radiation were recommended due to evidence of metastatic disease. Patient did not tolerate treatment well and a bone scan months later confirmed metastases to her spine. It is uncommon for EMPD of the vulva to metastasize to the spine, but patient deferred further work up to confirm and was placed in palliative care at that time.

Given that EMPD is very rare, accounting for only 1% of all vulvar cancers, little is known about the ideal treatment for EMPD.[2] Treatment is based on retrospective reviews and case studies, as there are no randomized control trials published for treatment of EMPD.[4] Treatment is typically surgical with adjuvant therapy dependent on stage of disease. Evidence based screening recommendations in the case of newly diagnosed EMPD include age-appropriate screening in addition to urine cytology, mammography, and PSA blood test (males).[5]

Conflicts of interest

None.

Author Contributions

D.A. contributed all path and path-related assessments; A.C. and M.P.A. contributed case and all other text.

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Study approval

N/A.

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