Extramammary Paget’s Disease in the Axilla of a Male Patient: A Case Report and Literature Review

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Abstract

An 82-year-old man was seen for a well-demarcated, erythematous plaque in the left axilla. Topical antifungals were used for months without improvement. After histological examination, the patient was diagnosed with extramammary Paget’s disease. Unilateral, isolated axillary extramammary Paget’s disease in a male patient is a rare finding, and accurate diagnosis requires careful clinical and histological examination.

Introduction

Mammary Paget’s disease was first described by Sir James Paget in 1874. Extramammary Paget’s disease was later described as a distinct entity by Radcliffe Crocker in 1889. The two conditions share similar histology and morphology and are primarily differentiated by anatomical location and likelihood of associated carcinoma.

Extramammary Paget’s disease (EMPD) is a rare, intraepithelial, neoplastic condition that most commonly affects areas with a high density of apocrine glands. Clinically, lesions appear as pruritic patches or plaques that are solitary, well-demarcated and erythematous, sometimes with scaling, crusting, erosion or ulceration.1,2 This nonspecific appearance can mimic more common, benign conditions such as psoriasis, dermatophytosis, irritant dermatitis, and eczema. Misdiagnosing EMPD can lead to a grave clinical outcome, as EMPD can be associated with an underlying carcinoma. Because EMPD can present similarly to other neoplasms on standard H&E, even the astute clinician who performs a biopsy to investigate the lesion may reach the incorrect diagnosis unless he or she is aware of the specific histologic features that can identify the condition.

Extramammary Paget’s disease most commonly affects the vulva, scrotum, and penis; it less commonly affects the face, axilla, buttocks, and thighs.3 The majority of cases of axillary extramammary Paget’s disease present bilaterally and concurrently with anogenital EMPD.4 Here, we present a rare instance of isolated, unilateral axillary extramammary Paget’s disease in a male patient, along with a review of literature and a discussion highlighting the importance of correctly diagnosing this entity.

Case Report

An 82-year-old man was seen for a well-demarcated, erythematous plaque in the left axilla. Topical antifungals were used for months without improvement. After histological examination, the patient was diagnosed with extramammary Paget’s disease. Unilateral, isolated axillary extramammary Paget’s disease in a male patient is a rare finding, and accurate diagnosis requires careful clinical and histological examination.

Clinical examination revealed a well-demarcated, erythematous plaque in the left axilla with no epidermal change (Figure 1). A 4-mm punch biopsy was taken at the posterior aspect of the lesion, which showed pagetoid spread with features concerning for Bowen’s disease (Figure 2). Given the overlapping features of Bowen’s disease and Paget’s disease, confirmatory staining for CK-7, CEA, and CAM5.2 was performed, and positivity of these stains confirmed the diagnosis of extramammary Paget’s disease while excluding other pathologies on the differential diagnosis (Figure 3).3,5

The patient was referred to Plastic Surgery. The lesion was excised, and the wound was closed with a rotational flap. Sentinel lymph nodes were negative for metastases.

Discussion

Extramammary Paget’s disease (EMPD) is an uncommon skin condition, with only a few hundred cases reported in world literature.7 EMPD is between 1.3 times and 4 times more prevalent in females, and the average age at diagnosis is 75.5,7 Among locations of extramammary Paget’s disease, the axilla is a rare site, with anogenital regions being much more common. Specifically, only 1% to 2% of cases of EMPD are found outside the labia majora, perianal skin, and male genitalia.1 A recent review of literature showed that among the 23 previously reported cases of axillary EMPD, mostly in the Japanese population, only 10 had isolated axillary involvement.4 Of note, the remaining 13 cases of axillary EMPD were found to be bilateral and concurrent with anogenital EMPD, the so-called “triple Paget’s disease,” which is more commonly reported in the literature.8 A study published in 2011 found only seven axillary EMPD cases published in the previous 20 years.3 Our case represents the exceedingly rare circumstance of an American male with isolated, unilateral axillary extramammary Paget’s disease.

When approaching a patient with a rare entity such as axillary EMPD, it is important to maintain a broad differential. The most common complaint associated with anogenital EMPD is long-standing, localized pruritus.1 In contrast, EMPD...
of the axilla presents with a non-specific clinical appearance similar to psoriasis, Bowen's disease, dermatophytosis, irritant dermatitis, or eczema. Patients may experience burning, itching, or no clinical symptoms. Thus, patients with axillary EMPD are commonly treated with a prolonged course of topical antifungals or corticosteroids and appear to have recalcitrant disease. In this clinical situation, biopsy is recommended. Diagnosis of EMPD requires immunohistochemical analysis of a punch biopsy of the lesion. Standard hematoxylin and eosin (H&E) staining shows Pagetoid cells, which are clusters of large, vacuolated cells with blue-tinted, finely granular cytoplasm located in the epidermis and adnexal epithelia. Pagetoid cells demonstrate a characteristic pagetoid spread in which abnormal cells are distributed singly and in small groups throughout an epithelial layer from deep to superficial. Other neoplastic conditions that demonstrate pagetoid spreads include Bowen's disease, superficial spreading malignant melanoma, mycosis fungoides, and Langerhans cell histiocytosis. Thus, a non-spreading malignant melanoma, mycosis fungoides, spreads include Bowen's disease, superficial neoplastic conditions that demonstrate pagetoid spread; however, the atypical cells of Bowen's disease are of squamous origin, while the atypical cells of superficial spreading malignant melanoma, Bowen's disease, mycosis fungoides, and Langerhans cell histiocytosis. Confirmatory immunohistochemical staining for EMPD includes positivity for CK-7, CEA, CAM5.2, EMA, and GCDFP-15 with epidermal spread of pagetoid cells of apocrine origin. Our patient's lesion stained positive for CK-7, CEA, EMA, and CAM5.2, ruling out other differentials. Assessing for invasion and metastatic disease remains a mainstay of treatment, and sentinel lymph node biopsy to assess for metastasis should be performed in individuals who demonstrate > 1 mm of dermal invasion by Paget cells. Our patient's sentinel lymph nodes were negative, and after surgical excision there have been no signs of ongoing disease process. Most patients do not have metastatic disease, and for these patients, wide surgical margin excision is almost always curative.

Conclusion

Isolated, unilateral extramammary Paget's disease is a rare neoplastic entity. It often presents non-specifically or as a pruritic lesion recalcitrant to corticosteroids and/or topical antifungals, as in our patient. Biopsy results with routine histology may be equivocal, as similar histopathological findings of atypical pagetoid cells may also be found in superficial spreading malignant melanoma, Bowen's disease, mycosis fungoides, and Langerhans cells histiocytosis. The nuclei of cells in EMPD which are clusters of large, vacuolated cells with blue-tinted, finely granular cytoplasm located in the epidermis and adnexal epithelium. EMPD demonstrates positive sentinel lymph nodes; however, 15% of cases of invasive EMPD without lymphadenopathy have shown positive sentinel lymph nodes, as well. Therefore, clinicians should assume metastatic potential for this entity regardless of lymphadenopathy.

References