Case Report

Extramammary Paget's disease of vulva: A rare entity

Sandhya Yadav, Pratik Gahalaut, Hardev Singh Soodan, Nitin Mishra, Madhur Kant Rastogi
Department of Dermatology, Shri Ram Murti Smarak Institute of Medical Sciences, Bhojipura, Bareilly, Uttar Pradesh, India

Address for correspondence:
Dr. Sandhya Yadav, House No. 390, Ground Floor, Sector-38, Gurgaon - 122 001, Haryana, India. E-mail: sndhydv1988@gmail.com

Abstract

Extramammary Paget's disease is a rare cutaneous, intraepithelial adenocarcinoma involving primarily the epidermis but occasionally extending into the underlying dermis. The condition typically presents as a red, velvety, pruritic skin rash of the vulva region which closely mimics a multitude of other, more common conditions. As a result, vulvar Paget's disease is frequently misdiagnosed, leading to an often lengthy lag time (an average of about 2–3 years) between the onset of symptoms and diagnosis.

Key words: Genital malignancy, intraepithelial carcinoma, non-healing ulcer

INTRODUCTION

Extramammary Paget's disease (EMPD) is a rare cutaneous, intraepithelial adenocarcinoma involving primarily the epidermis but occasionally extending into the underlying dermis[1] which accounts for <1% of carcinomas in vulva and majority of the patients are postmenopausal females.[2] It presents with a long-standing history of pruritic, erythematous, scaly, or velvety patches. The most frequent site is the vulva, but perineal, scrotal, perianal, and penile skin are also common areas.[3] The cancer cells in the neoplasm usually stay "*in situ*" and only rarely invade into the dermis to be metastatic through the lymphatic system.[2] Palpable lymph nodes are less frequently present in EMPD.[3]

CASE REPORT

A 55-year-old postmenopausal female presented with history of itchy and gradually progressive reddish lesion on genitals for 3 years. The patient was applying topical antifungals and steroids with partial relief. Cutaneous examination revealed a well-defined, moist erythematous plaque of size 15 cm × 10 cm with multiple erosions involving bilateral labia majora and clitoris [Figure 1]. On full body examination, no other lesion or lymphadenopathy was observed. On investigations, complete hemogram, chest X-ray and ultrasound whole abdomen were normal.

The clinical diagnosis of EMPD was confirmed by a 3 mm punch biopsy which showed cells within the basal layer of epidermis having nuclear enlargement with atypia, prominent nucleoli, and well-defined ample cytoplasm. These cells also extended downward to surround hair follicles. In addition, a focus of squamous cell compression and moderate inflammatory cell infiltrate was seen in the upper dermis [Figure 2]. Our patient was advised to undergo excisional surgery, but she refused and was lost to follow-up.

DISCUSSION

In 1874, Sir James Paget first described mammary Paget's disease[4] and in 1901, Dubreiuhl reported the first case of vulvar EMPD.[5] Vulvar Paget's is a rare disease and typically presents as a red, velvety, pruritic skin rash of the vulva region which closely mimics a multitude of other, more common conditions.[6] As a result, vulvar Paget's requires a biopsy for definitive diagnosis and is frequently misdiagnosed.

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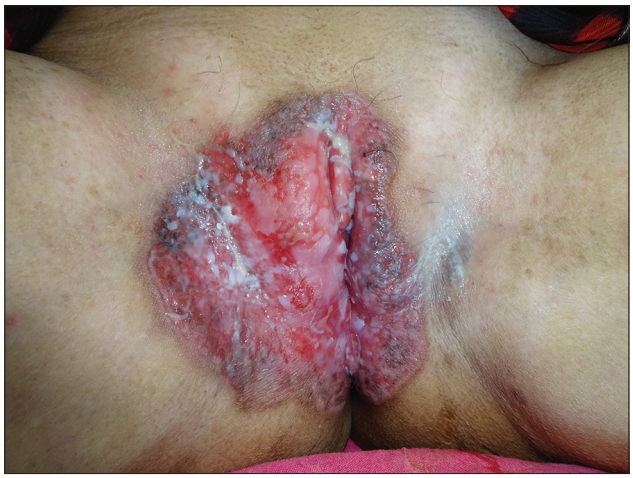

Figure 1: A well-defined erythematous plaque

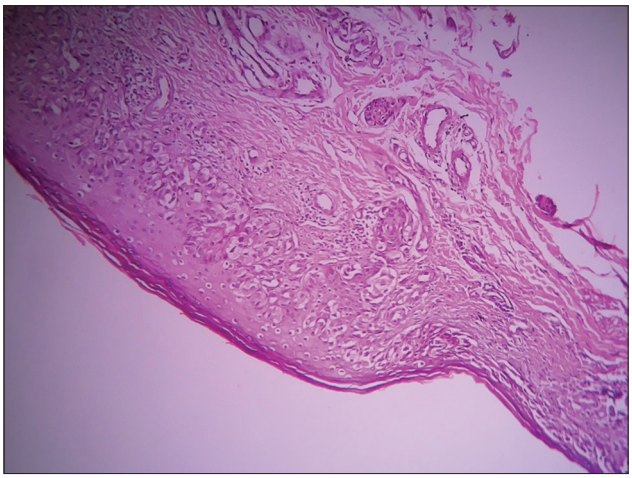

Figure 2: Numerous Paget's cells within the epidermis (H and E, ×40)

Paget's disease can be found in both men and women; however, it is most commonly seen in postmenopausal Caucasian females.[7] Typically, involved sites are the vulvar, perianal, scrotal and penile regions; rare sites include the thighs, buttocks, axilla, eyelids and external ear canal.[3] Grossly, the lesions appear as well-defined, moist and reddish plaque. The lesions can be white to red, scaling or macerated, can appear infiltrated, eroded or look like an ulcerated plaque.[2] Due to disease's relatively low incidence and since many cases are unreported, its true incidence remains unknown.[2]

The clinical differential diagnosis for EMPD includes psoriasis, contact dermatitis, fungal infections, lichen sclerosus, intraepithelial neoplasia and melanoma. The nonspecific clinical findings often lead to misdiagnosis and an average of 1 year can pass before a biopsy is taken and definitive diagnosis is made.[8]

It is characterized microscopically by the presence of specific tumor cells called Paget's cells. Paget cells are large cells with pale clear cytoplasm, large round hyperchromatic nuclei which tend to form clusters or solid nests. Paget cells can be found at all the levels of the epidermis. They can migrate within the epidermis in a horizontal as well as vertical manner.[5] Paget's cells have intracellular mucopolysaccharides, with EMPD having a greater amount of mucin as compared to MPD. As a result, cells frequently show positive staining for periodic acid-Schiff and diastase resistance, mucicarmine, alcian blue at pH 2.5, and colloidal iron.[3]

Surgery remains the treatment of choice for EMPD with recurrence rate of up to 44% with wide local excision. Other modalities include Mohs micrographic surgical excision, radiotherapy, topical agents including 5-fluorouracil, bleomycin, imiquimod and photodynamic therapy.[3]

This case is being reported for its rarity and to stress on the fact that an early biopsy should be advised to avoid misdiagnosing EMPD.

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Conflicts of interest

There are no conflicts of interest.

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