


Extramammary Paget's disease: Persistent pruritus vulvae is an alarming symptom in primary care

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Introduction

Extramammary Paget's disease (EMPD) is a rare, malignant cutaneous intraepithelial adenocarcinoma which occurs in sites rich in apocrine glands (*i.e.* vulva, perianal region, scrotum, penis and axilla). EMPD most often occurs in elderly women. The commonest area involved is the vulva (1). Nevertheless, EMPD is responsible for less than 01% of all vulval neoplasms. Etiology is not clearly known. The estimated incidence in Europe is about 0.7 cases per 100,000 women (2).

Clinical manifestations often mimic benign inflammatory dermatological conditions. This delays the early diagnosis. The definitive diagnosis is obtained by a skin biopsy of the vulva, which shows Paget cells. The treatment of choice is wide excision. Local recurrence is common even with a wide excision (1).

Case presentation

A 73-year-old postmenopausal married woman presented for the first time with a slow-growing intensely pruritic, plaque on vulva. It initially started as an extreme pruritus of vulva with mild erythema, which progressed gradually to present size and appearance over the period of 02 years. She had been on various treatments for several benign dermatoses.

Despite therapy, pruritus worsened with centrifugal extension of the lesion. She neither had fever nor other systemic symptoms. Patient had no pertinent medical history including diabetes mellitus or sexually transmitted diseases.

Dermatological examination revealed well-demarcated, scaly, lichenified vulvar plaque. Discrete and confluent grayish crusts and shallow erosions were also seen (Figure 01). There was no similar lesion anywhere else on the body. There was no regional lymphadenopathy. General and systemic examination revealed no abnormality.

Urgent incisional skin biopsy was performed for histopathological evaluation. Hematoxylin and Eosin (H&E) staining of the biopsy from the lesion showed large round cells with ample pale-staining cytoplasm, pleomorphic nuclei, and occasional prominent nucleoli, indicative of Paget cells (Figure 02). Inflammatory infiltrate with invasion of Paget's cells into the dermis at places was also seen (Figure 03). The clinical diagnosis of EMPD was confirmed histopathologically. Basic laboratory investigations were within the normal limits. Ultrasonography of abdomen and pelvis was normal.

She underwent vulvectomy and reconstruction surgery. Regular dermatological assessments were done in order to detect early signs of local recurrence (Figure 04 & 05).



Figure 01: Clinical presentation of the vulvar lesion



Figure 04: Post surgical appearance of vulva after one month

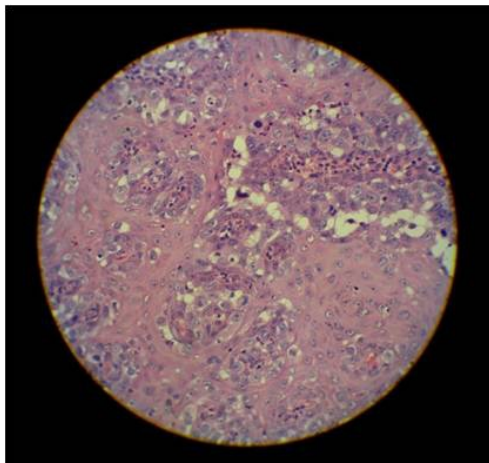


Figure 02: Paget cells (H&E Stain 40x magnification)



Figure 03: Inflammatory Infiltrate with invasion of Paget cells into the dermis (H & E stain 10 x magnification)



Figure 05: Post surgical appearance of vulva after one year

Discussion

The most common symptom of EMPD is intense pruritus. Majority of patients have no other symptoms. Remainder will experience non-specific clinical manifestations, which mimic benign dermatoses such as frictional-sweat dermatitis, contact dermatitis, flexural dermatitis, candida intertrigo, tinea cruris, lichen planus and lichen simplex chronicus (1).

The hallmark of this condition is relentless progression despite routine conventional therapy. As a result, the diagnosis is delayed with a median of 2 years from the onset of symptoms. Early diagnosis improves survival in EMPD. 10% of primary EMPD may have nodal involvement, which has the worst five-year survival rate of 0% to 24%. Yet, lesions which are detected early have a good prognosis with a five-year survival rate of 80% to 95%. Poor prognostic factors are deeper invasion, thickness of the tumour, vascular invasion and lymph node invasion (3).

Topical therapy has a variable response. 5% imiquimod cream, three times per week (on alternative days) for 16 weeks is widely used. 5% 5-fluorouracil cream is another topical agent, which can be used in EMPD. Twice a day application of 5% 5-fluorouracil cream for 04 weeks is the standard regimen. Radiotherapy and CO₂ laser ablation are some of the other non-surgical treatment modalities.

Treatment of choice for EMPD is wide local excision surgery or Mohs micrographic surgery. Recurrence rates are high which can range from 31% to 61% with local excision surgery. Even with Mohs micrographic surgery, recurrence rates vary from 08% to 26% (3). EMPD usually presents as a multifocal growth with subclinical extension into normal appearing skin. This highly irregular pattern of spread can have finger-like projections beyond the main body of the tumour. Interestingly, these subclinical projections may not be detected with routine H&E staining. Cytokeratin 7 immunostaining is required to visualize Paget cells that extend beyond the main body of the tumour (4). Thus, regardless of treatment method, long-term follow up is recommended to monitor for the local recurrence, development of internal malignancy or distant metastases (5).

Conclusions

High level of suspicion is always essential for an accurate clinical diagnosis of EMPD. Thus, Persistent pruritus vulvae must be considered as an alarming symptom in primary care. This case report reinforces the need for an early skin biopsy for histopathological evaluation from vulvar lesion which does not respond to conventional therapy.

Informed written consent has been obtained by the patient to publish this case report with photographs. Authors declare no conflicts of interests.

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