

# Paget's disease of the vulva, perineum, anal and gluteal region with underlying invasive and metastatic vulvar carcinoma.

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## INTRODUCTION

Extramammary Paget's disease is a rare neoplasm. Common sites of occurrence include vulva, perianal region, perineum and gluteal region. The International Society for the Study of Vulvar Disease classified vulvar Paget's disease as a non squamous intraepithelial lesion of the vulva. Unlike Paget's disease of the nipple, extramammary Paget's disease is not always associated with a subjacent or visceral malignancy.

## CASE REPORT

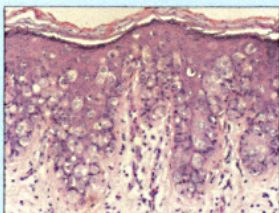
We report a case of extramammary Paget's disease, which involves vulva, perineum, perianal and gluteal region.

A 51-year-old woman was referred to department for vulvovaginal disease with 2 years history of a pruritic irritation and rash of vulva, perineum, perianal and gluteal region. Patient was treated by gynecologist and then by dermatologist without any effect by antibiotic and corticoids without biopsy.

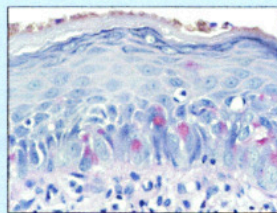
On clinical assessment, there was a large and extensive redish erythematous and eczematoid area with multiple superficial erosions, affecting the whole vulva, perineum, anal and gluteal region extending an area 25 cm x 18 cm. Inguinofemoral nodes were enlarged to 2 cm in left groin.



Clinical features of extramammary Paget's disease before resection.



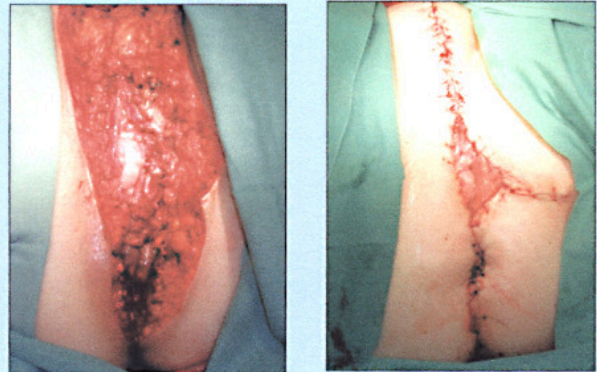
The epithelium is hyperkeratotic and diffusely infiltrated by large number of Paget's cells. All levels of the epidermis are involved.



The Paget's cells shows positive staining with diastase-periodic acid-schiff (PAS).

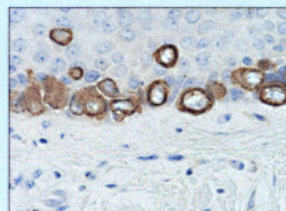
Biopsy of the lesion showed clusters of Paget's cells-PAS Alcian blue, CK, EMA, CEA positive and S 100P, HMB-45, vimentin negative. Proctosigmoideoscopy, cystoscopy and CT of abdomen showed no abnormality, full blood count, biochemical profile and chest X-ray were normal too.

Due to extent of disease and lymphadenopathy in left groin, radical vulvectomy with inguinofemoral lymphadenectomy were done.

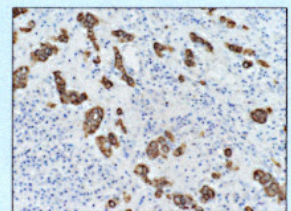


Radical Vulvectomy

Histopathological finding confirm Paget's disease of vulva, perineum, anal and gluteal region, but also area of invasive adenocarcinoma in left labium maius and metastatic adenocarcinoma in 4 inguinofemoral nodes in left groin. Positive margins of Paget's disease were describes in vulva, and gluteal region.



The Paget's cells show strong pancytokeratine expression.



Proctosigmoideal underlying invasive adenocarcinoma.

Due to positive margins and metastatic carcinoma in groin adjuvant radiotherapy to groins, vulva and pelvis followed. No complications either surgical treatment or radiotherapy were described.



Clinical features after surgical treatment and radiotherapy

Paget's disease is an important problem, often with long duration of symptoms, that calls for early diagnosis and adequate treatment. Cause of uncertain behaviour and tendency to locoregional relaps an effective long-term follow up is mandatory.