

Case Report

Extramammary Paget's disease: a man presented with bilateral groin rash

乳腺外佩吉特病：男患者雙側腹股溝部皮損

MC Wong 王夢貞 and KC Lee 李景中

A 67-year-old man presented with bilateral groin eruption for a few years. The diagnosis of extramammary Paget's disease was made based on clinicopathological findings. The patient defaulted follow up and hence any underlying associated malignancy could not be excluded.

患者，男，67歲，出現雙側腹股溝部皮損數年。臨床為病理學診斷為佩吉特病。因患者無接受隨診，可能關聯的潛藏惡性腫瘤未能排除。

Keywords: Extramammary Paget's disease

關鍵詞： 乳腺外佩吉特病

Introduction

Extramammary Paget's disease (EMPD) is an uncommon intraepithelial adenocarcinoma that affects apocrine gland-bearing skin. It may be associated with underlying internal malignancy

and adnexal adenocarcinoma. We reported below a man with bilateral groin EMPD.

Case report

A 67-year-old Chinese gentleman presented with itchy rash over bilateral groin areas and scrotum for a few years. He denied any urinary or gastrointestinal symptoms. For his past health, he has ischaemic heart disease and congestive heart failure. He is a non-smoker and non-drinker.

On physical examination, there were well-defined erythematous plaques over both groin areas with involvement of scrotum. The lesions were moist and there was whitish surface scaling

Social Hygiene Service, Department of Health, Hong Kong

MC Wong, MBBS, MRCP(UK)

Department of Pathology, Queen Elizabeth Hospital, Hong Kong

KC Lee, FRCPA, FHKAM(Pathology)

Correspondence to: Dr. MC Wong

Yau Ma Tei Dermatology Clinic, 12/F, Yau Ma Tei Specialist Clinic, 143 Battery Street, Kowloon, Hong Kong

mixed with erosions (Figure 1). The clinical differential diagnoses include intertrigo, tinea cruris, extramammary Paget's disease, Bowen's disease and squamous cell carcinoma.

Investigations including skin scrapping and fungal culture were negative. Swab culture from the lesion grew *Staphylococcus aureus* and *Pseudomonas aeruginosa*. An incisional skin biopsy was performed which showed the epidermis was extensively involved by isolated and clusters of pale cells forming pagetoid growth pattern (Figure 2). The pale cells exhibited oval nuclei with prominent nucleoli and pale cytoplasm (Figure 3). Immunohistochemical studies showed that these cells expressed epithelial marker (cam 5.2) but not S-100. The histopathologic features were compatible with extramammary Paget's disease. The patient was treated with a course of cloxacillin. He was not keen for surgery and defaulted follow up.

Discussion

EMPD is a rare intraepithelial adenocarcinoma that mimics inflammatory or infectious condition. It is a heterogenous condition representing either an intraepithelial adenocarcinoma in situ with



Figure 1. Left groin well-defined erythematous moist plaque with surface scaling and erosions.

invasive potential or pagetoid spread of an underlying adnexal tumour or of a regional internal malignancy. EMPD usually developed in the seventh decade.¹ It occurs most frequently in Caucasians with female predominant. However, most cases reported in Japan are men. There is predilection for apocrine gland-bearing areas

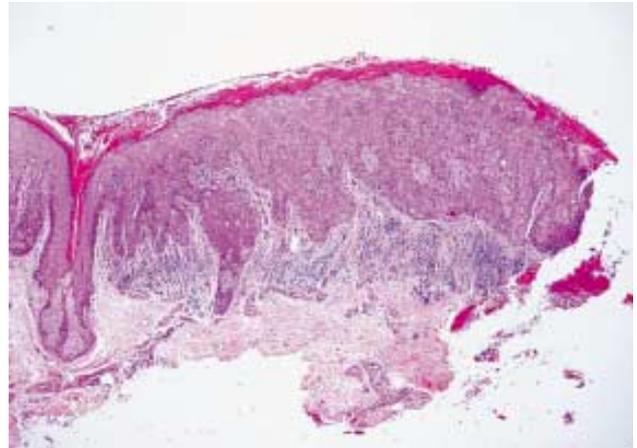


Figure 2. Low power showing isolated and small clusters of pale tumour cells scattered throughout the epidermis forming a classic Pagetoid growth pattern. (H & E, original magnification x 10)

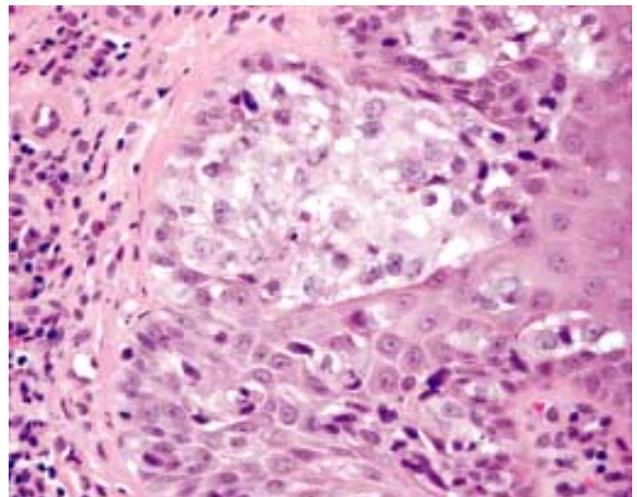


Figure 3. High power showing the pleomorphic pale tumour cells forming nests along the dermal epidermal junction. There is even cytoplasmic melanin pigment mimicking malignant melanoma. (H&E, original magnification x 20)

including vulva, perianal region, perineum, male genitalia. Rare sites include thigh, buttocks, axilla and external auditory canal.² Lesions developed in skin devoid of apocrine glands has been reported.³ Most commonly lesions present as well-defined, moist erythematous or white scaly plaques. Scattered white scales and erosions can give rise to 'strawberry and cream' appearance. Patients often complain of pruritus but may be asymptomatic. Bleeding, oozing, tenderness or a burning sensation can occur and symptoms may be present for months to many years.

Histologically, EMPD is characterised by distinct vacuolated Paget's cells in epidermis. Immunohistochemical staining for glandular cytokeratins, epithelial membrane antigens and carcino-embryonic antigens (CEA) should be performed. Diagnosis of extramammary Paget's disease involves two important steps. Firstly to ascertain the correct diagnosis and secondly try to determine if it is the primary cutaneous form or one associated with underlying tumour. Important differential diagnoses in this case are malignant melanoma and Pagetoid Bowen's disease. A combination of Cam 5.2 (a low molecular weight cytokeratin) and S-100 immunostaining is sometimes essential for a definitive diagnosis unless there is unequivocal mucin production or tubular structure formation. While it is generally agreed that a CK7 positive, CK20 negative and GCFDP-15 (BRST-2) positive profile favour a primary cutaneous form, other combination of results are considered non-specific. Moreover, definition on what to be considered positive (100% positive or a cut-off percentage) varies or not clearly defined in different studies making application on individual case difficult. As a result, while immunostaining can sometimes provide a suggestion for a possible primary cutaneous extramammary Paget's disease, it probably cannot replace a search for underlying malignancy especially for a perianal lesion in which the rate of having an associated internal malignancy is higher.

In a study of 197 cases of EMPD, 12% of patients have associated concurrent underlying internal malignancy and 24% have associated adnexal adenocarcinoma with higher mortality.⁴ Location of underlying internal malignancy appears to be closely related to location of EMPD. Perianal EMPD tends to associate with adenocarcinoma of digestive system whereas penile-scrotal-groin EMPD is related to genitourinary malignancy.⁵

Management of EMPD requires a thorough search for possible associated malignancy which affects the subsequent management and prognosis. For treatment of primary EMPD, first line therapy is wide local excision.⁴ Other modalities that have been employed include Mohs micrographic surgery (MMS), radiation, combined chemo-radiotherapy, photodynamic therapy, topical 5-fluorouracil, CO₂ laser and topical bleomycin.^{4,6-11} Recurrence or residual diseases are frequently seen because EMPD often have irregular margins, being multicentric and extend beyond visible clinical margins. Recurrent rates for vulvar EMPD range from 15% with vulvectomy, 27% with MMS and 43% with wide local excision; perianal diseases recur in 28% of patients after MMS and in 50% of patients after wide local excision.⁴

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