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Case Report

Extramammary Paget's disease of penis successfully treated by radiotherapy

放射治療成功治癒陰莖部位之乳房外佩吉特氏病

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A case of a 71-year-old Chinese man diagnosed to have Extramammary Paget's Disease (EMPD) was reported. He presented with a right-sided, recalcitrant groin ulcer. The diagnosis was made two years after first presentation. He was treated by radiotherapy alone and has no relapse since then. Till now, no internal malignancy was found in this patient.

本報告為一例乳房外佩吉特氏病，患者為 71 歲男性華人，其首發症狀為右側腹股溝頑固性潰爛，於發瘤病後兩年確診，單用放射治療，並無復發，無發現其他內臟併發惡性腫瘤。

Keywords: Chinese, Extramammary Paget's Disease, radiotherapy

關鍵詞：華人，乳房外佩吉特氏病，放射治療

Introduction

Extramammary Paget's Disease (EMPD) is a rare paraneoplastic cutaneous eruption that occurs mostly in the elderly. A 71-year-old Chinese male presented with a persistent penile root ulcer which confirmed to be EMPD. He was successfully treated with radical radiotherapy. Our experience is presented.

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Case report

A 71-year-old Chinese elderly male patient in February, 2002 presented with a two years history of chronic, non-healing right-sided groin ulceration. The ulcer was non-pruritic and non-painful. There was no concomitant skin eruption or ulceration elsewhere. Apart from the ulcer, the patient was well and enjoyed good past health. He denied any drug allergy. No history of trauma was reported. He denied any history of venereal exposure. On physical examination, there was a right-sided superficial ulcer measured 2 cm X 1.5 cm over the root of the penis. The ulcer was based on a larger erythematous irregular plaque with an advancing irregular margin (Figure 1) . The ulcer was increasing in size. There was no

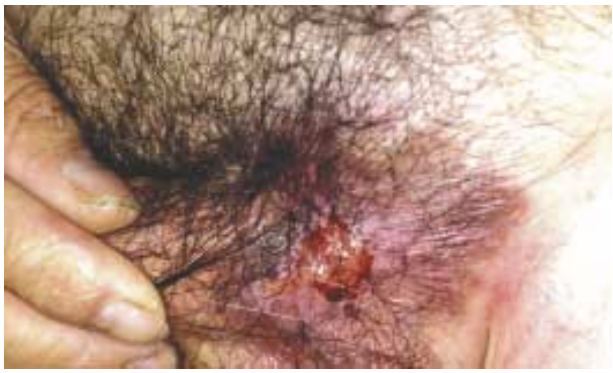

Figure 1. A right-sided chronic groin ulcer on an erythematous plaque with irregular border.

inguinal lymphadenopathy. An incisional skin biopsy was performed which showed multiple foci of Paget cells aggregated in the epidermis. The skin appendages of the upper dermis were also colonised by Paget cells. The tumour cells possess pleomorphic nuclei, prominent central nucleoli and moderate amount of vacuolar cytoplasm. A severe chronic inflammation was also noted. The histological features were compatible with EMPD (Figure 2). Screening test for sexually transmitted diseases revealed no abnormality. Screening for internal malignancy was performed by surgeons and this included colonoscopy, cystoscopy, ultrasound of the testes and abdomen, biopsy of the prostate gland. No internal malignancy was revealed. Test for serum chorionic-embryonic antigen (CEA) was also negative. Treatment options were discussed with our patient and radiotherapy was preferred than radical surgical excision.

A total of 15 sessions of radiotherapy to the right perineum was given at five treatments per week for consecutive three weeks. The lesion gradually subsided leaving residual hypo- and hyper-pigmentation of the skin (Figure 3). Subsequent follow-ups till the time of writing the report revealed no evidence of recurrence of internal malignancy.

Discussion

EMPD is defined as an intraepithelial adeno-carcinoma, usually of epidermal origin and

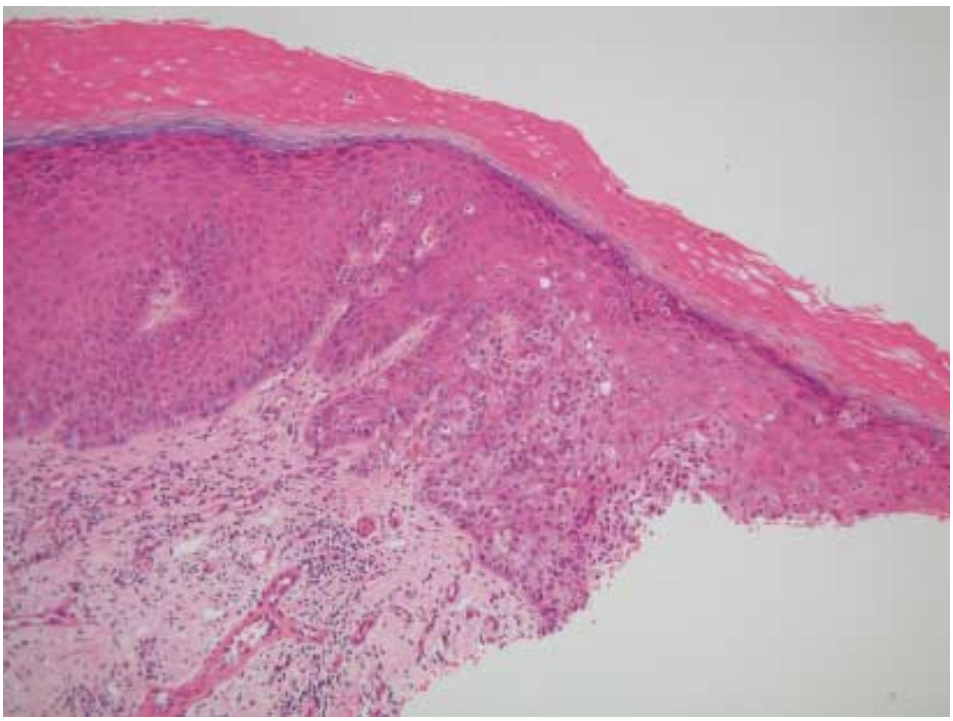

Figure 2. Histology showed multiple foci of Paget cells aggregated in the epidermis.

glandular differentiation. It may be associated with an underlying adnexal adenocarcinoma or internal malignancy.[1] It is an uncommon para-neoplastic skin condition; more often occurs in the female with a female to male ratio of 3.2 to 1. Most commonly occur in the elderly with an age range from 47 to 87 (Mean = 64) years old. The patient usually presents with non-descript symptoms like pruritus, tenderness, oozing from an ulcer and presence of a lump. EMPD most commonly occur in anatomical regions where apocrine glands are abundant like vulva, perianal area, genitourinary region of the male, buttock and very rarely; external auditory canal. Diagnostic delay is common. The duration of symptoms may range from one month to 30 years. Clinically important differential diagnoses included Bowen's disease, eczema, psoriasis and blistering dermatosis like pemphigus vulgaris.

The pathogenesis of EMPD is still speculative. The cell of origin of EMPD is unknown. It may arise de-novo within an epidermis or extended from an in-situ appendageal glandular tissue adeno-carcinoma.[2] The association of EMPD with an internal malignancy of apocrine origin raises the possibility that similarly derived embryogenic tissue susceptible to the same carcinogenic stimuli may undergo malignant degeneration simultaneously.

EMPD was associated with underlying internal malignancy. The figure reported in the literature ranged from 0-50%.[1] Twenty-four percent of cases of EMPD demonstrated association with a cutaneous adnexal carcinoma with a mortality rate of metastatic disease up to 46%.[1] Twelve percent of EMPD may present with a concurrent malignancy; 17% with a nonconcurrent malignancy; 29% with concurrent plus nonconcurrent malignancy and 71% with no malignancy.[1] The most frequent anatomical site of the internal malignancy associated with EMPD in the female in decreasing order is vulva, cervix, breast, Bartholin's gland and gallbladder. In the male; it is the genitourinary tract like the penis, scrotum, groin; perianal areas; prostate; kidneys

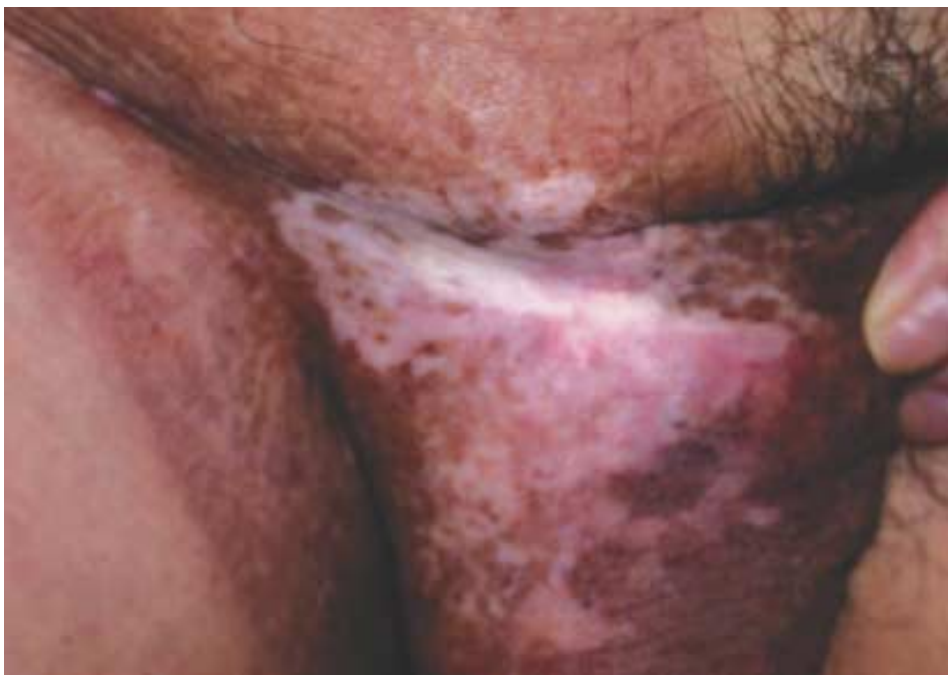

Figure 3. Complete remission of the right groin lesion after radiotherapy leaving hypo- and hyper- pigmentation.

and rectum. In all circumstances, a diligent search of an associated internal malignancy in a patient presented with EMPD is mandatory. So far, no internal malignancy was noted in our patient.

Despite the recommended first-line treatment of EMPD is complete surgical excision; our case was successfully treated with radiotherapy in the first instance without observed relapse. Surgical excision in EMPD may have two problems. Longstanding EMPD lesion may be extensive and diffuse, wide excision may not be feasible and may leave residual disease. A study involving 30 subjects showed that there was a relapse rate of EMPD as high as 40% in five years treated by surgical excision alone.[3] Most elderly patients, as in our case, suffered from EMPD may be reluctant to have surgery. Radiotherapy as compared with surgery required less post-treatment care and hospitalisation. As a result, wound sepsis and the cost of the treatment are less. There is increasing evidence that radiotherapy is a more suitable initial therapy of EMPD in the elderly. Three patients with primary EMPD treated with radiotherapy, none had evidence of recurrence 12-60 months later.[3] Combined treatment of EMPD by surgery and radiotherapy found no relapse in another 3 cases.[3] Stockdale found no signs of recurrence in four out of six patients treated by radiotherapy during 13-51 months of follow up.[4] Burrows also reported successful treatment of three cases of EMPD by radiotherapy without any relapse.[2] Our reported case provides further evidence that radiotherapy may be a first line treatment option for elderly patient with confirmed EMPD.

In conclusion, we have reported a case of EMPD in an elderly man treated by radiotherapy. Continuous careful monitoring of the patient revealed no evidence of internal malignancy or relapse. We suggested that radiotherapy should be offered as a first line treatment to the elderly especially those who are disabled by multi-system diseases.

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