Red Plaque on Groin

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A 60-year-old male presents with a persistent eruption on the left groin of one year's duration, despite steroid and antifungal medication.

Examination reveals a localized red plaque measuring 3 cm in diameter. There is no regional lymphadenopathy nor organomegaly. You decide to proceed with skin biopsy.

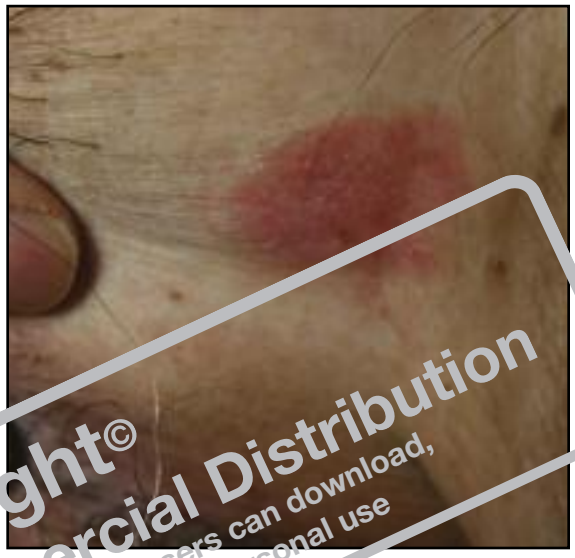

Figure 1: Red eruption on left groin

Clinical Investigation

CT scan of the abdomen and pelvis was unremarkable. Local wide excision was performed with rotational flap repair. Pathology revealed clear margins.

What is your diagnosis?

a. Extramammary Paget's disease
b. Fixed drug eruption
c. Tinea cruris
d. Scrotal cyst

Answer: Extramammary Paget's disease

About Extramammary Paget's disease

Extrammary Paget's disease (EMPD) is a primary cutaneous adenocarcinoma which presents with a similar clinical and identical pathological finding as Paget's disease of the breast.[1] However EMPD, as the name implies, occurs outside the mammary gland and has a predilection for the apocrine-bearing areas of the genital region. It was first recognized by Crocker in 1889 as a distinct and separate entity, following Paget's description of the breast in 1874. EMPD is a relatively rare tumour; less than 200 cases have been reported in the literature.[2]

Most patients develop EMPD in the fifth and sixth decades of their life. EMPD typically presents as a persistent, pruritic, eczematous eruption in the groin. Often, there is a preceding diagnosis of intertrigo or tinea cruris. However, it is nonresponsive to topical medication. EMPD afflicts more women than men (> 3:1 ratio), and the most common locations include the vulvar region and perineum.

The neoplastic tumour (Paget) cells have an epidermal origin and may display glandular differentiation. Approximately 25% of cases will show invasive adnexal carcinoma of underlying tissue. In addition, 10% of patients will have a concurrent internal malignancy, often related to and/or in close proximity to the site of the primary cutaneous neoplasm. Therefore, common associated malignancies include adenocarcinoma of the gastrointestinal as well as genitourinary tract. A thorough investigation for internal involvement must be undertaken.

Patients should have a complete history and physical exam with emphasis on the rectum, integument and lymph node system. Imaging exams include sigmoidoscopy and cystoscopy. Female patients require a breast exam, PAP smear and colposcopy as well as a pelvic ultrasound. A multidisciplinary approach may be required, involving coordinated care from a gastroenterologist, urologist, and colorectal, gynecologic and plastic surgeons.

Local wide excision, possibly combined with Moh's microsurgery, is the treatment of choice for cutaneous disease. Close follow up is required since there is a high incidence of recurrence, ranging from 10 to 60% following the first two years of diagnosis. It is difficult to establish a survival rate due to the rarity of the disease. The prognosis depends on early diagnosis and the presence of internal malignancy. Patients with localized cutaneous involvement have an excellent prognosis. The presence of invasive carcinoma and lymph node metastases are indicators of poor prognosis. Ultimately, 50% of patients with internal adenocarcinoma will succumb to their illness.

Take-home Message

Any chronic dermatitis that is nonresponsive to topical medication should be biopsied, to rule out malignancy. Dx

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References
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2. Chanda JJ. Extramammary Paget's Disease: Prognosis and Relationship to Internal Malignancy. J Am Acad Dermatol 1985 Dec:13(6):1009-14.

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