

## C A S E R E P O R T

# A case of extramammary inguinal Paget disease in a male patient: surgical treatment with an abdominal advancement cutaneous flap

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**Summary.** Extramammary Paget disease (EMPD) is a rare neoplasm. The clinical case of a 55-year-old man with a two-year history of a pruritic, painless erythematous skin rash on the inguinal region and scrotum is described. After a delay due to improper diagnosis and improper treatments, the patient came to the attention of the Division of plastic surgery. He underwent a punch biopsy and the pathology report came back as EMPD. Surgical excision was carried out, and an abdominal advancement cutaneous flap was performed for the defect repair. This is the first description of a reconstruction after surgical removal of inguinal EMPD with a flap of this type and we think that this type of treatment can be useful and reliable for disease localization in the groin area, especially for patients that present an excess of abdominal skin. ([www.actabiomedica.it](http://www.actabiomedica.it))

**Key words:** extramammary Paget disease, inguinal Paget disease, anogenital, Paget disease, surgical treatment

## Introduction

Sir James Paget, a British surgeon and physiologist, first described Paget's disease in 1874. In 1889, Radcliffe Crocker first described extra-mammary Paget's disease (EMPD) in a patient with urinary bladder carcinoma, who presented an eczematous lesion on the penis and scrotum. Darier and Coullaud described the perianal location of EMPD in 1893 (1).

The mammary type is rare and often associated with intraductal cancer (93-100% of cases). It is more prevalent in postmenopausal women (1). It appears as an eczematoid, erythematous, moist or crusted lesion, with or without fine scaling, infiltration and inversion of the nipple. Differential diagnoses are candidiasis, psoriasis, Bowen disease and chronic lichen simplex.

EMPD is an uncommon adernocarcinoma characterized by erythematous scaly patches or plaques occurring in apocrine gland-bearing skin such as the vulva, penis, scrotum, perineum, perianal area, and axilla (2).

Mammary and extramammary Paget disease are characterized by the presence of intraepithelial mucin-producing neoplastic cells known as Paget cells. However, the exact origin of these cells remains unclear (3).

The germline mutations in MMR genes are involved in the pathogenesis of EMPD and partially explain the genetic abnormalities for this disease (4).

The exact incidence of EMPD is not known, although it is more common in Caucasian postmenopausal women (1). EMPD represents just 6.5% of all Paget's disease and occurs mainly among in Caucasian women and Asian men in their 60s and 70s (5).

## Clinical Case

A 55-year-old man was referred to the Plastic Surgery Division of Siena (Italy) with a two-year history of a pruritic, painless erythematous chronic skin rash on the right emi-scrotum and inguinal region (Fig. 1).



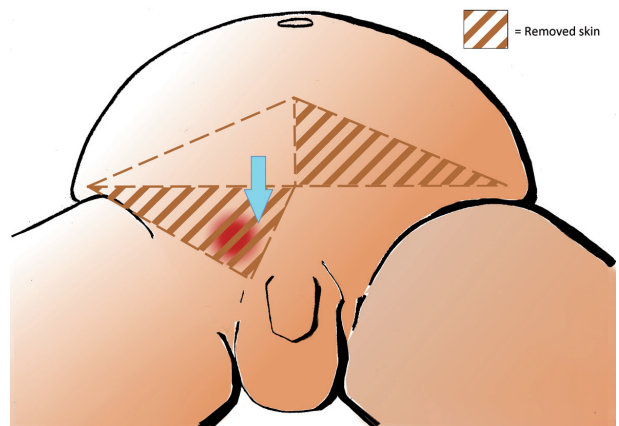
**Figure 1.** Pre-operative picture showing EMPD of the inguinal region

Because of progressive pruritus, erythema, and thickening of the affected area, he previously sought medical attention but misdiagnosis led to ineffective treatment (cortisone and antifungal topical agents). Local examination revealed an erythematous plaque extending over the base of the penis and 2-cm round peripheral rim onto the scrotum. The involved area contained whitish superficial exudates, and was scaly. No inguinal lymph nodes were evidenced by physical examination.

The patient initially underwent a punch biopsy and the pathology report came back as EMPD. Surgical excision was performed, and the repair of the defect was carried out by means of an abdominal advancement cutaneous flap (Fig. 2-3). The abdominal wall



**Figure 2.** Immediate post-operative picture



**Figure 3.** EMPD was surgically removed and an abdominal advancement cutaneous flap was used to cover the defect

was elevated immediately anterior to the underlying rectus and external oblique fascia caudally to the umbilical level.

The whole overlying skin and subcutaneous tissue immediately superficial to the fascia of the rectus abdominis and external oblique muscle was raised on the ipsilateral side of the defect. The flap was then moved caudally to cover the defect. The contralateral part of the flap was removed. This maneuver provided enough mobility for closing a gap of 12 cm. Superficial lymphadenectomy was performed at the same time. Lymph nodes were negative on histological examination. The patient, after 1 year of follow-up, is free from disease and has a normal life (Fig. 4).



**Figure 4.** Post-operative picture after 1 year of follow-up

## Discussion

The clinical case presented is of considerable rarity since in the Caucasians population men are rarely affected. Due to its rarity, differential diagnosis is difficult and diagnostic delays may substantially alter the prognosis.

In this case, the diagnostic delay resulted in treatment errors (cortisone ointment, antifungal creams) for suspected inflammatory diseases or fungal infections.

Current treatment for EMPD is surgical excision and Mohs' micrographic surgery. Studies using alternative therapies have been published. Photodynamic therapy, Imiquimod cream at 5% concentration, 5% 5-fluorouracil cream, CO<sub>2</sub> laser and the association of two or more therapeutic approaches showed variable results (6). Since relapses are very common with any of the available therapeutic options, close monitoring for long periods is mandatory. Metastatic EMPD systemic treatment includes FECOM 5-fluorouracil (5-FU, epirubicin, carboplatin, vincristine and mitomycin-C), 5-FU/cisplatin and single agent docetaxel or paclitaxel (7). Invasive pattern of strands/solid sheets is significantly associated with both lymph node metastasis and worse prognosis. Delay in diagnosis, depth of invasion, marked inflammation, lymphovascular invasion, and regional lymph node status are important prognostic factors (8).

Tabatabaei and McDougal already described the primary skin closure of large groin defects after inguinal lymphadenectomy for penile cancer using an abdominal cutaneous advancement flap (9).

To the best of our knowledge, surgical treatment of EMPD with an advancement abdominal cutaneous flap was never described before. Basic surgical technique is borrowed from classical abdominoplasty. The advancement abdominal cutaneous flap is a valid alternative to myocutaneous flaps for coverage of groin defects (10). In our opinion, this treatment option is an interesting approach for the treatment of such localizations in the groin area, especially when the patient shows an available excess of the lower abdominal skin.

## References

1. Lopes Filho LL, Lopes IM, Lopes LR, Enokihara MM, Michalany AO, Matsunaga N. Mammary and extramammary Paget's disease. *An Bras Dermatol* 2015; 90(2): 225-31.
2. Mun JH, Park SM, Kim GW, et al. Clinical and dermoscopic characteristics of extramammary Paget's disease: a study of 35 cases. *Br J Dermatol* 2015.
3. van der Linden M, Meeuwis KA, Bulten J, Bosse T, van Poelgeest MI, de Hullu JA. Paget disease of the vulva. *Critical reviews in oncology/hematology* 2016; 101: 60-74.
4. Kang Z, Xu F, Zhu Y, et al. Genetic Analysis of Mismatch Repair Genes Alterations in Extramammary Paget Disease. *Am J Surg Pathol* 2016 Aug 2. [Epub ahead of print] PubMed: 27487738.
5. Kang Z, Zhang Q, Zhang Q, et al. Clinical and pathological characteristics of extramammary Paget's disease: report of 246 Chinese male patients. *International journal of clinical and experimental pathology* 2015; 8(10): 13233-40.
6. Liao MM, Yang SS, Tan KB, Aw CW. Topical imiquimod in the treatment of extramammary Paget's disease: A 10 year retrospective analysis in an Asian tertiary centre. *Dermatologic therapy* 2016.
7. Phuoc V, Grothey A. Metastatic extramammary Paget's disease responding to weekly paclitaxel. *BMJ Case Rep* 2015; 2015.
8. Shu B, Shen XX, Chen P, Fang XZ, Guo YL, Kong YY. Primary invasive extramammary Paget disease on penoscrotum: a clinicopathological analysis of 41 cases. *Hum Pathol* 2016; 47(1): 70-7.
9. Tabatabaei S, McDougal WS. Primary skin closure of large groin defects after inguinal lymphadenectomy for penile cancer using an abdominal cutaneous advancement flap. *J Urol* 2003; 169(1): 118-20.
10. Ramasastry SS, Liang MD, Hurwitz DJ. Surgical management of difficult wounds of the groin. *Surgery, gynecology & obstetrics* 1989; 169(5): 418-22.

Received: 15 July 2016

Accepted: 25 August 2016

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