CASE REPORT

Reconstruction of the massive defect after excision of penoscrotal and perianal extramammary Paget’s disease

Yi-Shin Lu, Chu-Hsu Jeng*, Hwang-Chi Lin, Cheng-Han Hsieh, Yu-Hsien Lin, Kuo-Chuan Lo

Division of Plastic Surgery, Department of Surgery, Shin Kong Wu Ho-Su Memorial Hospital, Taipei, Taiwan

Received 7 March 2013; received in revised form 3 April 2013; accepted 5 June 2013
Available online 2 January 2014

KEYWORDS
extramammary Paget’s disease; gracilis myocutaneous flap; V-Y flap

Summary
Extramammary Paget’s disease (EMPD) is a tumor affecting mainly the genital region. Surgery remains the standard treatment for EMPD. Flaps were needed if there were massive defect after appropriate wide excision, especially when the tumor was very extensive. This study presents two cases of penoscrotal and perianal EMPD that needed flap reconstruction after wide excision of the tumor. Both cases were successfully reconstructed with flaps.

Copyright © 2013, Taiwan Surgical Association. Published by Elsevier Taiwan LLC. All rights reserved.

1. Introduction
Extramammary Paget’s disease (EMPD) is a tumor affecting mainly the genital region including the vulva, penis, scrotum, and perianal area. Most patients with EMPD have a good prognosis because the tumor cells generally extend only into the epidermis. For patients with an in situ tumor, wide local excision with a safe margin is the treatment of choice. However, a complete surgical excision is not always practical because the reported local recurrence rate is still high.1,2 Importantly, appropriate flap reconstruction after a wide local excision is needed if the tumor is very extensive. By contrast, a small proportion of EMPD is association with underlying or internal visceral carcinomaconsis. 3 Several investigators posit the existence of primary and secondary EMPD.6,7

In this study, we present two cases of penoscrotal and perianal EMPD that needed flap reconstruction after wide excision of the tumor.

Conflicts of interest: The authors have no conflicts of interest relevant to this article.

* Corresponding author. Division of Plastic Surgery, Shin Kong Wu Ho-Su Memorial Hospital, 95, Wen Chang Road, Shih Lin District, Taipei 111, Taiwan.
E-mail address: jeng8781@hotmail.com.tw (C.-H. Jeng).

http://dx.doi.org/10.1016/j.fjs.2013.06.007

Available online at www.sciencedirect.com
ScienceDirect
journal homepage: www.e-fjs.com

Formosan Journal of Surgery (2014) 47, 18–22
2. Case reports

2.1. Case 1

A 70-year-old male was admitted due to painless redness of the penoscrotal region for about 10 years. The patient complained of mild pain, heat sensation, and some discharge at the lesion site (Fig. 1). Outpatient treatment received elsewhere proved unsuccessful. Then the patient came to our outpatient department for help. The initial impression of condyloma was proven by biopsy on January 29, 2008. However, the symptoms failed to improve after 1 year. A second biopsy on July 24, 2009 showed Paget’s disease. The patient was then admitted for further management. The preoperative survey, included pelvic computed tomography and bone scan, showed no distal metastasis. Cystoscope revealed no other lesion in the bladder. Wide excision of the tumor (total penectomy) with a safety margin and reconstruction by a pedicled gracilis myocutaneous flap were done on August 11, 2009 (Figs. 2–4). Pathological examination showed infiltration of nests of pagetoid neoplastic cells in the lower epidermis with a margin free from disease (Fig. 5). The patient was discharged on September 1, 2009. There has been no local recurrence since this operation.

2.2. Case 2

A 69-year-old male with a history of diabetes mellitus and hypertension was admitted due to a perianal mass which had tended to bleed for about 2 years. He received treatment at another outpatient department, but results were not successful. There were perianal eczema and itching, followed by a tendency to bleed (Fig. 6). The biopsy showed Paget’s disease. No specific finding was found by rectal scope. No obvious distal metastasis was found by pelvic computed tomography scan. On October 1, 2009, a wide excision of the perianal tumor with a 2 cm safety margin was performed (Figs. 7 and 8). The wound was then reconstructed using a bilateral posterior thigh V-Y flap (Fig. 9). In addition, a temporary colostomy was done for wound care. The wound healing progressed smoothly and all sutures were removed after 3 weeks (Fig. 10). A physical examination of the anus showed adequate sphincter function and it was wide enough to accept an index finger, indicating an adequate diameter of the anus without serious scar contracture. No local recurrence has been noted.

2.3. Outcome

Both cases were successfully treated with a wide excision of the tumor with a 2 cm safety margin and reconstructed with a local flap.

3. Discussion

In 1874, Sir James Paget first described an intraepidermal neoplasm of the nipple, caused by an underlying breast carcinoma. Crocker described the first patient with EMPD involving the penis and scrotum in 1889. Perianal Paget’s disease was first described in 1893. Other sites of occurrence include the vulva, scrotum, groin, and axilla.

Due to different histogenesis, EMPD may be separated into two types. In primary EMPD, the most common type, there is no underlying carcinoma, and all lesions are located in the intraepithelial level. The malignant cell is suspected to originate from the intraepidermal apocrine glands or from pluripotent cells of the epidermis. There is the chance that the primary EMPD could invade the dermis and metastasize via lymphatic spread. By contrast, if the malignant cell is from an underlying apocrine carcinoma or...
internal malignancy, it is called secondary EMPD. Common visceral malignancies include carcinomas of the bladder, rectum, urethra, cervix, and prostate. About 15% of cases are associated with an underlying internal carcinoma.

EMPD often presents with a long-standing history of pruritic, erythematous patches over the anogenital and perineal areas, and always presents a poor response to topical treatment. Biopsy is frequently delayed. Lesions typically involve apocrine gland bearing skin such as the vulva, perineal, scrotal, perianal, and penile skin.

The prognosis for primary EMPD without deep dermal invasion is good with appropriate treatment. However, invasive EMPD has a high rate of metastasis. The depth of invasion is an important prognostic factor. The overall survival rate is poor; if the invasion extends into the subcutaneous tissue, and then regional lymph node metastases may occur. In cases of secondary EMPD, prognosis is related to the underlying carcinoma. According to the data of the Veterans General Hospital, Taipei, Taiwan, six of 29 patients (20.7%) died of the scrotal EMPD and three (10.3%) died of associated malignancy. But there are still insufficient data about associated malignancy or the histological characteristics of each lesion.13

Surgery remains the standard treatment for EMPD. However, high local recurrence rates are seen after a wide surgical excision with an adequate margin. This could be due to irregular margins or the multifocal nature of EMPD. Several papers have shown an overall recurrence rate of about 40% with a wide local excision.12,14 Local recurrence rates are higher in cases of invasive disease compared to those limited to intraepithelial involvement. More radical and extensive surgeries are associated with lower rates of local recurrence.

Mohs micrographic surgical excision (MMS) both improves cure rates and spares tissue around critical genitourinary structures.15 The recurrence rate after treatment with MMS is reported in 16% of primary EMPD. In 97% of cases treated with MMS, a safe margin of about 5 cm should be excised from the clinical tumor margin. With a surgical margin of 2 cm, only 59% of tumors are cleared, and the other 41% of EMPD are treated with the standard wide excision.

![Figure 4](image1.png)

**Figure 4** Case 1’s wound 1 week post operation.

![Figure 5](image2.png)

**Figure 5** Pathology of Case 1. (A) The biopsy specimen shows groups, clusters, or single cells within the epidermis (hematoxylin and eosin, 100×) that (B) shows nuclear enlargement with atypia, prominent nucleoli, and pale-staining cytoplasm (hematoxylin and eosin, 400×).

![Figure 6](image3.png)

**Figure 6** Local perianal mass with bleeding tendency in Case 2.
The two cases in this study had noninvasive primary EMPD. Due to the large tumor, both cases accepted a wide excision with a 2 cm safety margin. The defect was so big that a local flap was used for reconstruction. If we choose the 5 cm safety margin, the destruction around the critical genitourinary structures wound is very severe. We need to think about the balance between the benefit and the destruction of a large safe margin. Even if we use the 5 cm safety margin, local recurrence still could happen. It has not been proven whether more extensive surgery would be more effective in preventing local recurrence; the value of excision wider than a 3 cm safe margin is controversial. The recurrence rate has been reported to be about 12.5% in 24 patients who received wide local excision and the excision margin varied from 1 cm to 5 cm.13

Although surgery remains the traditional treatment for EMPD, radiotherapy may be used when patients are poor surgical candidates or when the level of genitourinary function after extensive surgery is a concern.16 Radiotherapy has also been used for local recurrence after surgery or as an adjuvant therapy in patients with a high risk of local recurrence.16

The experience of chemotherapy for EMPD still relies on case reports. Systemic chemotherapy has been used to treat patients with invasive and metastatic disease and may be considered in rare cases when surgery and radiotherapy are contraindicated. Limited reports of systemic chemotherapy to treat EMPD have been described, including a combination of low dose 5-fluorouracil and cisplatin17; a combination of 5-fluorouracil, cisplatin, mitomycin C, epirubicin, and vincristine18; and docetaxel.19

In conclusion, most EMPD cases have a good prognosis, with noninvasive EMPD being treated with surgical excision and careful evaluation of the tumor margin. Invasion level and lymph node metastasis are important prognostic factors in EMPD. Because no effective therapy yet exists for widely metastatic disease, the therapeutic strategy for advanced EMPD requires further investigation.

References