

## Perianal Paget's Disease

Chang Woo Kim, Yon Hee Kim<sup>1</sup>, Min Soo Cho, Byung Soh Min, Seung Hyuk Baik, Nam Kyu Kim

Division of Colon and Rectal Surgery, Department of Surgery, Severance Hospital, Yonsei University College of Medicine, Seoul; <sup>1</sup>Department of Pathology, Severance Hospital, Yonsei University College of Medicine, Seoul, Korea

The incidence of extramammary Paget's disease (EMPD) is very low. An 84-year-old Korean man was treated with topical and oral medications at a local dermatologic clinic for a year, but the symptoms did not improve. He visited Severance Hospital and underwent a perianal skin biopsy and was finally diagnosed with EMPD. The authors performed a wide local excision according to a 1-cm margin around the lesion. For the skin and the soft tissue defects, bilateral inferior gluteal artery perforator flap transpositions were performed. The size of the lesion was 14 cm<sup>2</sup> × 9 cm<sup>2</sup>, and the lateral and the basal margins were all disease free.

**Keywords:** *Extramammary Paget's disease; Perianal Paget's disease*

### INTRODUCTION

Perianal Paget's disease (PPD) is a subgroup of extramammary Paget's disease (EMPD), which is an uncommon intraepithelial neoplasm. The rate of PPD associated with malignancy ranges from 33% to 86% [1]. The true incidence of the disease is difficult to estimate due to its rarity, but it is known to represent less than 1% of all anal diseases and 6.5% of all cases of Paget's disease [2]. The authors report a case of a patient with PPD.

### CASE REPORT

#### Patient

An 84-year-old Korean man with hypertension and a history of appendectomy presented with perianal pruritus and a rash. He was treated with topical and oral medications at a local dermatologic clinic for a year, but his clinical symptoms did not improve. He visited Severance Hospital and underwent a perianal skin bi-

opsy, and was finally diagnosed with EMPD. The 6.5 cm<sup>2</sup> × 8 cm<sup>2</sup>-sized skin lesion was oval shaped and located from under the coccygeal area to the perineal body. The lesion abutted the anoderm 1 cm from the anal verge, but had not invaded the penis, testes, or both thighs. Abdomino-pelvic computed tomography and rectal magnetic resonance imaging were performed to evaluate preoperative staging and to detect other diseases. They showed no abnormalities, including no regional and inguinal lymph-node enlargement. A colonoscopy showed no intraluminal abnormalities. Thus, the authors decided to perform a wide local excision and to make flaps for the large area of skin and the soft tissue defects.

#### Surgery

The patient received a bowel preparation with polyethylene glycol one day before surgery because of the possibility of conversion to an abdominoperineal excision. He was placed in a jack-knife position on the table (Fig. 1A). Surgical margins 1 cm around the main lesion were demarcated based on a preoperative multiple-punch biopsy at eight locations which were all proven to be basal pigmentation or eczematous change. A wide local excision was performed (Fig. 1B). The size of the lesion was 6.5 cm<sup>2</sup> × 8 cm<sup>2</sup>. However, we carefully resected the lesion over the demarcation because the patient was too old to tolerate the time required for the frozen biopsy to confirm the proper margins. As a result, the final size of the specimen was 9 cm × 14 cm (Fig. 1C). For the skin and the soft tissue defects, bilateral inferior gluteal artery perforator flap transpositions were performed (Fig. 1D). Two closed suction drains were inserted at both flap sites. Repair and reinforcement of the dentate line with the skin were performed. A 28-Fr rectal tube was inserted into the

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Correspondence to: Seung Hyuk Baik, M.D.

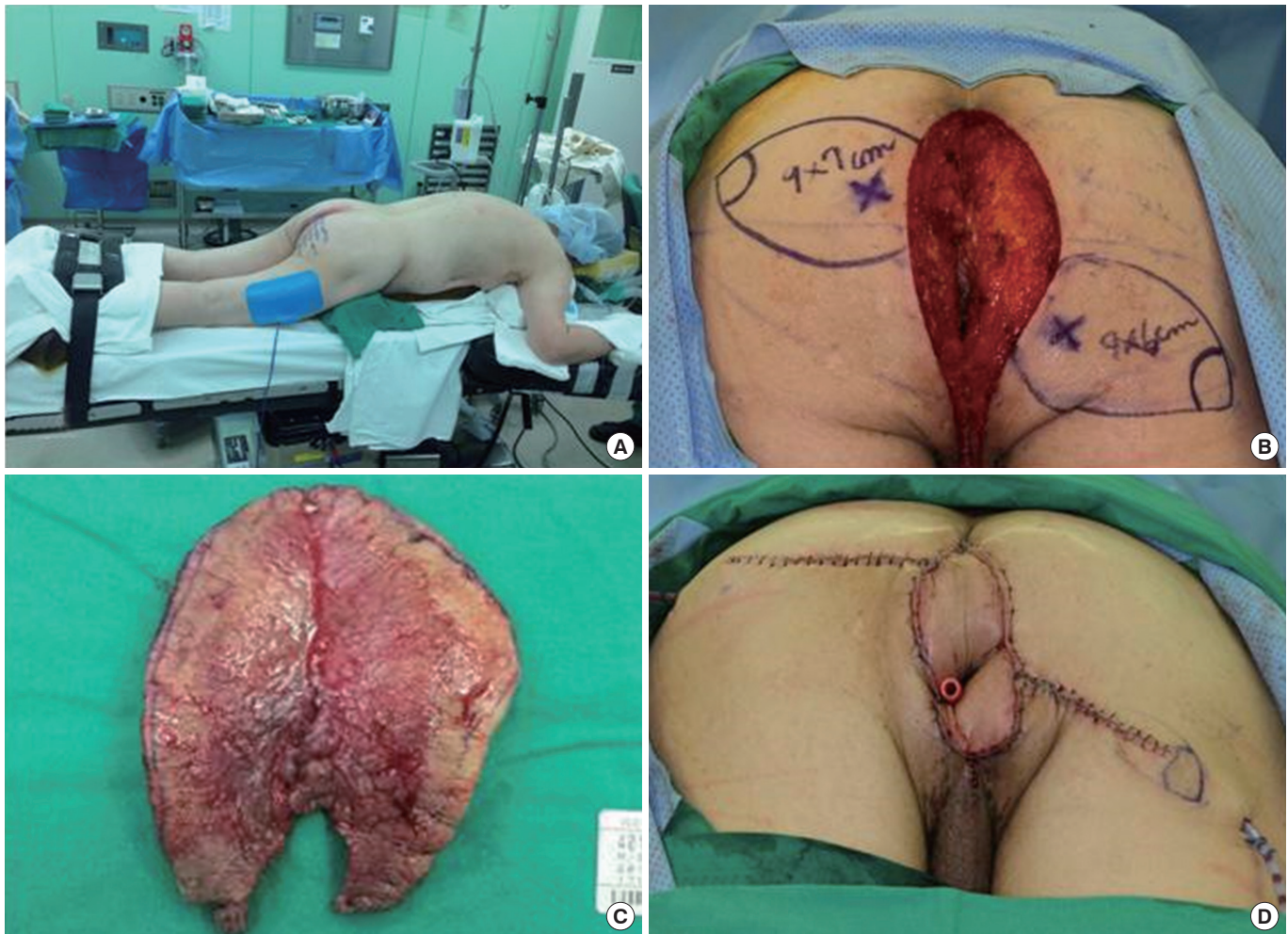
Division of Colon and Rectal Surgery, Department of Surgery, Severance Hospital, Yonsei University College of Medicine, 50 Yonsei-ro, Seodaemun-gu, Seoul 120-752, Korea

Tel: +82-2-2228-2100, Fax: +82-2-313-8289

E-mail: [whitenoja@yuhs.ac](mailto:whitenoja@yuhs.ac)

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**Fig. 1.** Position of the patient (A), after the wide local excision (B), the surgical specimen (C), and after the skin-flap transpositions (D).

anal canal. Aseptic dressing was done, and the patient was sent to the recovery room without any events. The total operation time was 182 minutes, and the estimated blood loss was 20 mL.

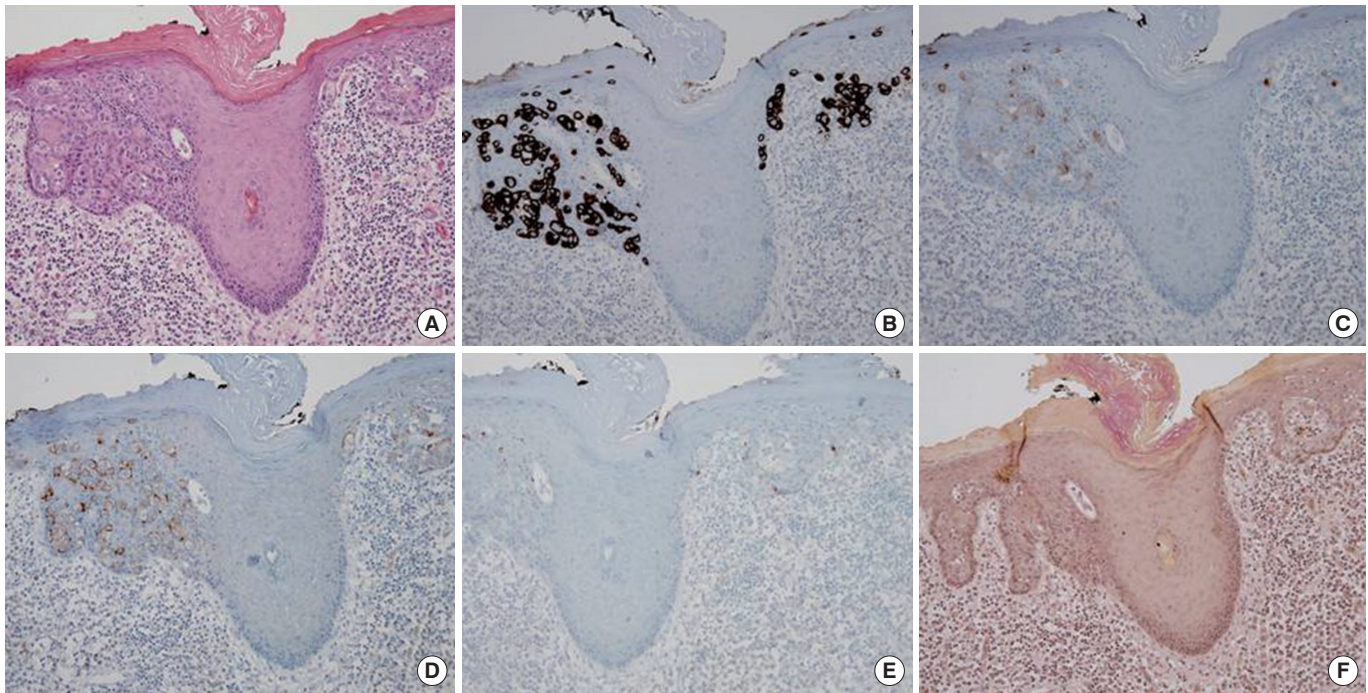
#### Postoperative course

The patient recovered without any complications. He sustained the prone position for the integrity of flaps and was kept on nil per os (nothing by mouth) with intravenous nutritional support alone for 12 days after surgery to prevent wound infection from the stool. The right drain was removed at postoperative day (POD) #6 whereas the left drain was removed at POD #12. The rectal tube was removed at POD #7. The patient started his diet at POD #13 without any problems. The pathology was proven as EMPD. The immunohistochemistry was positive for cytokeratin 7 (CK7), carcinoembryonic antigen, and C-erb B2 while it was negative for HMB-45 (Fig. 2). The lateral and the basal margins were all disease free (0.7 cm and 0.1 cm, respectively). The patient was discharged at POD #20.

#### DISCUSSION

Darier and Couillaud reported the first case of PPD 19 years after the first report of Paget's disease by Sir James Paget in 1874 [3]. Fewer than 200 cases of PPD have been reported in the literature, and the true incidence is difficult to estimate. PPD has also rarely been reported in Korea. Fewer than 20 articles on EMPD have been in Korea; moreover, no article on PPD has been found until now. A multicenter study on EMPD in Korea revealed only seven cases (3.6%) of PPD during 5 years [4].

The diagnosis of PPD can be obtained by using a histological examination showing the presence of Paget cells. In addition, CK7, CK19, and C-erb B2 are favorable immunohistochemical markers for the diagnosis of EMPD [5, 6]. However, it can be misdiagnosed easily without a histological confirmation due to its rarity and nonspecific clinical manifestation, which include exfoliative, exudative, verrucous, or hypopigmented patches that are accompanied by pruritus or burning sensations, which ultimately



**Fig. 2.** Immunohistochemistry examinations: (A) H&E ( $\times 100$ ), (B) cytokeratin 7 ( $\times 100$ ), (C) carcinoembryonic antigen ( $\times 100$ ), (D) C-erb B2 ( $\times 100$ ), (E) HMB-45 ( $\times 100$ ), and (F) mucicarmine ( $\times 100$ ).

mimic treatment-refractory eczema [4]. In the present case, the first impression at a local clinic was eczema and dermatitis. A treatment of topical steroid and oral antihistamine was applied, but was ineffective for a year. If a conventional treatment for general dermatitis is not effective for a certain period, PPD should be suspected, and a biopsy of the lesion should be considered.

Reaching a consensus on the treatment for PPD is difficult because PPD is rare, and most of the reports in the literature are case reports. Some authors have suggested nonsurgical treatment first. Kobayashi et al. [7] reported the effect of intralesional interferon. Cohen et al. [8] reported the results of the topical imiquimod in 9 cases of EMPD. Guerrieri and Back [9] suggested radiotherapy to control locally the disease for selected cases. However, many authors have recommended surgical treatment, including a wide local excision through abdominoperineal excision, for PPD. Shutze and Gleysteen [10] established a staging classification of PPD and recommended several treatment options. The present case corresponds to stage I according to their classification: Paget's cells found in the perianal epidermis and adnexae without a primary carcinoma. The recommendation for stage I and IIA is a wide local excision.

A wide local excision was performed safely after the demarcation of a margin of 1 cm based on a skin-punch biopsy. A safe resection margin of 2 cm is often recommended, although a clear margin of 1 cm could be safe for EMPD [1, 11]. The final pathologic report confirmed disease-free lateral and basal margins after the wide local excision. Skin or a muscle flap is usually used for

skin and soft tissue defects. Shutze and Gleysteen [10] described the defect as being covered with skin flaps for the skin and the defects in their two cases of PPD.

Long-term follow-up of patients with EMPD is important because of the possibility of recurrence or development of an associated cancer. Our patient recovered and was discharged without any complications. We are following up on the patient, and he has been alive for 3 months without relapse. A multicenter study on EMPD in Korea demonstrated that the clinical features of EMPD were consistent with reports from other Asian population-based studies, but the study showed that the rate of association with malignancy (14.4%) in the Korean population was higher than that observed in other Asian, such as 3%-5% in China [4].

## CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

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