PERIANAL PAGET'S DISEASE: REPORT OF A RARE CASE

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ABSTRACT

Introduction: Perianal Paget's disease is a rare skin disorder of unknown aetiology, which is frequently associated with malignancy. We aimed to analyze clinical and pathological characteristics of perianal Paget's disease (PPD) with considering relevant literature.

Case report: We present a 44-year-old male patient admitted to the hospital with perianal eczema-like lesions and pruritus. Following the incisional biopsy of a perianal skin was showed as perianal Paget's disease, the patient underwent a surgical en bloc resection of the lesion with wide negative margins and the defect closed with a local transposition flap. The histopathologic examination revealed a perianal Paget's disease. The patient made an uneventful recovery and was discharged home on the eight postoperative days. No adjuvant therapy was given. After fifteen months follow-up, the patient has no evidence of local recurrence with good anal function and cosmetic result was good.

Conclusion: Although, the choice of the optimal treatment for perianal Paget's disease (PPD) remains controversial, surgery is still the main stay of radical treatment modality.

A thorough initial evaluation and long-term follow-up is very important to identify both an underlying primary malignancy and recurrence.

Key words: Paget's disease, extramammary, perianal disease

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INTRODUCTION

Perianal Paget's disease (PPD) is an uncommon intraepidermal adenocarcinoma, characterized by the presence of atypical Paget's cells(1). Paget's disease has been reported in several extramammary sites, including the axilla, perineum, groin, thigh, scrotum, and vulva(1). Perianal Paget's disease firstly was reported in 1893, by Darier and Coulillaud (1). The most publications of the reported cases have appeared as case report and up to date, total about 200 cases of perianal Paget's disease have been described in the literature (2).

Here in, we report a 44-year-old patient with PPD disease and attempt to review the relevant publications.

CASE REPORT

A 44-year-old man was admitted to our clinic because of perianal eczema-like lesions and pruritus (Figure 1).Despite of the patient received medical treatment with topical cortisone cream for two months, it was not seen any improvement. At clinical assessment, there was large, whitish, eczematoid plaque affecting the whole perianal area. An eczematous erythema was measured and found to be 5-6 cm around the perianal skin, involving the anal verge. Histologic examination of the incisional biopsy reported as PPD disease.

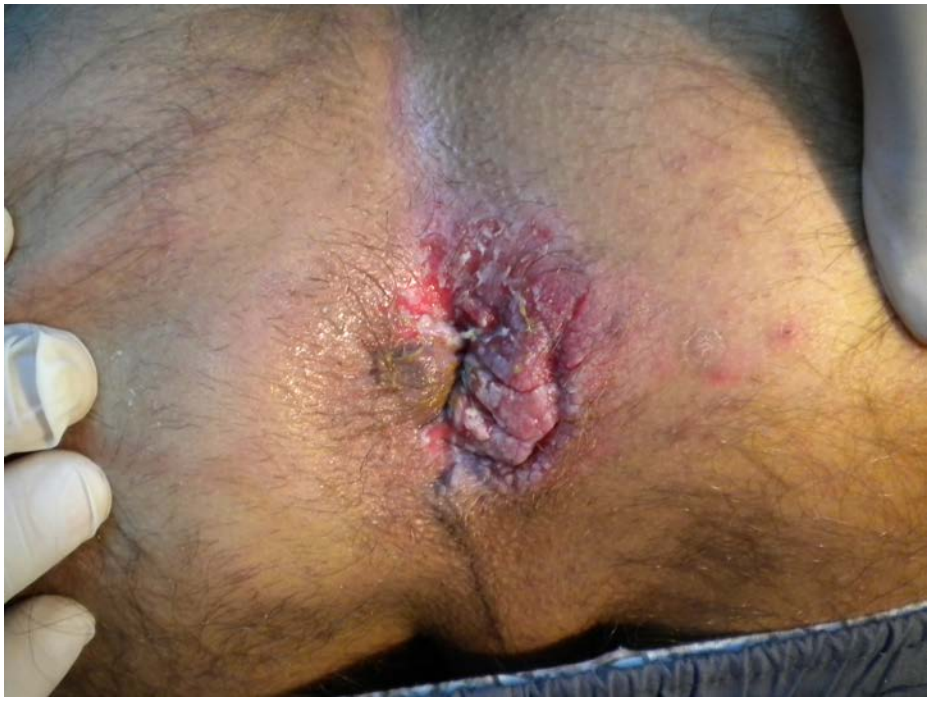

Figure 1. Perianal Paget's disease (preoperative appearance). It is seen as the skin lesion that is sharply marginated erythematous plaque with silvery-whitish areas.

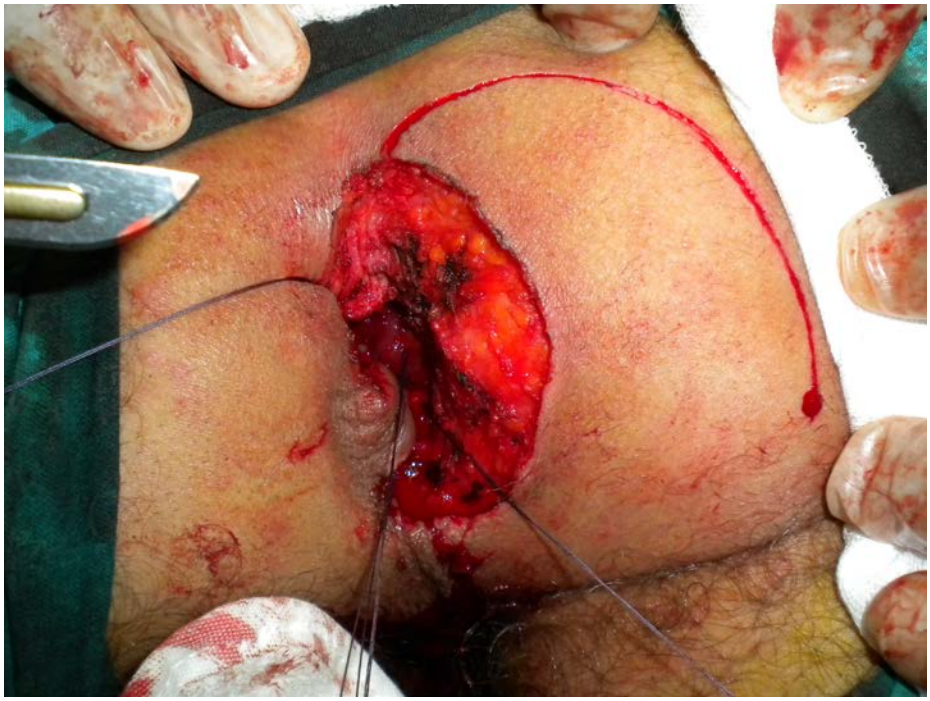

Figure 2. **The lesion is removed totally with the negative margins both on frozen section and permanent section.** A skin rotation flap is prepared to cover large perianal skin defect.

The patient was operated in the lithotomy position. A wide excision of the subcutaneous tissue was made with a safety margin of approximately 3 cm. A rhombic gluteal fold was prepared a full-thickness pedicle flap graft (Figure 2). Microscopic examination of excised specimens showed incidental Paget's disease of the anal skin. The epidermis showed a follicular focal infiltration by large, round Paget's cells that had swollen nuclei and bright bodies (Figure 4).All surgical margins were found to be negative. The patient made an uneventful recovery and was discharged home on the eight postoperative days. No adjuvant therapy was given. Follow-up at 15 months showed good cosmetic result (Figure 4).

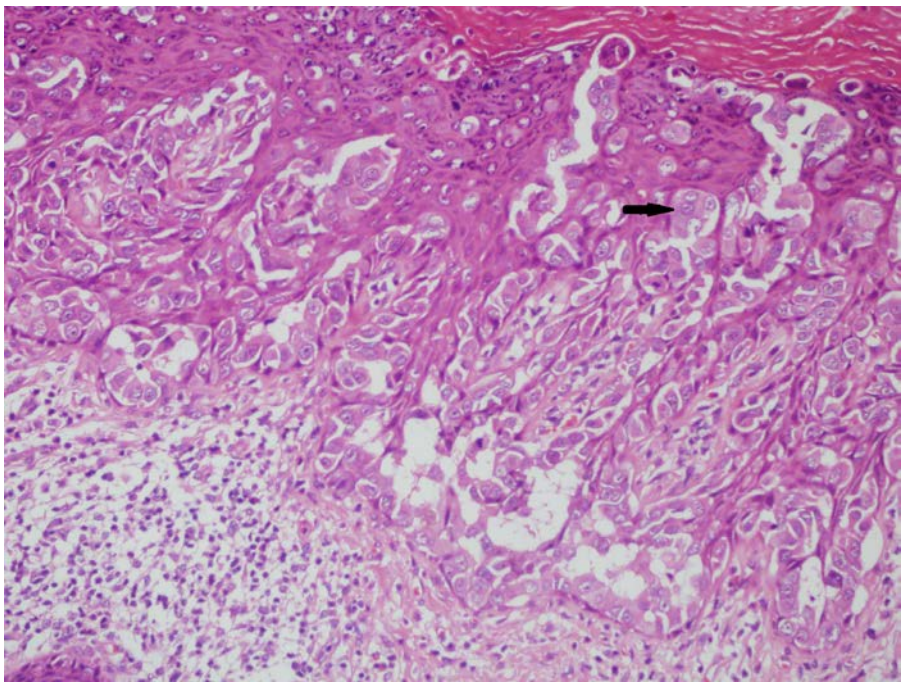

Figure 3. **Histopathological images of perianal Paget's disease. (H&E, x240).** Many Paget's cells with clear cytoplasm and large pleomorphic nuclei within dermis are present.

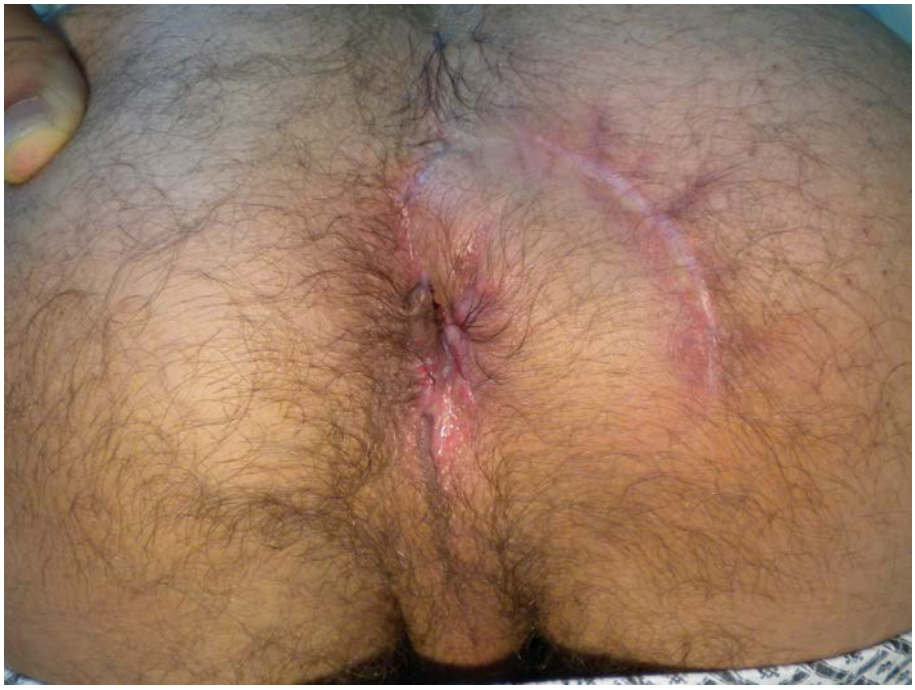

Figure 4. Three months post-operative appearance of the case.

DISCUSSION

Perianal Paget's disease is a very rare intraepithelial adenocarcinoma, arising from dermal apocrine sweat glands (2). It has been reported that more commonly in women than in men and seems to be more frequently found between 50-70 years of age. The true incidence of this disease is not exactly known due to its rarity (3). The incidence of associated malignancies with PPD ranges from 33 to 86% (4).

Clinical features of PPD are non spesific and include well demarcated, slightly raised and erythematous lesions, pruritus, irritation, and rash. Licheniced, leucokeratotic or leucoplakia-like patches may also be seen in some patients. Therefore, the diagnosis of PPD is often delayed. It has been suggested that the ano-genital eczematoid lesions are not responsive to 6-8 weeks of topical therapy should be biopsies (5-7).

It is reported that local, regional and systematic extent of the PPD is the most important factor in determining treatment modality choice (2). Currently, surgical treatment methods are still the mainstay in the management of PPD. Shutze et al, in 1990, recommended the stage and management of PPD disease, as shown to table 1(8).

Wide local excision with free surgical margin is the mainstay of PPD treatment in the absence of invasive cancer. A wide skin excision can be caused with circumferential loss of skin all around the anus, which is the case with our patient, requires a skin graft for reconstruction. The various techniques included myocutaneous flaps of the gluteal and thigh muscles, transposition or rotation local skin flaps and V-Y island flaps have been described in the literature for the treatment of these defects (2,7).We used rotation local skin flaps in our case. In contrast to, an inadequate surgical margin will cause local recurrence. However, local recurrence rate was reported between 31% and 61% because surgical margins can be interpreted as appearing negative intra-operatively yet proving to be positive in later permanent histological analysis (7).

Although several authors have advocated the benefits of non-operative other modalities of treatment including radiotherapy chemotherapy, photodynamic therapy, topical imiquimod, topical chemotherapy and topical ALA and CO_2 laser, the long-term outcomes of this therapy modalities are not known due to its rarity (8).Radiotherapy can be used as the primary therapy in patient's not fit or refusing surgery. Brown et al reported an article about roles for radiotherapy and indications in PPD. They proposed that radiotherapy of PPD has two main roles, the first is as primary treatment for in situ or invasive disease and the other is following surgical relapse of in-situ or invasive disease (7).

Recurrence or progressions of PPD remain one of the most important problems after surgery. It may be seen even after wide local excision at obtaining disease-free resection margins (8). Mc Carter et al reported, the largest series from the Memorial Sloan-Kettering Cancer Center including 27 patients treated between 1950 and 2000

(1).They had a recurrence rate of 30% in patients with WLE and they identified an invasive cancer in 44% of the patients. In this study, the overall disease-free 5-year survival rate of patients without an invasive component was 64% compared with 59% in those with an invasive component. In this study, it has been clearly shown that the high incidence of recurrence and poor outcome associated with invasion.

Conclusionally, PPD are often associated with gastrointestinal adenocarcinoma synchronous, so that a full assessment of the intestinal tract should be done. Although surgery remains the mainstay of treatment at present, due to the rarity of this condition, the optimal treatment is unclear. It should be kept in mind that Paget's disease has the capacity to become invasive. The patients with Paget's disease have risk of developing recurrent cancer and careful prolonged follow-up is needed.

No conflict of interest was declared by the authors.

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