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Total vaginectomy and radical vulvectomy for extension of extra-mammary Paget's disease

Byung Su Kwon[1], Yong Il Ji[2], Min Hyung Jung[3], Young Sun Kim[4]

[1] Department of Obstetrics and Gynecology, Seoul National University, College of Medicine, Seoul
[2] Department of Obstetrics and Gynecology, Haeundae Paik Hospital, Inje University College of Medicine, Busan
[3] Department of Obstetrics and Gynecology, Kyung Hee University, College of Medicine, Seoul
[3] Department of Obstetrics and Gynecology, Soonchunhyang University Gumi Hospital, Soonchunhyang University College of Medicine, Seoul (Republic of Korea)

Summary

Extra-mammary Paget's disease of the vulva is a rare non-invasive adenocarcinoma that usually occurs in postmenopausal women. Histologically, it often extends beyond the visible lesion, leading to positive surgical margins and frequent recurrences, but can be managed by simple vulvectomy or wide local excision. Although current evidence supports the use of radical surgery as an alternative to the generally performed wide local excision in the treatment of widely extended extra mammary Paget's disease of the vulva, nonetheless there controversy still exists regarding the extent of an adequate resection margin. Here the authors present a case of successful radical vulvectomy with total vaginectomy without adjuvant treatment on a delayed diagnosis of extra-mammary Paget's disease, extending from the vulva to the apex of vagina.

Key words: Paget's disease; Total vaginectomy; Non-invasive adenocarcinoma; Vulvectomy.

Introduction

Extra-mammary Paget's disease of the vulva is a rare tumor accounting for less than 2% of all vulvar malignancies mainly affecting postmenopausal women [1-3]. The lesion is defined as an intra-epidermal neoplasm and may be accompanied by an invasive or in situ adenocarcinoma of the apocrine glands [4]. Although the clinical manifestation of extra-mammary Paget's disease varies, it begins insidiously with pruritus and a burning sensation with observed flaking, oozing, and maceration [4]. Differential diagnoses includes Bowen disease, superficial fungal infection, psoriasis, leukoplakia, and eczematous dermatosis [5].

Wide local excision with or without inguinofemoral lymphadenectomy is initially performed as the primary treatment in all patients. While adjuvant radiotherapy is considered in patients with positive surgical margins or lymph node metastasis [1]. Nonetheless, surgical treatments for non-invasive vulvar extra-mammary Paget's disease accounts for significant local recurrence rate ranging between 30–60%. This may be due to clinically ill-defined areas of extension and multicentric foci of the tumor cells [6].

In this paper, the authors present a case of successful radical vulvectomy with total vaginectomy without adjuvant treatment on a delayed diagnosis of extra-mammary Paget's disease, extending from the vulva to the apex of vagina.

Case Report

A 76-year-old postmenopausal woman presented to the dermatology department with recurrent vulvar skin irritation, itching of the labia major, and minora of two years duration. The patient had a history of papillary urothelial carcinoma of bladder and underwent an open radical cystectomy and hysterectomy with both salpingo-oophorectomy. She was treated with topical steroids and antimycotics for one month, without any improvements.

During physical examination, a distinct vulvar lesion was seen in the labia minora and majora involving the upper part of the vagina (Figure 1a). Four quadrants vulva and two vagina excisional biopsies were taken. Histopathological finding showed non-invasive extramammary Paget's disease in all quadrants of the vulva, extensively extending to the upper vagina mucosa (Figure 2). She was referred to the obstetrics and gynecology department. Pelvic magnetic resonance image showed no evidence of local metastasis (Figure 1b). Positron emission tomography-computed tomography also showed no abnormal hypermetabolic lesion indicative of metastasis (Figure 1c).

Surgical treatment including radical vulvectomy with total vaginectomy for Paget's disease that originated in the vulva and extended to the vaginal margins was performed. Involvement of vaginal resection margin was performed by frozen sectionings (Figure 3a) followed by bilateral fasciocutaneous advancement flap coverage by the plastic surgery department (Figure 3b). One year later, she had a no recurrence on a follow-up direct vulva and vagina biopsy and pruritus resolved with improvement in quality of life.

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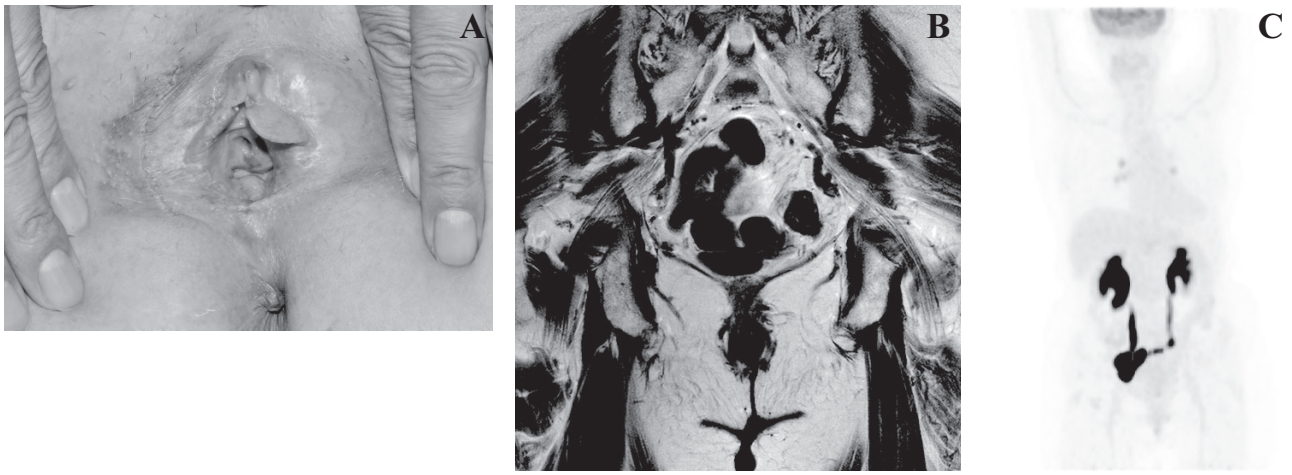

Figure 1. — a) Preoperative photograph of the lesion. b) Pelvic magnetic resonance image showing no evidence of local recurrence or metastasis. c) Positron emission tomography-computed tomography showing no abnormal hypermetabolic lesion indicative of local metastasis.

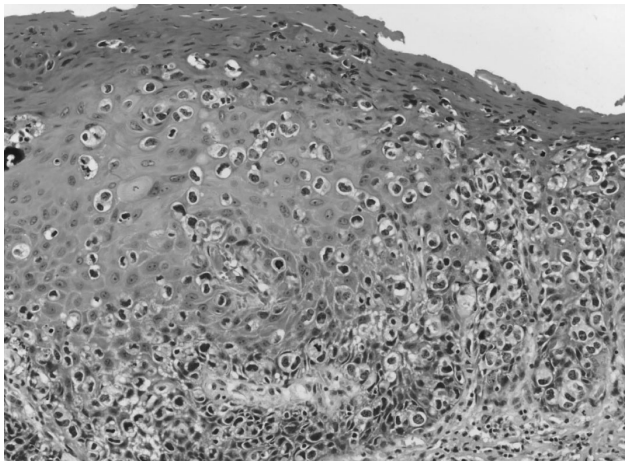

Figure 2. — Epidermis with keratinization and pagetoidcell infiltration of epidermis by malignant cells with a clear cytoplasm (H&E ×200).

Discussion

James Paget's published the first description of Paget's disease in 1874 that involved the nipple and areola, while Crocker first described extramammary Paget's disease as a rare disorder in 1889. The most common site of involvement is the vulva, followed by the axillae, penis, and, less frequently, eyelids, umbilicus, and groin [4]. It often spreads in an occult fashion, with margins extending beyond the apparent edges of the lesion [7].

Diagnosis may be delayed because of non-specific clinical findings and could lead to several inappropriate treatments including extended periods of topical steroids. About one year can be spent before biopsy is taken and definitive diagnosis is made.

Although extension of extra-mammary Paget's disease of the vulva into the upper vagina and cervix has only rarely been described in a delay in diagnosis, previous reports have

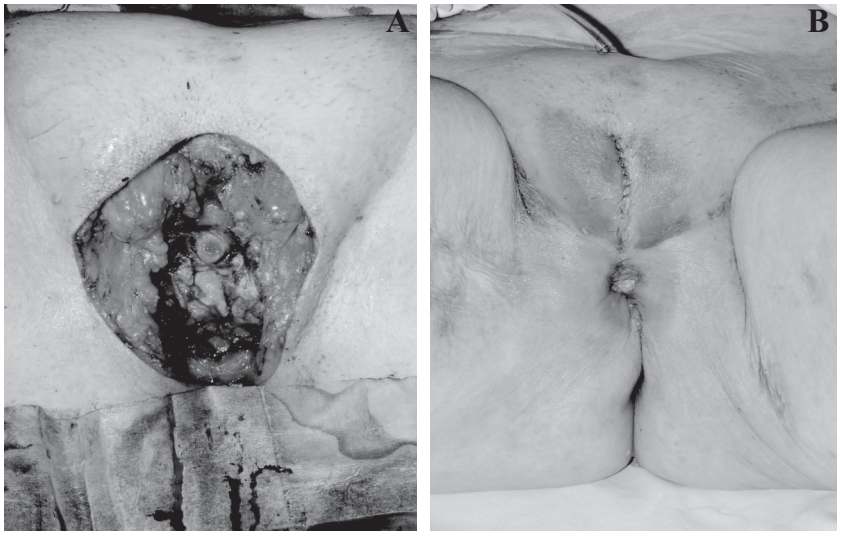

Figure 3. — a) Intraoperative photograph post resection showing a wide resection margin. b) Closure of the wound after cystostomy.

indicated that a delay in diagnosis for >12 months was associated with a multifocal lesion, which required extended resection [8, 9]. For this reason, all patients with non-specific vulva findings should undergo clinical evaluation and early biopsy by a skilled physician if there is no response to standard therapy [9]. In this case, the patient had recurrent dermatitis in the labia minora and majora for two years, with no symptom improvement despite treatment. Histopathological examination of vulvar and vaginal biopsy specimens showed non-invasive extramammary Paget's disease in all labia minora and majora of the vulva, extensively extending to the apex of vagina mucosa.

Even though complete surgical resection is the current treatment of choice, Paget's disease of vulva is generally non-invasive so often can be managed by simple vulvectomy or wide local excision. However, this disease is notorious for its high local recurrence rate (30–60%) mainly due to clinically ill-defined areas of extension and multicentric foci of the tumor cells [6].

Some studies found that positive margins is the risk factor of increasing recurrence rate [9, 10]. However, controversies still exist about it. The mean time of recurrence was shorter when resection margin is positive. Also, in many cases, local recurrence usually occurred outside the previous margin. It remains the most challenging feature in the management of vulvar intraepithelial Paget's disease.

Some suggested using intraoperative frozen section analysis of biopsies from the perimeter of the planned area of resection to obtain negative pathologic margin, while decreasing the risk of positive surgical margin status. Unfortunately, frozen-section analysis can be misleading, appearing negative intraoperatively but proving to be positive on later permanent analysis. Therefore, intraoperative frozen section evaluation of the surgical margins may not be helpful in reducing the recurrence rate [9, 10].

Here, the authors recommend performing radical vulvectomy and total vaginectomy from the vulva to the apex of vagina mucosa to prevent local recurrence instead of wide local excision. Long term follow up is necessary to exclude recurrence and newly developing associated cancer.

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Address reprint requests to:
YOUNG SUN KIM, M.D.
250, Gongdan-dong, Gumi
Gyeongsangbuk-do
Republic of Korea
e-mail: chacha0725@naver.com