Paget's disease of the vulva - a review of our experience

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Summary

Objective: The aim of our study was to review our experience with Paget's disease of the vulvar relative to initial examination, treatment and oncological outcome.

Methods: Ten women with extramammary Paget's diseases of the vulva were treated during the 10-year period. The charts of these patients were reviewed and data were collected regarding patient demographics, symptoms, previous Paget's disease, surgical treatments and time to recurrence.

Results: Eight women (80%) were treated with wide local excision or partial vulvectomy, and two patients (20%) required radical resection for invasive adenocarcinoma. The group of women who had invasive diseases also underwent inquinofemoral lymphadenectomy and no lymphatic metastases were noted. Three women (30%) experienced recurrence. The mean time to relapse was 30 months (range 3-88 months).

Conclusion: Recurrence is very common and long-term monitoring is recommended with careful examination of any abnormal vulvar lesion.

Key words: Paget's disease; Vulva.

Introduction

Paget's disease of the vulva (PDV) is a type of intraepithelial neoplasia that arises from multipotential stem cells in the epidermis and epidermally derived adnexal structures [1]. PDV is thought to be commonly associated with an underlying subjacent glandular apocrine adenocarcinoma analogous to the situation with Paget's disease of the breast [1]. PDV affects mainly postmenopausal women of European origin with a mean age at diagnosis of 64 years [1].

It begins in the apocrine gland-bearing regions of the vulva, genital folds, and perianal region and may be multifocal [2].

The condition presents with pruritus in over 50% together with soreness, burning, bleeding, and pain [2]. It may either appear as red velvety areas with white islands of hyperkeratosis or be pinkish and scaly. The diagnosis is made by biopsy and there is distinctive histological appearance with pathognomonic Paget cells [2, 3].

Methods

Ten patients with extramammary Paget's diseases were treated at the Institute of Gynecology and Obstetrics, Clinical Center of Serbia during the period 1995 to 2000. The charts of these patient were reviewed and data were collected regarding patient demographics, symptoms, previous Paget's disease, surgical treatments and time to recurrence.

Wide local excision was performed in cases of lesions with a small amount of subcutaneous tissue. Radical vulvectomy with node dissection was performed when invasive disease was found. The gross lesion was outlined with a 1-2 cm border, and strips of tissue beyond this border were excised.

Invasive disease was defined as Paget's disease that had spread beyond the basement membrane.

Patient follow-up consisted of furthur care in our institution with long-term monitoring and repeated excision of symptomatic lesions.

Results

The ten women with Paget's disease of the vulva were treated at our Institute during the period 1995 to 2000.

The average age of patients was 58 years (range 46-84 years).

The most frequent symptoms were a burning sensation (88%), lesion (76%), itching (75%) and watery discharge (55%). Also dysuria was found in 5% of cases. Symptoms presented for an average of 20 months (range 2-27 months). The gross extent of disease varied greatly from 2 cm to involvement of almost the entire vulva.

Two of our patients (20%) had been treated previously for Paget's disease before our involvement.

There were two patients (20%) with other primary cancers; one had had breast cancer and another one had had bladder cancer. These cancers were diagnosed and treated before the diagnosis of Paget's disease of the vulva.

Treatment included wide local excision or vulvectomy depending on the extent of disease. Patients with invasive disease underwent radical vulvectomy.

Eight women (80%) were treated with wide local excision or partial vulvectomy, and two patients (20%) required radical resection for invasive adenocarcinoma. The group of women who displayed invasive diseases also underwent inquinofemoral lymphadenectomy and no lymphatic metastases were noted. In these two patients invasive disease was identified by biopsy before surgery. Both of the patients with invasive disease had had previous treatment for Paget's disease.

Revised manuscript accepted for publication July 3, 2006

Three women (30%) experienced reccurence. The mean time to relapse was 30 months (range 3-88 months). One of them had had invasive disease at the initial surgery. Each symptomatic recurrence was treated with wide local excision.

Discussion and Conclusion

Paget's disease of the vulva remains a rare condition that accounts for < 1% of vulvar neoplasms [3]. It was originally described as a breast lesion with associated underlying invasive ductal adenocarcinoma. It was later classified according to the location on the body as mammary or extramammary disease [3].

In most cases, PDV is an eczematous dermatosis with a characteristic lesion which is histologically confined to the epidermis and epidermal appendages [4]. The lesion is apocrine in origin and usually confined to the epithelium. However, invasive disease is present in up to 15% to 25% of patients.

Occasionally, a clinically similar lesion of the vulvar skin is associated in 25% of cases with an adenocarcinoma that arises from the underlying apocrine or eccrine glands. Commonly associated malignancies are breast, basal cell, rectal, genitourinary and cervical carcinomas [5].

The management of PDV is defined by the need to exclude invasive PDV, an underlying adnexal adenocarcinoma and a concurrent carcinoma [6]. Thus women with PDV should undergo investigations for other malignancies including at least colonoscopy, mammography and Papanicolaou smear [6]. Any suspicious symptoms of other malignancies should guide additional evaluation.

The need to exclude invasive PDV and an underlying adnexal adenocarcinoma means that the tumor must be excised [6, 7]. If such a tumor is known to be present, the treatment should be the same as for invasive adenocarcinoma of the vulva. If it is not known whether or not there is an underlying carcinoma, the vulvar skin and underlying appendages should be removed [6]. Previously radical vulvectomy was performed, but has been replaced by more conservative surgery, including skinning vulvectomy with split-thickness skin-graft, hemivulvectomy or simple vulvectomy [6, 7].

Paget's disease classically has microscopic extension beyond the gross lession. To ensure clear margins, frozen section may be used during the surgical procedure [8]. A problem with these efforts is that Paget's disease often has a patchy type of distribution with multiple islands of abnormal epithelium that are not connected [8].

The consensus regarding treatment is that wide local excision is adequate for specimens without invasive disease [8, 9] as was carried out in our patients' cases. Wide margins around the visible abnormality seem sensible because the histological abnormality extends much wider than the visible abnormality. Some authors recommend selective use of intraoperative frozen section analysis of the vulvectomy specimen, not for margin status but to determine whether invasive disease is present [9]. Grossly negative margins are probably adequate, and it seems reasonable to limit the extent of resection in favor of the preservation of the clitoris, urethra, and anus [9].

When an underlying carcinoma is present the prognosis is poor. Patients with non invasive PDV do not die of their disease, but do develop recurrent disease and may develop invasive cancer [8, 9]. PDV may reccur in an area previously totally excised and transplanted with autologous skin. Usually recurrences are amenable to local therapy and treatment options include further local excision, as was done in our patients, laser ablation and topical 5-fluorouracil cream [8, 9].

After treatment about one-third of women experience local recurrences over many years which is close to our results (30%). These patients require long-term follow-up and careful examination of any abnormal vulvar lesion that develops [9].

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