The incidence of vulvar pathology and vulvectomy within risk and non-risk groups

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Summary

At the Institute of Gynecology and Obstetrics over a period of five years, more than 180,000 women were examined; 580 simple and 178 radical vulvectomies were performed.

Key words: Simple vulvectomy; Radical vulvectomy.

Introduction

Vulvar neoplasms

Malignant tumors of the vulva are uncommon, representing about 3%-4% of malignancies of the female genital tract [4]. Most tumors are squamous cell carcinomas, with melanomas, adenocarcinomas, basal cell carcinomas, and sarcomas occurring much less frequently [4].

Squamous cell carcinoma of the vulva occurs mainly in postmenopausal women, and the mean age at diagnosis is 65 years [4]. A history of chronic vulvar itching is common [4]. Vulvar cancer tends to be found more frequently in patients who are obese and in those who have hypertension, diabetes mellitus, or arteriosclerosis, although these conditions do not appear to be specific risk factors. Other primary malignancies have been reported in up to 22% of cases, the most common primary site being the cervix [3].

Recent studies suggest two different etiologic types of vulvar cancer [1]. One type, seen mainly in younger patients, is related to human papillomavirus infection and smoking and is commonly associated with vulvar intraepithelial neoplasia (VIN). The more common type, seen mainly in elderly women, is unrelated to smoking or human papillomavirus infection and concurrent VIN is uncommon. Carcinoma in situ of the vulva (VIN III) appears to carry a significant risk of progression to invasive cancer if left untreated [2].

About 5% of patients have positive results on serologic testing for syphilis. In the latter group of patients, vulvar cancer occurs at an earlier age and carries a graver prognosis [2]. Although rarely seen in the United States, vulvar cancer also occurs in association with lymphogranuloma venereum and granuloma inguinale.

Intraepithelial neoplasia

The International Society for the Study of Vulvar Disease recognizes two varieties of intraepithelial neoplasia: squamous cell carcinoma in situ (Bowen's disease) or VIN III and Paget's disease [2].

Squamous cell carcinoma in situ: VIN III

During the past two decades, the incidence of carcinoma in situ of the vulva has increased. Younger patients are being affected, and the mean age is approximately 45 years [4].

Itching is the most common symptom, although some patients present with palpable or visible abnormalities of the vulva [4]. Approximately half of the patients are asymptomatic and there is no absolute diagnostic appearance. Most lesions are elevated, but the color may be white, red, pink, gray, or brown. Approximately 20% of the lesions have a "warty" appearance, and the lesions are multicentric in about two-thirds of cases [4].

Careful inspection of the vulva in a bright light, with the aid of a magnifying glass if necessary, is the most useful technique for detecting abnormal areas. In a patient with pruritus vulvae and no gross abnormality, colposcopic examination of the entire vulva after the application of 2% acetic acid is helpful. The toluidine blue dye test (Collin's test) may also help direct biopsies. Because the dye fixes to cell nuclei, false-negative results may be seen in the presence of hyperkeratosis, and false-positive results may be seen in the presence of excoriations. A liberal number of directed biopsies must be taken to establish the diagnosis and rule out invasive carcinoma.

A number of methods of treatment are used for carcinoma in situ of the vulva. In the past, total vulvectomy was usually performed. It is now clear that the incidence of recurrence following total vulvectomy (about 30%), is not less than that following local excisions of the individual lesions. Because of the distressing psychological consequences of vulvectomy, local superficial excision is now considered the mainstay of treatment.

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Microscopic disease seldom extends significantly beyond the macroscopic lesion, so margins of about 5 mm are usually adequate [3]. For extensive lesions involving most of the vulva, a "skinning' vulvectomy in which the vulvar skin is removed and replaced by a split-thickness skin graft, may be used. Because subcutaneous tissues are not excised, the cosmetic result is superior to that obtained with vulvectomy.

Topical 5-fluorouracil cream is effective in about 50% of cases, but patient tolerance is low because of the painful ulceration that results [5]. Laser therapy is also effective, particularly for multiple small lesions. When large areas of the vulva are treated with laser therapy, postoperative pain is severe and patient tolerance for this procedure is low [6]. Topical chemotherapy and laser therapy offer the optimal cosmetic outcome, but because no specimen is available for histologic study, several biopsies must be taken before treatment to exclude invasive disease.

Bowenoid papulosis of the vulva

Bowenoid papulosis is a clinical entity that usually affects younger individuals. It is characterized clinically by multiple reddish brown or violaceous papules on the vulva or penis and histologically, it is indistinguishable from carcinoma in situ. A viral cause has been postulated but not proved [6]. Treatment should be by local excision or laser therapy. Some lesions may regress spontaneously after pregnancy.

Paget's disease

Paget's disease of the vulva predominantly affects postmenopausal white women [4]. Paget's disease also occurs in the nipple areas of the breast.

Itching and tenderness are common and may be long-standing. The affected area is usually well demarcated and eczematoid in appearance, with the presence of white plaque-like lesions. As growth progresses, extension beyond the vulva to the mons pubis, thighs, and buttocks may occur; rarely, it may extend to involve the mucosa of the rectum, vagina, or urinary tract. In 10% to 20% of cases, Paget's disease is associated with an underlying adenocarcinoma [4].

The disease is characterized by large, pale, pathognomonic Paget's cells, which are seen within the epidermis and skin adnexa. They are rich in mucopolysaccharide, a diastase-resistant substance that stains positive with periodic acid-Schiff. The intracytoplasmic mucin may also be demonstrated by Mayer's mucicarmine stain. The histogenesis of this lesion has been controversial, but it is currently believed to be a type of adenocarcinoma in situ [4]. Paget's cells are typically located adjacent to the basal layer, both in the epidermis and in the adnexal structures.

Unlike VIN III, the histologic extent of Paget's disease is frequently far beyond the visible lesions. Hence, wide local excision is required to clear the lesion, and frozen sections should be obtained on the margins of resection. Recurrences still occur in approximately 30% of patients [4] and may be treated by further excision or laser therapy. If an underlying invasive carcinoma is present, the treatment should be the same as for other invasive vulvar cancers, usually requiring a radical vulvectomy and bilateral inguinofemoral lymphadenectomy.

Materials and Methods

Women living in Serbia and Montenegro in their reproductive and non-reproductive period of life, were analyzed over a 5-year period. Vulvar and cervical screening were regularly performed. Vulvar microscopic examination was included into routine cervical examination by colposcopy.

Results and Discussion

At the Institute of Gynecology and Obstetrics over a period of five years, more than 180,000 women were examined. There were approximately 25,000 admissions per year. The number of gynecological and colposcopic examinations was in the range between 15,000 and 18,000 a year.

By analyzing the number and type of vulvectomies, we obtained significant data pointing to the necessity of introducing vulvoscopy into routine examinations. Over a period of five years, 580 simple and 178 radical vulvectomies were performed.

Table 1. — *Former International Federation of Gynecology and Obstetrics Clinical Staging for Vulvar Cancer (1969).*

Stage	Clinical Findings
Stage 0	Carcinoma in situ, e.g. VIN III, noninvasive Paget's disease.
Stage I	Tumor confined to the vulva up to 2 cm in diameter, and no suspicious groin nodes.
Stage II	Tumor confined to the vulva more than 2 cm in diameter, and no suspicious groin nodes.
Stage III	Tumor of any size with adjacent spread to the urethra or vagina (or both), the perineum, and the anus; or clinically suspicious lymph nodes in either groin, or a combination.
Stage IV	Tumor of any size infiltrating the bladder mucosa, or the rectal mucosa, or both, including the upper part of the urethral mucosa, or fixed to the bone, or other distant metastases, or a combination.

Table 2. — *International Federation of Gynecology and Obstetrics Staging of Vulvar Carcinoma (1989).*

Stage	Clinical Findings
Stage 0	Carcinoma in situ, intraepithelial carcinoma.
Stage I	Tumor confined to the vulva or perineum, or both (2 cm or less in greatest dimension); no nodal metastasis.
Stage II	Tumor confined to the vulva or perineum, or both (more than 2 cm in greatest dimension); no nodal metastasis.
Stage III	Tumor of any size with adjacent spread to the urethra or vagina, or both, or the anus, or unilateral regional lymph node metastasis, or a combination.
Stage IVa	Tumor invades any of the following: upper urethra, bladder mucosa, rectal mucosa, pelvic bone or bilateral regional node metastasis, or a combination.
Stage IVb	Any distant metastasis including pelvic lymph nodes.

By analyzing the age of 580 patients who underwent simple vulvectomies, we obtained very discriminating data. Over 70% of the women were younger than 40 years of age. In 45% of the cases, cervical loop diathermy was performed with CIN I (35%) and CIN II (25%) diagnoses; in 40% of the cases other diagnoses were established (condylomata accuminata, acid white epithelia, mosaicism, portio vaginalis uteri).

Within 14 to 18 months, patients who had undergone cervical loop diathermy presented with obvious problems in the vulvar region during routine examination. Since cultural habits for intimate hygiene within our population do not necessarily mean depilatory removal of hair in the region, vulvoscopy is not always adequate and the examination is performed only when problems are clearly evident. It should be mentioned that after surgery, during follow-up examination of the cervix, if subjective difficulties such as itching, burning, irritation or a reddish color of the vulva are detected, primary diagnosis of the pathology can be established. Out of all performed vulvectomies, 40% of the women had already been hospitalized and treated by surgery.

Histological findings in simple vulvectomies indicated intraepithelial neoplasias in 87% of the cases. In 13% of the cases, repeated surgery was performed and the process was spread to radical vulvectomy.

In 79% of the simple vulvectomy cases, histological examinations pointed out the existence of squamous cell carcinoma in situ (VIN III), while Paget's disease was present in 11% of the cases. In 10% of the cases, histological analyses pointed to vulvar Bowenoid papulosis.

Considering that 13% of repeated surgeries occurred in the group of women with simple vulvectomies, histological findings were analyzed after the surgery. Stage II of the disease was confirmed in all these patients.

The necessity of introducing ex tempore histological analysis during all gynecological procedures was confirmed in order to increase the healthcare standards of both patients and physicians.

Conclusion

Since almost half of the patients undergoing simple vulvectomies were not in the risk group, it is our opinion that vulvar examination in combination with colposcopy examination should be a part of routine screening of the population.

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