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Case Report

Cancer of the vulva in a 28-year-old woman



Janina Lipińska^{a,b,*}, Marek Stefanowicz^{a,b}, Tomasz Waśniewski^{a,b}, Piotr Martyn^b, Karolina Loewenau-Samusionek^b

^aClinic of Gynecology and Obstetrics, Gynecological Oncology and Gynecological Endocrinology, Faculty of Medical Sciences, University of Warmia and Mazury in Olsztyn, Poland ^bDepartment of Gynecology, Obstetrics and Gynecological Oncology, Provincial Specialist Hospital in Olsztyn, Poland

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ABSTRACT

Introduction: Prevalence of vulvar cancer in women under 30 years of age is very low. Human papillomavirus DNA is found in 20%–60% of women with invasive vulvar cancer and in 70%–80% of patients with vulvar neoplasia. In a young woman treated for recurrent, extensive vulvar condyloma acuminate, squamous cell carcinoma of the vulva was found. *Aim:* The aim of this work is to demonstrate that cancer of the vulva may occur in young women. It demonstrates the need for particular diligence in the diagnosis and treatment of vulvar diseases and building a good relationship with the patient in order to minimize the fear of invasive treatment and encourage compliance.

Case study: The patient, a 28-year-old female, after surgical treatment of condyloma acuminata, was admitted with a 3 cm tumor of the vulva. The tumor was excised with a margin of tissue free from infiltration – keratinizing squamous cell carcinoma was diagnosed. The patient did not return for extension surgery. After 5 years she was admitted with 4 cm loco-regional recurrence. Radical resection of the vulva with bilateral inguinal lymphadenectomy was performed. Four lymph node metastases were found. Adjuvant radiotherapy was implemented. Observation time was 4 years after the surgery for recurrence and 10 years after first diagnosis of cancer.

Results and discussion: Vulvar lesions cause anxiety in women, associated with sexual activity and the appearance of external genitalia after surgical treatment. Doubts of both the physician and the patient, particularly young, associated with invasive diagnostic methods result in prolonged ineffective conservative treatment. In the presented case wide local excision of the tumor proved ineffective. Local recurrence metastasized to lymph nodes bilaterally and contralaterally.

Conclusions: Treatment results of vulvar cancer are frequently dependent on patients' willingness to comply and minimizing the fear of consequences of the treatment. Women with oncological conditions of the vulva should be diagnosed and treated in specialized centers.

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^{*}Correspondence to: Clinic of Gynecology and Obstetrics, Gynecological Oncology and Gynecological Endocrinology, Faculty of Medical Sciences, University of Warmia and Mazury, Department of Gynecological Oncology, Provincial Specialist Hospital in Olsztyn, Żołnierska 18, 10-516 Olsztyn, Poland. Tel.: +48 89 538 64 83.

E-mail address: lipinskajoan@yahoo.com (J. Lipińska).

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1. Introduction

Vulvar cancer is the fourth most common type of gynecologic cancer. It predominantly affects older women. The highest incidence rates are observed in patients aged 70 or older.¹ Prevalence in women under 30 years of age is very low.¹³

In accordance with data obtained from the studies conducted on the population of U.S. women, in common with increasing number of new cases of vulvar cancer, a more than fourfold increase in vulvar intraepithelial neoplasia (VIN) was noted.⁸ According to several authors human papillomavirus DNA is found in 20%–60% of women with invasive vulvar cancer and in 70%–80% of patients with VIN. HPV infections are considered sexually transmitted diseases that typically affect women under 45 years of age, smokers and with early initiation of sexual activity.¹⁴ A subclinical infection may be diagnosed by vulvoscopy. Types of viruses of high oncogenic potential, e.g. HPV 16, 18, 31, 33, 66, may contribute to the development of cervical cancer, cancer of the vulva, vagina, anus, penis and larynx.¹²

The first results of a vaccine against HPV antigens, proving high effectiveness in the prevention or reduction of HPV infections, were published in 2002. Published in 2005, the results of a prospective, randomized, double-blind clinical trial FUTURE II confirmed overall effectiveness of quadrivalent HPV vaccine in the prevention of benign and malignant lesions.^{10,15}

2. Aim

The aim of this work was to demonstrate that cancer of the vulva may also occur in young women and that particular diligence in the diagnosis and treatment of female genital lesions, also in very young subjects, is essential. In addition, the role of building a good relationship with the patient is emphasized, that may result in willingness to comply, following recommendations and minimizing the fear of invasive treatment.

3. Case study

The patient, a 28-year-old female, was admitted to the Department of Gynecologic Oncology with a vulvar tumor measuring 3 cm in diameter, located in half of the length of the right labia majora. Several years before, after numerous attempts of conservative treatment, the patient underwent extirpation by electrocautery of condyloma acuminatum. At that time, the histopathological diagnosis was condyloma accuminatum, koilocytosis without nuclear atypia, which suggested viral etiology of the disease. Following the admission to the Department of Gynecologic Oncology, the tumor was totally excised with a margin of surrounding tissue free from macroscopic infiltration. Pathological report confirmed squamous cell carcinoma G2, size of $2.8 \times 2.0 \times 0.8$ cm, excised with margins of 4×8 mm and 10 mm deep. The patient was provided with detailed information on the treatment options in such cases, but she definitely refused to consent for extended resection. She was discharged with a recommendation of further regular follow-up in Gynecologic Oncology Outpatient Clinic. Within a year after the surgery no lesions on the vulva or inguinal lymph nodes were observed. After this period of time the patient did not return for follow-up, despite definite recommendations.

After 5 years the patient was admitted to the Department of Gynecologic Oncology again, with vulvar lesion in the form of malignant tumor, 4 cm in diameter, at the site of the postoperative scar. Radical surgical resection of the vulva with bilateral superficial and deep inguinal lympadenectomy was performed. Pathological report confirmed keratinizing squamous cell carcinoma G2. Cancerous lesion sized $3.7 \times 2.3 \times$ 1.1 cm was excised with a margin of 0.50×1.20 cm. Twentyfive lymph nodes were resected, including 13 ipsilateral and 12 contralateral. Metastases were found in 3 ipsilateral superficial inguinal lymph nodes and in 1 contralateral deep inguinal lymph node. The patient was classified as stage III B according to FIGO 1988. Post-operative wounds healed by primary intention. Patient was discharged home after 6 days. After 3 months she underwent adjuvant radiotherapy and received a planned dose of 50 Gy to the area of the vulva and inguinal and iliac lymph nodes, X₆ 16 MeV; E 18 MeV. A three degree radiation-induced skin reaction was a complication of treatment. Recurrent thrombophlebitis was also observed. Therefore, anticoagulant treatment with Acenocumarole was induced. Currently, the patient remains under the care of Oncology Outpatient Clinic, which she attends regularly.

4. Results and discussion

Incidence of pre-cancerous vulvar lesions is unknown. There is no obligation for its registration. Thus, it is difficult to follow the natural course of malignant transformation.

Lesions developing on the vulva cause high anxiety in women, associated with sexual activity and the appearance of external genitalia after the treatment, particularly surgical. A good cooperation with the patient, meticulous diagnostic and treatment process and elimination of fear associated with the disease are therefore necessary. Doubts of both the physician and the patient, associated with invasive diagnostic methods that allow histopathological diagnosis, result in prolonged ineffective conservative treatment. It concerns particularly young females.

The classic and still applicable method of surgical treatment of vulvar cancer is radical resection of the vulva with superficial and deep inguinal lymphadenectomy. According to numerous works the most important prognostic factors in squamous cell carcinoma of the vulva are tumor size, depth of stromal invasion and presence of lymph node metastases.

Recurrence rates in the literature cover a wide range of values. In a multicentre study conducted in Italy, after the treatment of 502 patients, recurrence was reported in 37.3% of cases, including: 53.4% of local recurrences, 18.7% of nodal recurrences, 5.7% of pelvic lymph nodes recurrences.¹¹ The analysis of the treatment results in Centre of Oncology in Warsaw demonstrated that recurrence occurred in 33.3% of patients and in 75.0% recurrence occurred in the first year of follow-up.³ Time of local recurrence after surgical treatment for advanced stages of the disease was 5–8 months.⁷ Recently,

approach to surgical treatment of vulvar cancer has changed – from radical vulvectomy in each stage of the disease to the less radical methods of intervention, such as wide local excision of the vulva around the primary lesion. Resection of inguinal lymph nodes from one side, superficial lymph nodes or sentinel node labeling were attempted.^{2,7} In the presented case local excision of the tumor approximately 3 cm in size proved ineffective. Local recurrence, treatment of which was delayed by the patient until it grew almost to 4 cm, gave bilateral and additionally contralateral lymph node metastases. Metastases were found in three out of eight ipsilateral resected inguinal lymph nodes and in one out of four contralateral femoral lymph nodes. Eight contralateral superficial inguinal lymph nodes were also removed, in which metastases were not found.

There are documented cases of contralateral lymph node metastases in lesions below 2 cm (1.2% of cases) and primary site more than 2 cm in diameter (1.8%).^{4,6} Cases of contralateral inguinal lymph node recurrence after surgical treatment with unilateral lymphadenectomy have also been reported.^{5,9}

Pathological lesions on the vulva, lymph nodes or internal organs are not observed either in clinical examination, or in diagnostic imaging. Total observation time after radical surgery and adjuvant radiotherapy of recurrent vulvar cancer is 4 years and 10 years after first diagnosis of cancer.

5. Conclusions

Treatment results of vulvar cancer are highly dependent on patients willingness to comply, including subjects at younger age, and minimizing the fear of the method and consequences of treatment.

Women with oncological conditions of the vulva should be diagnosed and treated in specialized oncology centers.

Wide use of HPV vaccine would create an opportunity to significantly reduce development of persistent HPV viral infection and, in consequence, vulvar and cervical intraepithelial neoplasia and vulvar cancer.

Conflict of interest

None declared.

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