



Case Report

A rare case of vulvar squamous cell carcinoma; case presentation

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Abstract

Objectives. Vulvar cancer is a rare gynecological malignancy, with an incidence of 1.5 per 100 000 women/year. The most common vulvar cancer is developed in squamous cells, the most encountered type of skin cells.

Case report. We report a case of a 72-year-old female admitted in the Department of Plastic Surgery of Emergency Clinical Hospital “Prof. Dr. Agrippa Ionescu” with a 5/4.2 cm painful ulcerated tumoral mass located in the vulvar area. The lesion slowly increased in size over the past 12 months. The tumour was surgically removed with oncological safety margins and sent for histopathological evaluation. The histopathological examination revealed an ulcerated squamous carcinoma with lymphovascular and perineural invasion, but with negative margins. Postoperative results were favorable, and no local or general complications were observed.

Conclusion. We highlight this case due to its unusual presentation in the clitoral area. Moreover, considering the potential for recurrence we point out the importance of the radical vulvectomy with regional lymphadenectomy and histopathological examination, in order to put a precise diagnosis and ensure the best possible treatment for the patient.

Keywords : vulvar carcinoma, squamous cell carcinoma, radical vulvectomy

Highlights

- ✓ Vulvar cancer is a rare and aggressive tumour, lowering the quality of life of affected women by deforming the perineal area (with physiological and aesthetic implications).
- ✓ Prevention/ early detection makes possible vulvar cancer detection in the first stages decreasing the morbidity and mortality.

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Introduction

Vulvar cancer is considered to be the fourth gynecologic malignancy, representing 5% of all tumours of the female genital tract. 95% of them are squamous cell carcinomas (SCC), the rest 5% being melanomas, sarcomas and basal cell carcinomas (1). The annual incidence is 1.5 per 100.000 women/ year, increasing constantly with age, the average age at diagnosis being 75 years old (2). Generally, the prognosis is good if it is diagnosed and treated in early stages, the cornerstone being represented by a radical local wide surgical excision (1).

There are two types of vulvar squamous cell carcinoma. The first one affects younger women and it is associated with high-risk types of Human Papillomaviruses (HPV), most of the time HPV genotypes 16, 18 and 33 (3). It is also connected to warty and/or basaloid vulvar intraepithelial neoplasia, representing 30% of the vulvar squamous cell carcinoma. The second type and the most frequent affects elderly women and it is not linked to any type of Human Papillomaviruses. In general, this type of squamous cell carcinoma is associated with lichen sclerosus that is characterized with pruritus and in time scratching leads to hyperplasia of squamous cells and further to atypia and vulvar intraepithelial neoplasia (4). This vulvar epithelial neoplasia, if it is not discovered, might evolve to an invasive cancer (1).

The clinical examination displays pruritus, the most common symptom for vulvar cancers, vulvar bleeding, dysuria and discomfort. Local exam shows a lesion that can be described as a fleshy or warty tumour mass, with possible ulcerations (4).

Vulvar squamous cell carcinoma, in first stages has a horizontal growth with invasion in adjacent organs like vagina, urethra or anus followed by lymphatic spreading to regional lymph nodes (inguinal, femoral and pelvic). In advanced cases, distant metastases might be also present in liver, lungs or bones (5). Prognostic factors are considered the size of the lesion, the depth of the invasion and the disease stage (1).

Considering the histological examination vulvar squamous cell carcinoma can be classified in three types: basaloid, warty and keratinizing. The keratinizing is the most common type and appears after menopause despite the other types that appear before menopause (3).

Primary treatment consists in wide surgical excision associated if necessary with regional lymphadenectomy. Radiotherapy and chemotherapy are also recommended in particular cases (1).

Case report

A 72-year-old woman, fair skin type, was referred by the gynecologist to our plastic surgery department with a tumour mass situated on the vulvar region. The tumour appeared one year before and increased in size, being also associated with local discomfort, pain and dysuria. Local examination showed a warty tumour mass of 6/5 cm that involved almost all the anatomical parts of the vulva including labia majora and minora, clitoris, vestibule, vaginal introitus, and the urethral meatus (Figure 1). The clinical evaluation of regional adenopathy was positive with palpation of bilateral inguinal enlarged lymph nodes.



Figure 1. Preoperative aspect of the tumour

The patient had no other medical history. General clinical examination found a normal weight patient, in a good general condition without any particular findings. Complete blood test evaluation showed slightly elevated leucocytes and urinalysis revealed leucocytes and erythrocytes in urine. Further tests showed also the presence of a urinary infection. According to antibiogram, antibiotic treatment was started and a biopsy of the tumour was realized.

In order to evaluate the extension of the tumour a computed tomography was performed. The CT scan highlighted the tumour mass measuring 58,5/25 mm, with irregular borders, located on the anterior part of the vulvar region with inguinal metastatic lymph nodes with the biggest diameter of 18,5 mm on the right part and 24,5 mm on the left side.

Histopathological examination of the tissue fragments, following incisional biopsy was suggestive for a squamous cell carcinoma.

Considering the size and the tumor's aggressivity, surgical treatment of the tumour was performed under spinal anesthesia. Radical vulvectomy was realized with complete removal of the vulva to the level of pubis periosteum, thigh deep fascia and inferior sheath of the urogenital diaphragm. The excision was performed with a 1.5 cm safe margin in order to minimize local recurrence. Excision of 2 cm of the urethra was also realized being forced to reconstruct it using oral mucosa (Figure 2). Reconstruction of the vulvar region was completed using local fasciocutaneous advancement flaps continuously with reconstruction of the distal part of urethral meatus (Figure 3).

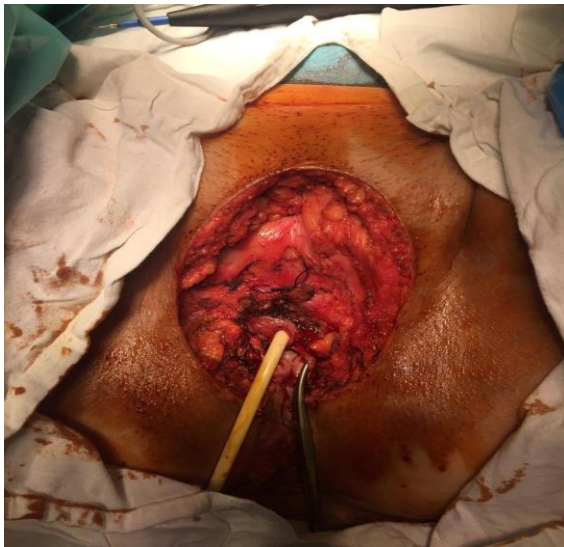


Figure 2. Intraoperative aspect after resection of the tumour



Figure 3. Postoperative aspect first day after surgery

Considering the presence of palpable bilateral inguinal lymph nodes, a full bilateral inguinofemoral lymphadenectomy was realized through separate

incisions in the inguinal area. The patient made a good postoperative recovery. No local complications like edema, hematoma or infection were encountered.

Histopathological examination showed keratin pearls, apoptotic and dyskeratotic cells with atypical mitosis that involved the dermis and epidermis (Figure 4). The diagnosis revealed an ulcerated squamous cell carcinoma well differentiated (G₁) measuring 5/4,2 cm, with a thickness of 22 mm, located in the clitoral area with extension to the urethral meatus. Lymphovascular and perineural invasion were found. All margins were free of tumour. From all the inguinal lymph nodes that were dissected 4 of them presented metastasis of squamous cell carcinoma, 3 nodes from the left side and 1 from the right part. According to TNM stabilization our patient had SCCs stage IIIB (T₂N_{2b}M₀).

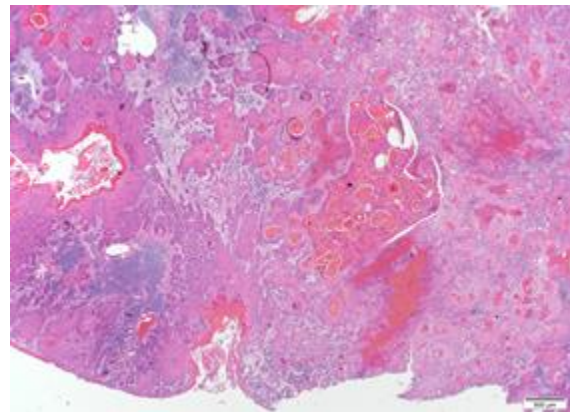


Figure 4. Mucosal invasion of the squamous cell carcinoma with good differentiation (G₁). HE staining

Despite the oncologist recommendation our patient refused adjuvant radiotherapy. Follow up at three and six months showed no local recurrence or local discomfort.

Discussion

Vulvar cancer is a rare disease that affects especially elderly women. The prognosis is favorable when is diagnosed in an early stage with a five-year survival rate of 80 to 90% (6).

There are some characteristics of SCCs that influence the prognosis such as differentiation grades, location, tumour size and depth and perineural invasion. Our patient presented SCCs in first stage of differentiation (G₁) that has a good prognosis compared with stages 3 and 4 that recur and metastasize faster (6). In general, the SCCs located in the genital region are considered high risk and those in the vulvar area require special attention of the treatment due to the aggressiveness (7).

The size of the lesion is also important in risk assessment. SCCs larger than 2 cm are more likely to recur and metastasize than those with a diameter of less than 2 cm. The size of the primary tumour is not the most important factor in defining prognosis, but the thickness. The thickness of the tumour is considered one of the most important factors, suggesting that a depth bigger than 6 mm have a higher risk for lymph node involvement and a rate of metastasis of 15% (8). In the case above, the histopathological examination showed a thickness of 22 mm, with invasion of the regional lymph nodes. Risk factors for lymph node metastasis include also age, degree of differentiation, tumour stage, presence of capillary-lymphatic space invasion. Almost 30% of patients with operable disease have lymph nodal extension (8).

Perineural invasion, known as neurotropism, is described as tumour cells in the perineural space (9). It is a poor prognostic factor (7) that was also present in our patient in addition with lymphovascular invasion and indicates the necessity of adjuvant therapy.

The staging of this type of skin cancer can be realized using American Joint Committee on Cancer classification (AJCC with TNM) or the International Federation of Gynecology and Obstetrics classification (FIGO). The information required are the dimension and extent of the tumour, node status and presence of distant metastasis. The case that we reported presented a tumour less of 5 cm with invasion in the vulva and lower urethra (T₂), spread in 4 regional lymph nodes (N_{2b}) with no distant metastasis that was classified as stage III_b according to AJCC and FIGO classifications (1, 10).

In this stage the recommended treatment is wide local excision with regional lymphadenectomy associated with adjuvant radiotherapy (8). In our case radical vulvectomy was necessary considering the tumour size and invasion. This was also associated with separate incision for realizing inguinal lymphadenectomy.

Adjuvant radiotherapy is recommended in high risk cases with tumour size over 4 cm, lymphovascular invasion, lymph node invasion and close or positive surgical margins (11). Radiotherapy of the inguinal region is also indicated in patients that have only an invaded lymph node (1). Our patient refused any adjuvant therapy, being more predisposed to local recurrences.

Prognosis is relative good in early stages, the most important factor, like in breast cancer, being the presence of regional lymph nodes metastasis (12). The 5-year survival rate is 70-90% for patients with negative nodes compared to 25-41 % for those with lymph nodes metastasis. Other negative prognostic factors are

perineural invasion, stage and older age. Considering FIGO classification, our patient has a 1-year survival rate of 74% and a 5-year survival rate of 43% that is probably reduced due to radiotherapy rejection. Recurrences are more difficult to treat and can lead to distant metastasis (13, 14).

Conclusions

Vulvar cancer is a rare and aggressive tumour having a disabling effect on women, lowering their quality of life by deforming the perineal area that can affect their relationships, body image and confidence. Prevention followed by early detection and histological examination of any suspect vulvar lesions makes possible vulvar cancer detection in the first stages decreasing the morbidity and mortality.

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