

PATIENT WITH MALIGNANT VULVAR NEOPLASM: CASE REPORT

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ABSTRACT

Background: Vulvar carcinoma most often occurs on the outer surface area of the female genitalia. The vulva is the area of skin that surrounds the urethra and vagina, including the clitoris and labia. It is quite rare cancer of the female reproductive system and accounts for 3-4% of all genitourinary tract neoplasms. Though it can occur at any age, vulvar cancer is most common in older adults. When present in young women it is mostly associated with human papillomavirus (HPV)-related dysplasia. The most common histology is squamous cell carcinoma of the vulva.

Case study: This report presents a patient with verified planocellular vulvar carcinoma who underwent radical hysterectomy with adnexectomy and lymphadenectomy due to cervical neoplasm (squamous cell carcinoma). After chemo-radio therapy, the patient underwent radical vulvectomy and two months later, resection of the distal urethra and vulvar and vaginal reconstruction with gracilis muscle. Full-thickness skin graft was used to reconstruct the distal part of the urethra.

Conclusion: Combination of flaps and full-thickness skin graft can be used in reconstruction of vulva and urethra.

Keywords:

Malignant vulvar neoplasm, planocellular carcinoma, reconstructive techniques, vulvectomy

INTRODUCTION

Vulvar cancer is rare cancer of the female reproductive system and accounts for only 3-4% of all genitourinary tract neoplasms. The peak of its incidence is in the 7th and 8th decade of life. In most cases (90%) it is planocellular carcinoma, second most common is basocellular carcinoma (4%), while the rest (6%), are cases of rare carcinoma types, such as melanoma, Paget's disease, Bartholin gland carcinoma [1-2].

There are at least two types of planocellular vulvar carcinoma. The first type is usually a result of VIN (vulvar intraepithelial neoplasia) progression, most commonly occurs in younger women and is associated to HPV infection, promiscuity, and condyloma. Vulvar intraepithelial neoplasia is premalignant dysplasia of vulvar squamous epithelium which differs from carcinoma because the epithelial basement membrane

is intact. Depending on a height of epithelium it is divided into three stages. In the first stage, VIN 1, only one-third of the thickness of the surface layer of the vulva is affected; in the second stage, VIN 2, two-thirds are affected, while in the third stage, VIN 3 (carcinoma in situ) the full thickness of the surface layer of the vulva is affected [3-5].

The second type most commonly occurs in obese, diabetic, and women with hypertension. Besides already mentioned, risk factors are also considered to be smoking, lichen sclerosus, squamous hyperplasia, chronic granulomatous diseases, and squamous vaginal or cervical carcinoma. The labia majora and clitoris are the most common sites. Vulvar cancer usually presents with palpable, slow-growing mass, and patients visit their doctors because of itching, pain, bleeding, vaginal discharge, or burning sensation during urination [1-3].

Vulvar cancer can spread locally (ex. urethra, urinary bladder, vagina, anus or rectum), hematogenous, in inguinal lymph nodes or from inguinal lymph nodes to pelvic and paraaortic lymph nodes [5,6].

Treatment consists of radical wide excision and removal of inguinal lymph nodes (in patients with early stage disease) while chemoradiation is an alternative to radical vulvectomy with en bloc inguino-femoral lymphadenectomy for advanced disease [5,6].

CASE STUDY

A 64-year-old patient was admitted to the Department of Plastic, Reconstructive, and Aesthetic Surgery at the Clinical Hospital Center Zagreb for the excision of the malignant vulvar neoplasm.

In April 2011, the patient underwent radical hysterectomy with adnexectomy and lymphadenectomy in Department of Gynaecology and Obstetrics at Clinical Hospital Center Zagreb due to cervical neoplasm (squamous cell carcinoma). Radiotherapy (administered for 29 consecutive days), brachytherapy (50 hours), and concomitant chemotherapy (cisplatin 40 mg/m², once a week, 4 consecutive weeks) followed.

In May 2012, the patient underwent radical vulvectomy after pathohistological analysis confirmed planocellular carcinoma with negative margins. Two months later, resection of the distal urethra and vulvar and vaginal

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reconstruction with gracilis muscle were performed. Full-thickness skin graft (Wolf-Krausse) was taken from the left thigh and was used to reconstruct the distal part of the urethra. This is somewhat more risky procedure than using split-thickness graft because of the much higher percentage of graft failure and consequent graft contraction. The area from which the graft was taken heals faster and less painful than in case of split-thickness graft. Very appreciated characteristic of full-thickness graft is also a lower incidence of secondary contraction, which enables great reconstructive results. In this case, the full-thickness graft was taken from the left thigh, and distal urethral part was reconstructed. During the surgical procedure, the patient was under general anesthesia, a fusiform skin graft was taken from thigh using inguinal crease as long axis. Perineal skin defect was covered using Thiersch split-thickness skin graft. In May 2018 the patient had discovered lesion and experienced difficulty with urination. She was referred to the department of gynecology and obstetrics, University Hospital Centre Zagreb. Gynecological examination revealed cystic formation, located left paraurethral, movable size 4 cm. The biopsy was performed. Cytological as well as microbiological analysis was done. Preoperative abdominal ultrasound revealed hypoechoic zone with posterior accumulation caudal from urinary bladder towards rectum, without Doppler signal (used for vascularity assessment), possible cystic mass/fluid retention in postoperative scar tissue. Removal of the superficial structures revealed tumorous cystic formation (figure 1).

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According to the results, differential diagnoses included malignant neoplasms of the vulva. Surgical removal, excision (without lymphadenectomy), was performed (figure 2). Pathological and cytological analysis, were made. A smear test was positive for malignant cells.

Postoperative pathology reported lots of abnormal squamous cells, less specialized with degenerative changes and necrotic cellular debris. According to the clinical cytologist the lesion was planocellular carcinoma. Pathohistological examination of tumor-associated material showed abnormal tissue size 7x1.5x1.3 cm, constructed of a poor cellular connective tissue with cystic formation, partially thickened with atypical moderately differentiated epithelium. Described areas were also found in the stromal connective tissue. Skeletal muscle tissue was described marginally as well.

Postoperative course was uneventful. The patient was discharged from the hospital in good condition, on the 7th postoperative day.

DISCUSSION

Female urethral reconstruction is complex, and one must carefully evaluate patients. There are some differences from male urethral reconstruction because of the different urethral length. The female urethra is shorter than the male urethra, leading to a higher

risk of incontinence. Reconstructive techniques can be categorized as anastomotic, flap-based or graft-based. The surgical approach for these techniques of repair can be described by its relative position to the urethra; dorsal (that is 12 o'clock), ventral (that is 6 o'clock) and circumferential. Ventral (supravaginal) and dorsal accesses have been the most frequent ones, although the small number of patients have undergone "tubularized urethroplasty"[5-7].

The advantage of using a ventral approach is the minimal mobilization of the urethra, however, this access lead to the higher risk of developing a urethrovaginal fistula.

Potential benefits of the dorsal approach would include the avoidance of a vaginal incision and its associated post-operative complications, including issues with urethrovaginal fistula and wound complications. The dorsal access provides some more positive facts such as good mechanical support and well-vascularized base for receiving the transplant. However, surgeons confront some disadvantages using a dorsal approach as well. The injury of the sphincter mechanism causing incontinence is possible, as well as sexual dysfunction caused by a neurosensory disorder [5-7].

Female urethral reconstruction can be performed with lingual flap and buccal mucosal graft urethroplasty, but the highest success rates have been reported to treat a heterogeneous group of urethral disorders, such as urethral stricture and urethral loss. Urethral replacement can be accomplished by in situ tubularized full thickness buccal mucosal graft based on the clitoris creating the neourethral tube. Neourethra is then covered with the gluteus tissue, or the periurethral tissue can be used as well. The disadvantage of the buccal mucosal graft is a low, but significant risk of the salivary glands injuries [6-9].

The disadvantage of the full-thickness graft is a higher risk of the transplant rejection. Also, the healing process is slower due to full-thickness skin graft.

Several more reconstructive techniques have been described. These have included vaginal or labial flaps. Flaps are the most common and the earliest published reconstructive technique utilized for female urethral stricture. Flaps are particularly advantageous because of their mobility and good vascularity, meaning they can be raised with relative ease. In this case, these techniques were not possible due to radical vulvectomy [7-9].

CONCLUSIONS

The prognosis for vulvar carcinoma is fairly poor due in part to their rare nature and a relative lack of evidence-based management strategies, but the most important is early diagnosis. Early diagnosis can improve prognosis in vulvar carcinoma or other aggressive tumors and, in the case of recurrences, may lead to changes in therapy. Early diagnosis followed by prompt surgical operation remains the best treatment for patients at present.

CONFLICT OF INTEREST:

The authors declare that there is no conflict of interest.

The patient gave her informed consent prior to her inclusion in case report.

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FIGURE CAPTIONS

Figure 1: Cystic tumorous mass found during procedure



Figure 2: postoperative status (after cystic mass removal)

