

Case 1 – Vulvectomy plus posterior exenteration with plastic reconstruction in advanced cancer – a case report

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Abstract

This case report describes a 47-year-old patient with advanced vulvar cancer diagnosed in May 2014. Treatment included a vulvectomy, inguino-femoral and pelvic lymphadenectomy combined with posterior exenteration and plastic reconstruction. The choice between radical surgery and radio-chemo irradiation, both of which are valid therapeutic options in locally-advanced vulvar cancer, is discussed.

Key words: posterior exenteration, squamous cell carcinoma, vulvar cancer, vulvectomy

Introduction

Invasive squamous cell carcinoma of the vulva is primarily a disease of postmenopausal women [1]. Squamous cell carcinomas account for about 90% of all primary vulvar malignancies, most squamous carcinomas of the vulva occur at the labia majora and minora (60%) [2]. The incidence of vulvar cancer is 2.2 cases per 100,000 women. The National Cancer Institute has reported that vulvar cancer is one of the twelve malignant tumors that have increased in incidence over recent years [3]. All patients with tumors that have >1 mm of stromal invasion require inguino-femoral lymphadenectomy [4]. Stromal invasion of >2 mm and extracapsular lymph node spread are the most important predictive factors for local and regional recurrence [5]. Independent risk factors for recurrence include the size of the tumor, lympho-vascular space involvement, multifocality of the tumor, status of surgical margin, and the presence of a concurrent vulvar intraepi-

thelial neoplasia [6]. Overall, about 30% of patients with operable disease have lymph nodal spread [7].

Until the 1980s the standard therapeutic approach for invasive locoregional vulvar carcinoma was radical surgery, including complete resection of the vulva and regional lymph nodes using the butterfly technique. In tumors clinically confined to the vulva or perineum, radical local excision with a margin of at least 1 cm has replaced radical vulvectomy and separate incision has replaced en-bloc inguinal node dissection [8]. Pelvic exenteration (PE) is a therapeutic option in advanced primary or relapsed vulvar cancer, offering median- to long-term survival for many patients. Carcinomatous spread to regional lymph nodes and complete resection are the most important prognostic factors [9]. PE remains a therapeutic option with an acceptable rate of complications and postoperative mortality [10]. Patients with lymph node-positive vulvar cancer are at high risk of disease recurrence. A recently published large multicentre study observed that adjuvant radiotherapy was associated with improved prognosis in lymph node-positive patients. However, outcome after adjuvant radiotherapy still remains poor compared with lymph node-negative patients [11]. Radiation therapy can be used for patients who are unable to tolerate surgery because of extended disease or comorbidities [12].

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Clinical course and treatment

In May 2014, a 47-year-old woman was referred to our department for treatment of locally advanced vulvar cancer. Rectoscopy and cystoscopy did not reveal any evidence



Fig. 1. Vulvar cancer.

of tumor involvement or other pathological conditions of the rectum or urinary bladder. Gynaecological examination showed an ulcerated vulvar cancer on the right labium major, extending to the anus and passing over to the contralateral side (Figure 1). Recto-vaginal examination showed that the tumor infiltrated the lower third of the vagina. Also there was clear evidence of infiltration into the anal-sphincter and the septum rectovaginale. Histological examination of a biopsy specimen confirmed an invasive large-cell, non-keratinizing, squamous cell carcinoma of the vulva.

The patient was discussed at a meeting of the institutional tumor board. It was considered that neoadjuvant radio-chemotherapy could risk loss of anal sphincter function, and the patient was reluctant to undergo radiotherapy. Therefore, the board's final decision was to that vulvectomy with a posterior exenteration was best treatment option for the patient. The inter-disciplinary operation started with the inguino-femoral lymphadenectomy (Figure 2),

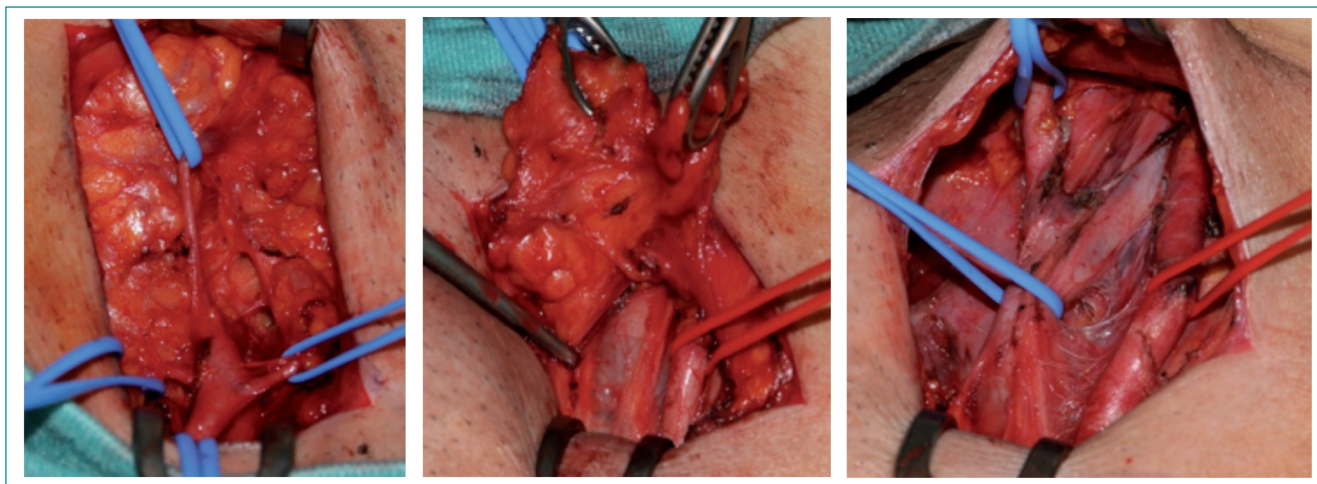


Fig. 2. Inguino-femoral lymphadenectomy.

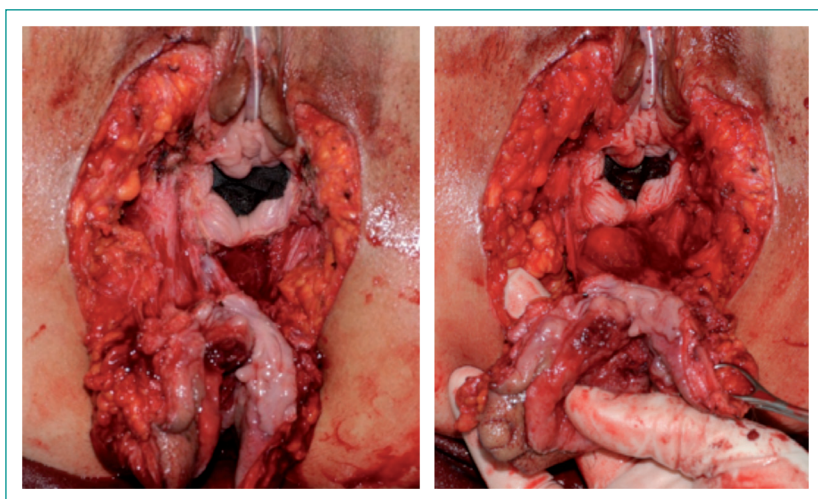


Fig. 3. Vulvectomy.

followed by the vulvectomy (Figure 3). Then posterior exenteration, including an abdomino-perineal extirpation of the rectum with implantation of a permanent sigmoideostomy, was performed. The next part was pelvic lymphadenectomy. A total of 35 lymph nodes (LN) were removed: 8 right inguinal LN, 7 left inguinal LN, 5 LN from the right iliaca externa arteria, 7 LN from the left iliaca externa arteria, 1 LN from the right obturatoric region, 1 LN from the left obturatoric region and 6 LN from the perirectal region. The final part of the operation was performed by a plastic surgeon. He used a dorsal-pedicu-



Fig. 4. Plastic surgery reconstruction with transposition-flap from both upper legs.

lated transposition-flap from both upper legs (Figure 4) for reconstruction of the defect. The final histological result showed a low differentiated squamous cell carcinoma of the vulva, Grade 3. TNM classification was pT2 (tumor size: 8 × 5 cm) N0 (0/35 LN) L1 V0 Pn0. The margins of resection were free of tumor cells on all sides. The minimal distance from the tumor to the margin of resection

was 0.5 cm. Therefore, according to current guidelines, no adjuvant local radiotherapy or chemotherapy was necessary. The patient recovered and was scheduled for 3-monthly follow-up visits.

The first follow-up was in July 2014. Primary wound healing was satisfactory. There was a lymphedema in the left inguinal region from which 35 ml of clear, serous liquid was aspirated. As a consequence of the surgery the patient had a voiding dysfunction and needed to use a urinary catheter. The next follow-up was in November 2014. Primary wound healing showed a strong tendency to keloid scar formation. Measurement of nerve conduction velocity still showed some voiding dysfunction but this had improved since the last follow-up.

Conclusion

Complications such as wound healing problems, lymphedema and functional deficiencies, resulted in a trend went towards more limited surgical approaches for vulvar cancer, often combined with radiation therapy [13]. Given the young age of the patient in this case, radical surgical treatment was considered the best choice because it could provide the best cure rate. It has been proven that tumor diameter and tumor-free margins of the resected tissue are the most important prognostic factors. In our opinion, surgery in this patient with locally advanced extension of vulvar cancer was associated with good recovery and quality of life. Even in difficult situations, vulvectomy with posterior exenteration followed by plastic reconstruction can have beneficial outcomes for patients [14].

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Commentary

In the case of a locally advanced vulvar cancer, when the urethra and/or anus-rectum are involved and/or the tumor is large, primary surgical management can be feasible even if it often requires anterior and/or posterior pelvic exenteration, complex reconstructive surgery, and urinary or intestinal diversion [1].

In these cases, primary radiotherapy (combined with concurrent chemotherapy) has been more frequently offered as an alternative to upfront surgery to decrease morbidity and disfiguration (increasing trend from 18% in 1988 to 30% by 2008) [2]. Neoadjuvant chemoradiation also seems to be beneficial in these patients: several observational studies, two phase II trials by the Gynecologic Oncology Group (GOG) and a Cochrane review have described the outcomes of primary chemoradiation followed by surgical resection, showing a decrease of tumor burden, often allowing less radical resection [3-5]. Moderate (desquamation requiring treatment interruptions, deep venous thrombosis) and severe (rectovaginal fistula, bowel perforation/obstruction, avascular hip necrosis, toxic deaths) complications have been reported, and wound breakdown can occur in up to 20-31% of patients who undergo surgery after chemoradiation [6-8].

Pelvic exenteration, however, remains a reliable treatment option for locally advanced vulvar cancer in “young” patients without extra-inguinal node or distant metastases and with resectable disease allowing adequate surgical margins. The mortality for this procedure is sufficiently low, and the rate of complications could be acceptable. Plastic reconstructive procedures play an important role in this surgical approach [9, 10].

The Cochrane review confirmed no differences in survival or recurrence rates between primary radiotherapy and surgery, with patient age being the only significant prognostic factor [5].

I read with interest the case described by Schmid and colleagues and two further considerations can be made: a) the upfront exenterative surgery was a feasible, if not the most preferred, option (“young” age, no extra-inguinal node and distant metastases, resectable disease); and b) given that the lack of extra-inguinal node involvement is a prerequisite for exenterative surgery, a FDG-PET could be included in the preoperative workup.

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