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A RARE VULVAR MULTIFOCAL IN-SITU SQUAMOUS CELL CARCINOMA: CASE REPORT

ENDER GÖRÜLEN VULVAR MULTİFOKAL İN-SİTU SKUAMÖZ HÜCRELİ KARSİNOMA: OLGU SUNUMU

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ABSTRACT

Introduction: Vulvar squamous cell carcinoma (VSCC) is the fourth most common type of gynecological cancer, though it is almost very rare in younger group.

Material and Methods: In this case report we would like to present a twenty-two years old, non-virgin, a white female with multifocal (approximately more than a hundred) epidermal VSCC at mostly from labia majora through to anal area.

Results: All skin lesions including epidermis, dermis, hypodermis and superficial

fascia were excised at spinal-epidural combined anesthesia conditions. Healing period was ended uneventfully. In permanent pathological evaluation multifocal in-situ squamous cell carcinoma observed and secure skin margins were obtained. Smear examination showed that our patient had HPV 16 positive

Conclusions: VSCC often related with HPV positivity. Therefore generalization of vaccination programs had an important role at preventive treatment. The other genital structures must be evaluated in which having HPV positivity. We recommend that in case determination of any suspicious or newly developed skin lesions at female external genitalia must be referred to related surgery department for excisional biopsy in order to early diagnose of a cancer beginning.

Keywords: Vulvar squamous cell carcinoma, vulvar lesion, female external genitalia

ÖZET

Giriş: Vulvar skuamöz hücreli karsinom (VSHK) 4. sıklıkta görülen jinekolojik kanser olmasına rağmen genç yaş gurubu kadınlarda oldukça ender görülür.

Gereç ve Yöntem: Bu olgu sunumunda 22 yaşında, beyaz bir kadında, çoğunluğu labia majorada yer alan ve anal çıkıma doğru yayılan multifokal epidermal VSHK olgusu anlatılmıştır.

Sonuçlar: Tüm cilt lezyonları epidermis, dermis, hipodermis ve süperfisiyal fasyayı da içerecek şekilde spinal-epidural kombine anestezi şartlarında eksize edildi. İyileşme döneminde herhangi bir sorunla karşılaşılmadı. Patolojik inceleme sonucunda multifokal in-situ skuamöz hücreli karsinom tanısı kesinleştirildi. Cerrahi sınırlarda herhangi bir patolojik invazyon tespit edilemedi. Smear değerlendirmesinde hastamızda HPV 16'nın pozitif olduğu görüldü.

Tartışma: HPV pozitifliği sıklıkla VSHK'a eşlik etmektedir. Bu yüzden aşılama programlarının yaygınlaştırılması önleyici tedavide önemli bir yere sahiptir. HPV pozitif hastalarda diğer genital yapıların da dikkatli şekilde değerlendirilmesi gerekmektedir. Önerimiz kadın dış genitalyasında saptanan şüpheli, yeni ortaya çıkmış ya da şüphe yaratmayacak bir deri lezyonu dahi ilgili bölümlerce eksizyonel biyopsi yöntemleri kullanılarak olası bir kanser olgusunun erken yakalanabilmesi amacıyla tanılandırılmalıdır.

Anahtar Sözcükler: Vulvar skuamöz hücreli karsinom, vulvar lezyon, kadın dış genitalya

INTRODUCTION

Vulvar squamous cell carcinoma (VSCC) is the fourth most common type of gynecological cancer and affects the female external genitalia. It accounts for approximately 3-5% of all gynecological malignancies, with an incidence rate of 1–2/100 000.[1] VSCC is a disease that occurs in the sixth to seventh decade of life, and increase in incidence all over the world owing to aging population. But it is almost very rare in younger group.

Over the past decade, the prevalence of vulvar intraepithelial neoplasia (VIN) in young women has increased significantly.[2] VIN is clearly a premalignant finding and is associated with HPV infection, particularly subtypes 16 and 18.[3]

The most frequently reported symptom of vulvar cancer is a long history of pruritus especially in older patients. Less common presenting symptoms include vulvar bleeding, discharge, dysuria, and pain. The most common presenting sign of vulvar cancer is a vulvar lump or mass. Rarely, patients present with a large, fungating mass. On physical examination, the vulvar lesion is usually raised and may be fleshy, ulcerated, leukoplakic, or warty in appearance.[1] Most of the squamous cell carcinomas are unifocal and occur on the labia majora. Approximately 5 percent of cases are multifocal, and the labia minora, clitoris, or perineum may be involved as primary sites. Therefore it is difficult to determine a VSCC case in a young female which resort to clinic with multifocal spreading vulvar lesions, as our patient.

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CASE REPORT

Twenty-two years old, non-virgin, a white female presented with multifocal epidermal lesions at mostly from labia majora through to anal area (Figure 1).

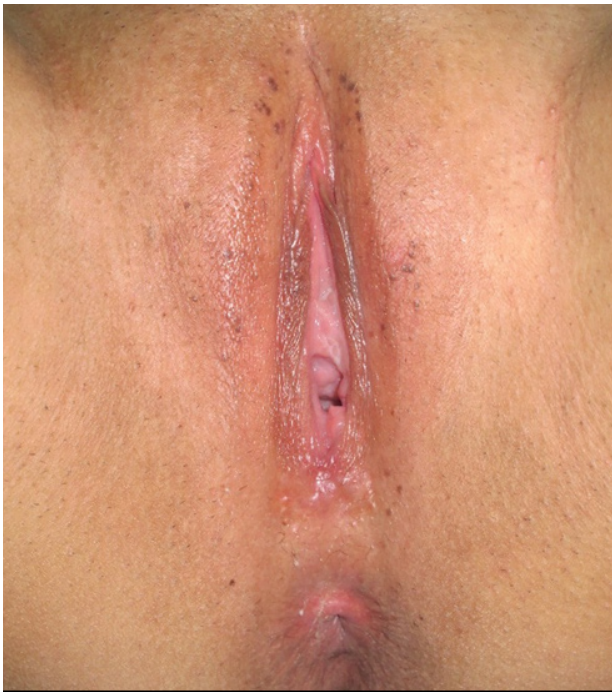

Figure 1. Multifocal epidermal lesions spreading from labia majora through out to anal area.

Our patient had a pathological result, which was taken by a dermatologist with shave biopsy procedure were vulvar in-situ squamous cell carcinoma and the tumor suppressor gene p53 mutation positive. Sheave biopsy is an improper technique that's why we planned a total surgical excision with vulvar tissue sparing, without sentinel lymph node sampling in order to justify previous pathological result. Because the previous pathological result was pointed out an in-situ carcinoma, serum tumor marker levels were negative. Abdominal ultrasonography examination revealed a single fusiform reactive lymph node on the right inguinal chain, although neither abdominal computerized tomography scans nor positron emission tomography evaluation found any metastatic focus or suspicious lesions. Afterwards surgical excision applied under spinal-epidural combined anesthesia, all lesions removed with full cut (Figure 2) skin margins and after undermining of the skin edges primary closure can achieved without tension (Figure 3). The surgically resected specimen consisted of epidermis, dermis, hypodermis and superficial fascia (Figure 4). Healing period was ended uneventfully.

RESULTS

Multifocal vulvar in-situ squamous cell carcinoma revealed that in permanent pathological evaluation and all lesions can be removed. Skin margins were obtained with safety margin. Afterwards gynecological consultation requested and smear examination showed that our patient had HPV 16 positive due to this, cervical entrance excision by conization technique planned and applied in order to eradicate and diagnose of a probable cervical cancer.

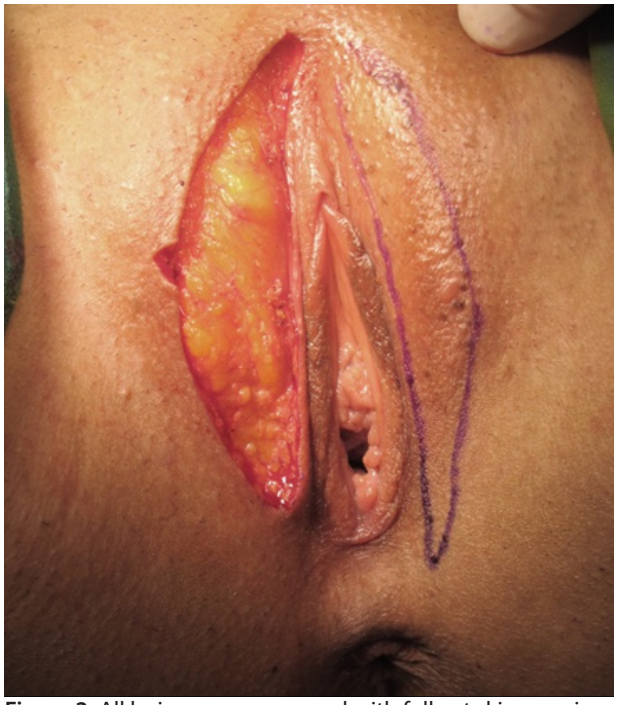

Figure 2. All lesions were removed with full cut skin margins.

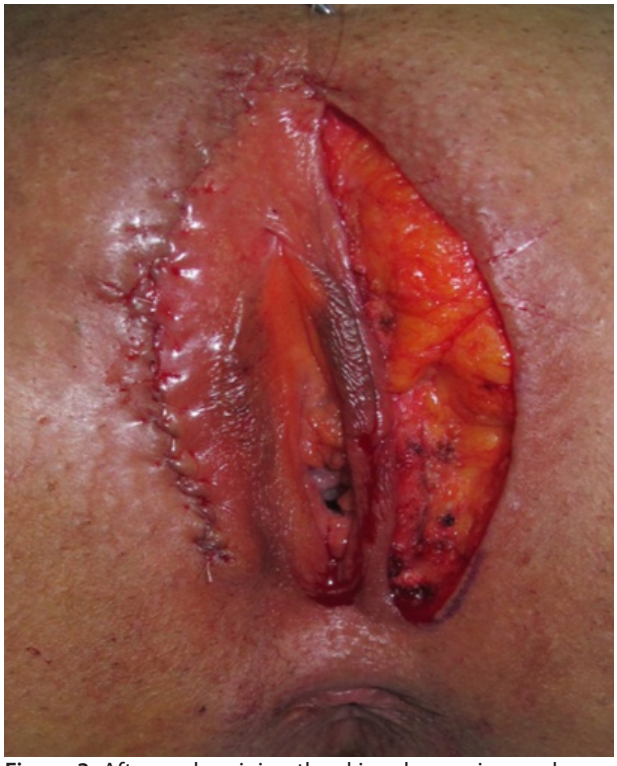

Figure 3. After undermining the skin edges, primary closure was achieved without tension.

Later on the surgeries, oncologic consultation requested for presence of dissemination to body and systemic treatment necessity. The oncology depart-

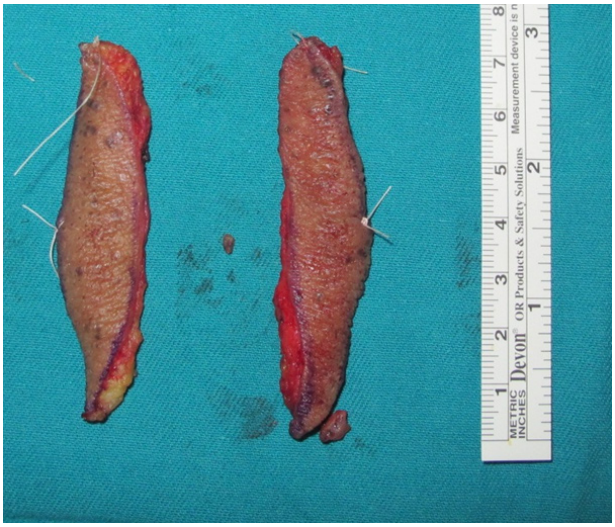

Figure 4. Surgically resected area consisted of epidermis, dermis, hypodermis and superficial fascia.

ment did not found any evidence in terms of systemic dissemination and only recommended close follow up. Additionally psychiatry consultation requested for mood alterations. Total treatment process ended healthfully and close monitoring suggested. The patient was reminded to be cautious for new developing lesions and their early resection. In the 11-month follow-up period, no new or suspicious lesions were assessed with dermatoscopic evaluations. In the 3, 6 and 9 month follow up abdominal ultrasonography revealed that the single lymph node which displayed before surgery was indifferent from first assessment.

DISCUSSION

The diagnosis of early invasion in vulvar carcinoma remains one of the most contentious areas in vulvar pathology, and both over-diagnosis and under-diagnosis are common.[8]

Brown vulvar lesions are quite common in the general population. Although the great majority of these lesions are benign, those that cannot be definitively diagnosed clinically should be biopsied. Reviewing difficult cases with colleagues who specialize in vulvar lesions is highly recommended, given that in the vulva some lesions that grossly, dermoscopically and microscopi-cally appear atypical can actually be normal variants and thus are not associated with risk of malignancy. The benefit of careful, experienced evaluation is significant for the patient's physical and emotional well-being, preserving the skin when possible, but this must be balanced against the potential risk of invasive, life-threatening malignancy.[9]

Invasive SCC of the vulva is an uncommon disease that develops in older women and is quite rarely observed in women younger than 30 years of age [4]. Our case is 22 years old. Despite her genital multifocality (vulva, perineum, and perianal region) of squamous cell carcinoma, our patient showed a problemless clinical course in terms of local and systemic dissemination.

The prognosis of patients with vulvar cancer is generally good when early and appropriate treatment is initiated in a timely fashion. The overall five-year survival is 70 percent and correlates with the stage of disease and lymph node status. The number of positive groin nodes is the important prognostic factor. Other factors that influence prognosis are tumor size and tumor ploidy (i.e., number of pairs of chromosomes).[5]

In recent years, vulvar carcinoma has emerged as a disease heterogeneous with respect to HPV expression, histological subtype, and patient age.[6] HPV expression varies in tumors with different histological growth patterns.[6] For example, carcinomas which are exophytic ("warty carcinoma") are more likely to contain HPV than flat carcinomas.[6] In older patients, cancers are more likely to be HPV-negative than in younger patients.[6] Heterogeneity in HPV expression may also exist in vulvar dystrophy in the presence and absence of vulvar carcinoma.

The authors concluded that ongoing vaccination programs will most likely result in a notable reduction of precursor lesions of vulvar cancer and anticipated that the reduction will translate into lower rates of vulvar cancer.[7]

The treatment of vulvar carcinoma has evolved during the last decades towards to more conservative and individualized surgery. Beware of radical treatment procedures increasingly become popular hence it is hard to reconstruct genital structures once damaged.

CONCLUSION

We recommend that women with any suspicious or newly developed vulvar lesions must be referred to related surgery department for incisional or excisional biopsy in order to early determination of a cancer beginning. VSCC is visible very few among young women but it is important to keep in mind that if a lesion observed which is looking benign should be remove for distinguish from malignancy.

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