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Vulvar Cancer – A Rare Diagnosis in Identical Twins

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Abstract

Vulvar cancer is a rare neoplasm, accounting for 4% of gynecological cancers, 90% of which are squamous cell carcinomas. This tumour is more frequent in postmenopausal women, with a peak incidence in the 6th to 7th decades of life. Clinically, patients may present with vulvar ulcer, bleeding, pain, edema and leukoplakia but severe pruritus is the most common symptom. Diagnosis is based on the histological examination and the treatment is surgical. Vulvar cancer might be curable in early stages, but in locally advanced disease is associated with a poor 5 year survival rate. The authors describe two case reports of identical twins with malignant vulvar neoplasia. In literature, only one case of this malignancy is described in two twin sisters. This fact raises the hypothesis of a genetic component in the etiology of the disease.

Keywords: vulvar cancer, squamous cell carcinoma, sarcoma, identical twins

Introduction

Vulvar cancer is a rare neoplasm, accounting for 4% of gynecological cancers, 90% of which are squamous cell carcinomas, representing 0.6% of all cancers in women [1-3]. Vulvar cancer is more frequent in postmenopausal women, with a peak incidence in the 6th to 7th decades of life. However, there has been an increase in the incidence in younger women, probably due to the increasing rates of human papillomavirus (HPV) infection and smoking [1, 4]. The most common site of vulvar cancer is labia majora [1]. Two pathways have been described for vulvar cancer pathogenesis [5-7]. The first subtype is related with HPV infections that causes vulvar intraepithelial neoplasia (VIN) and the tumours are of the basaloid or condylomatous type, accounting for approximately 60% of vulvar cancers [5-7]. This subtype of vulvar cancer occurs in younger patients (35–65 years of age) and is associated with other risk factors such as smoking, history of other sexually transmitted infections and immunosuppression [5-7]. The second subtype is independent of HPV and is associated with lichen sclerosus or lichen planus and the tumours are usually of the keratinizing type. It is more common in elderly patients (55-85 years) and a mutation in p53 gene is commonly present [5, 6].

Clinically, patients may present with vulvar ulceration, bleeding, pain, edema and leukoplakia but severe pruritus is the most common symptom [1, 6]. Incisional biopsy with histological exam is mandatory for diagnosis [5]; imaging exams are not indicated for the diagnosis of vulvar cancer, but are important for staging of the disease [6]. The treatment of vulvar cancer is surgical, through local resection of the tumor or radical vulvectomy, depending on tumor extension, with or without excision of local lymph nodes [6, 8]. Vulvar cancer might be curable in early stages, but in locally advanced disease is associated with a poor 5 year survival rate, inferior to 40% [4´1].

Case Report

The authors report clinical cases of malignant vulvar neoplasia in two monozygotic twin sisters. A 78-year-old woman, with a previous history of myelodysplastic syndrome, was diagnosed with epithelioid vulvar sarcoma. She went to the emergency department because of a genital blood loss and exophytic hardened and ulcerated lesion on the right labia minora with 4 cm (Figure 1). It was performed an incisional biopsy which revealed a vulvar epithelioid sarcoma. The thoraco-abdomino-pelvic computed tomography (CT) revealed

"multiple scattered lung nodules of about 1 cm; apparently reactive adenopathies in the inguinal regions, without adenomegaly of the obturator, iliac and retroperitoneal chains." Palliative vulvectomy was decided without inguinal emptying (Figure 2). The definitive anatomopathological exam confirmed the diagnosis of vulvar epithelioid sarcoma. Post-operative was complicated with dehiscence of the operative wound and pulmonary thromboembolism and the patient died 25 days after surgery. A 77-year-old woman, with a history of vulvar sclero-atrophic lichen, had been submitted to radical vulvectomy with inguinal emptying when she was 62 years old, whose anatomopathological exam study revealed a well differentiated and invasive squamous cell carcinoma, with negative margins and with no ganglionar invasion. The patient went to the Gyneacological consultation and presented vulvar lesion suggestive of tumor recurrence, confirmed by incisional biopsy. Abdomino-pelvic CT revealed an adenopathy on her left inguinal region. Surgical excision of vulvar recurrence lesion and left inguinal adenopathy was performed and she underwent adjuvant treatment with radiotherapy and brachytherapy. Six months after surgery the patient had bone pain complaints. An abdomino-pelvic CT was performed which revealed "suspected iliac-obturator adenopathies", confirming epidermoid carcinoma metastases after biopsy. The patient underwent palliative chemotherapy, after group decision on an Oncology Gynecological consultation.

Discussion

The incidence of vulvar cancer is 3 per 100,000/year [5]. The most common type of vulvar cancer is squamous cell carcinoma accounting for about 90% of the cases [1]. Vulvar sarcomas are very rare in adults and the prognosis is poor, depending on the histological type, local extent of invasion and mitotic activity; the majority of these tumors metastasize hematogenously and lymphatic invasion is uncommon [9]. Vulvar cancer spreads by local extension, lymphatic embolization and hematogenic dissemination. Regional lymphatic invasion is very common, even for small tumours, due to the high number of lymphatic vessels in vulva: the first lymphatic invasion occurs in the superficial inguinal ganglia, followed by the femoral ganglia and then to the pelvic ganglia [10]. As noted previously, with the exception of vulvar sarcomas, hematogenous metastasis is rare and late, occurring primarily to the lungs, liver and bone [10]. The most important prognostic factors for vulvar cancer are the stage of disease, the size of lesion, and the depth of invasion [11]. Standard treatment for vulvar cancer is surgical through local resection of the tumor or radical vulvectomy and excision of local lymph nodes, depending on tumor extension and invasion [6, 8]. There is a high rate of complications associated with surgery, around 60%, usually with dehiscence of the suture, infection, lymphocele and chronic lymphedema. Surgery with sentinel node evaluation has led to a decrease of this complication rate [4]. Approximately 80% of recurrences occur within the first 2 years after treatment, particularly in patients with invaded lymph nodes. Thus, a close follow-up is critical to early diagnosing recurrent disease: every 3 months in the first 2 years, then every 6 months to the 5th year and annual after the 5th year [12]. In literature, only one case of this malignancy is described in two twin sisters [13]. This fact raises the hypothesis of a genetic component in the etiology of the disease.

Conclusion

Vulvar cancer is a rare neoplasm and in literature only one case of this malignancy has been described in two twin sisters [13]. This fact raises the hypothesis of a genetic component in the etiology of the disease.

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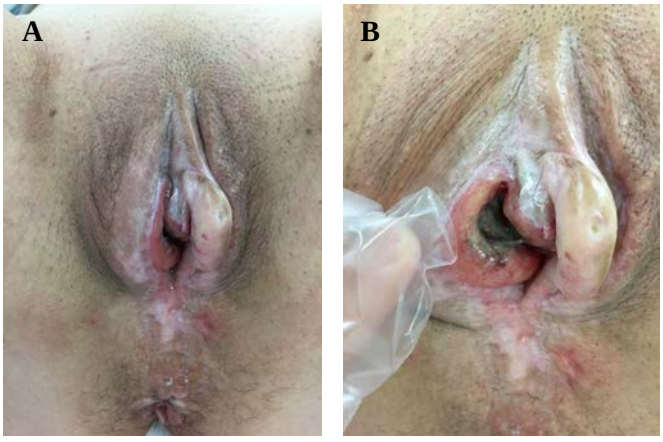

Figure 1 (A, B): ulcerated lesion on the right labia minora with 4 cm and induration of left labia minora.

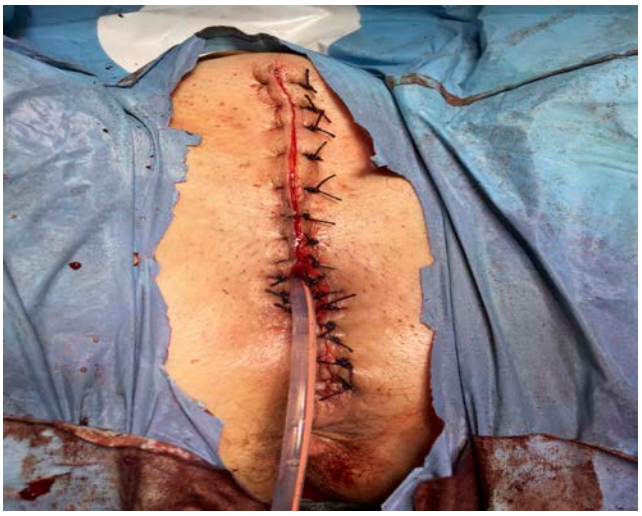

Figure 2: Post-operative result of palliative vulvectomy.