Basal cell carcinoma of the vulva: case report

Introduction: Basal cell carcinoma (BCC) of the vulva is a rare condition that accounts for less than 0.4% of BCC cases and 2% to 4% of vulvar neoplasms. BCC of the vulva is more common among white, multiparous, and postmenopausal women, especially in the seventh decade of life. The objective is to report a case of BCC of the vulva in which aspects of diagnosis and treatment were discussed. Case Report: A 63-year-old woman, G1P1A0, arrives at the office in January 2022 for treatment of a persistent lesion on her vulva. An incisional biopsy was performed which showed that it was likely nodular basal cell carcinoma with invasion of the dermis. The patient underwent tumor resection with free macroscopic margins and primary suture. The surgery had no complications preoperatively or postoperatively. The histopathology of the surgical specimen showed that it was a nodular basal cell carcinoma with an irregular, flat, white area, measuring 0.7x0.4cm, with the lateral margins 7.0 and 5.0mm apart and deep, 5.9mm; all free. Conclusion: The reported case is rare, with surgical resection of BCC of the vulva with margins being successful. Fourteen months after surgery, the patient has no evidence of local or regional recurrence.

Keywords: Skin neoplasms; Carcinoma, basal cell; Vulva; Diagnosis, differential; Case reports.

ABSTRACT

Introduction: Basal cell carcinoma (BCC) of the vulva is a rare condition that accounts for less than 0.4% of BCC cases and 2% to 4% of vulvar neoplasms. BCC of the vulva is more common among white, multiparous, and postmenopausal women, especially in the seventh decade of life. The objective is to report a case of BCC of the vulva in which aspects of diagnosis and treatment were discussed. Case Report: A 63-year-old woman, G1P1A0, arrives at the office in January 2022 for treatment of a persistent lesion on her vulva. An incisional biopsy was performed which showed that it was likely nodular basal cell carcinoma with invasion of the dermis. The patient underwent tumor resection with free macroscopic margins and primary suture. The surgery had no complications preoperatively or postoperatively. The histopathology of the surgical specimen showed that it was a nodular basal cell carcinoma with an irregular, flat, white area, measuring 0.7x0.4cm, with the lateral margins 7.0 and 5.0mm apart and deep, 5.9mm; all free. Conclusion: The reported case is rare, with surgical resection of BCC of the vulva with margins being successful. Fourteen months after surgery, the patient has no evidence of local or regional recurrence.

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RESUMO

Introdução: O carcinoma basocelular (CBC) de vulva é uma condição rara que corresponde a menos de 0,4% dos casos de CBC e de 2% a 4% das neoplasias de vulva. CBC de vulva é mais comum entre mulheres brancas, multiparas e na pós-menopausa, especialmente na sétima década de vida. O objetivo é relatar um caso de CBC de vulva no qual discutiram-se os aspectos do diagnóstico e tratamento. Relato de Caso: Mulher de 63 anos de idade, G1P1A0, chega ao consultório em janeiro de 2022 para tratamento de lesão persistente em vulva. Realizou-se biópsia incisional que mostrou tratar-se de provável carcinoma basocelular nodular com invasão da derme. A paciente submeteu-se a uma ressecção do tumor com margens macroscópicas livres e sutura primária. A cirurgia não teve complicações no pré-operatório e no pós-operatório. O histopatológico da peça cirúrgica mostrou tratar-se de carcinoma basocelular nodular com área irregular, plana, branco, medindo 0,7x0,4cm, com as margens laterais distando 7,0 e 5,0mm e profundas, 5,9mm; todas livres. Conclusão: O caso relatado é raro, tendo sido o tratamento de ressecção cirúrgica do CBC de vulva com margens bem-sucedido. Catorze meses após a cirurgia, a paciente encontra-se sem evidências de recidiva local ou regional.

Descritores: Neoplasias cutâneas; Carcinoma basocelular; Vulva; Diagnóstico diferencial; Relatos de casos.
INTRODUCTION

Basal cell carcinoma (BCC) of the vulva is a rare condition, as it represents less than 0.4% of all BCC cases and 2% to 4% of vulvar neoplasms. It is estimated that around 6,190 cases occur annually in the world and an increase of 4.6% in the number of cases is expected every 5 years. The highest prevalence occurs in white, multiparous, and postmenopausal women, especially in the seventh decade of life, therefore, advanced age is a risk factor, but it can also occur in young patients1,2.

Regarding the clinical presentation, the lesions can vary from solitary pinkish-red papules or plaques on the labia majora to bilateral, multiple lesions, and in advanced stages, ulceration occurs. Regarding etiology, since the affected region is located in an area protected from solar radiation, there is possibly an association with the following factors: chronic inflammation, exposure to ionizing radiation and arsenic, immunosuppression, pelvic radiation or trauma; that is, damage that is not related to ultraviolet radiation, but that can also generate mutations. Therefore, it is important to research secondary conditions such as lichen sclerosus, nevoid syndrome, and Paget’s disease, since there are documented cases in the literature of BCC in these diseases3.

Symptoms are nonspecific and include itching, pain, ulcerations, and bleeding. These are part of a condition with late and variable clinical presentation and are therefore factors that can delay diagnosis and treatment. The most common type is nodular, to the detriment of other rarer variants such as superficial, infiltrative, and mixed3.

Dermoscopy presents characteristics similar to those of other sites: oval, bluish-gray nests, arborizing telangiectasias, and leaf-shaped structures with bright clear areas and ulceration4. Treatment is surgical, but recurrence may occur because the edges of the tumor are not easily well delineated, making it difficult to obtain free surgical margins and increasing the chances of performing incomplete excisions5,6.

OBJECTIVE

The objective of the study was to report a case of BCC of the vulva in which aspects of diagnosis and treatment were discussed.

CASE REPORT

A 63-year-old woman, G1P1A0, presented to the office in Teresina, PI, in January 2022 for treatment of a persistent lesion on the vulva. She noticed the presence of the nodule after an ingrown hair persisted while waxing the area. Her history included systemic arterial hypertension and a total abdominal hysterectomy, with preservation of the ovaries, performed in 2006 due to symptomatic myoma and smoking for around 30 years.

The gynecological examination revealed a raised tumor in the upper third of the labia majora on the left, measuring approximately 1.0x0.8cm, without ulceration and slightly irregular edges, in addition to the absence of suspected inguinal lymphadenopathy on clinical and ultrasound examination (Figure 1).

A transvaginal ultrasound and vulvoscopy were indicated. A small simple cyst on the left ovary and a hyperchromic lesion with a smooth surface on the left labia majora were identified, respectively, suspected of neoplasia. The histopathology of the material from the incisional biopsy of the lesion (Figure 2) showed that it was likely nodular basal cell carcinoma, with invasion of the dermis and without perineural invasion.

The patient underwent resection of the vulvar tumor with free macroscopic margins and primary suture. During the surgery, there were no complications and the patient was discharged one night after the procedure. In the immediate and late postoperative period, there were no complications, such as dehiscence and/or inflammation. The histopathology of the surgical specimen (Figure 3) showed a nodular basal cell carcinoma with an irregular, flat, white area, measuring 0.7x0.4cm, with the lateral margins 7.0 and 5.0mm apart and the deep margins, 5.9mm; all free. The patient, 14 months after surgery, has no evidence of local or regional recurrence.

Figure 1. Injury to the vulva presented by the patient.
The study was approved by the Research Ethics Committee of the State University of Piauí (CEP-UESPI), Opinion No. 4,311,835/2020. The patient signed the Free and Informed Consent Form (TCLE) for the publication of the case.

DISCUSSION

Currently, some techniques such as the use of a dermatoscope and reflective confocal microscopy are being highlighted as diagnostic advantages in the context of BCC. This is because they provide visualization of characteristics inherent to BCC, such as linear telangiectasias, arborizing veins, and nests of oval bluish-gray cells, in addition to pearly structures. In the present case, as the main suspicion was neoplasia based on vulvoscopy, it was decided to perform a punch biopsy. It should be noted that the later the diagnosis, the greater the chances of mutilating surgeries that compromise the patient’s quality of life.

Also, according to validation in a series of recent cases, the site most affected by BCC of the vulva is the labia majora, in around 90% of cases, and the average age of affected women is 71.9 years, being the most affected ethnic group, the Caucasian. As stated above, our patient is within the age range most frequently affected by BCC of the vulva, and the affected site was considered the most frequent in the literature (labia majora), although there are cases in which the lesion is located on the clitoris, in the labia minora, in the perineum, and the vaginal introitus.

The initial symptoms may only be itching or non-specific superficial lesions, however, if not diagnosed and treated properly, they may progress to ulceration, bleeding, and pain, since cancer in the vulva can have a more aggressive behavior than in photo-exposed areas, with more perineural invasion and local infiltration. It is worth noting that patients with basal cell nevus syndrome, that is, a mutation in the \( PTCH \) gene, are more likely to develop the disease and require regular skin exams as a follow-up.

As for types, the most common BCCs are nodular types, followed by superficial ones. Generally, there is no need for a lymph node approach, as metastatic involvement is rare. Inguinal lymphadenectomy is only indicated in the presence of clinically evident lymph node metastasis. BCC is an entity that grows indolently and has a low propensity to metastasize. Imaging studies are reserved for advanced cases to evaluate the involvement of adjacent structures and help with surgical planning.

The diagnosis is made by biopsy with the aid of histopathological analysis using hematoxylin and eosin dyes or the detection of typical markers by immunohistochemistry, with CK20 being the most indicated, which was not performed in our case due to the more morphological nature of the findings, which can generate questions, and is mostly used for academic purposes. The presentation can be confused with other diseases, such as psoriasis, dermatitis, lichen sclerosus, vulvar intraepithelial neoplasia, and Paget’s disease.
Regarding the development of cancer, the p53 and BCL2 genes are involved, participating in the regulation of the cell cycle. The Ki-67 and PCNA markers are linked to cancer predisposition. Mutations in P53 can lead to the malignancy of pre-existing lesions and in BCL2 to the immortalization of the cell. As for the PCNA marker, it was not used in our case because it is reserved for identifying injuries related to UV light, with the present case being located in a non-photoexposed area. Ki-67 provides prognostic information about the tumor and was not used in our case because it is a non-ulcerated lesion with no signs of severity

Treatment can include topical application of 5% imiquimod, cryotherapy, curettage, or wide local excision, also more recently including the Mohs technique, with lower associated local recurrence rates, a problem better related to location, and not to the size of the lesion.

It is worth noting that there are no guidelines that corroborate the superiority of any technique in the treatment of these tumors and recurrence rates can reach 20%, although some authors suggest the surgical approach by excision with free margins as effective. Systemic therapy can be recommended in cases of positive margins, metastases, and advanced cases, with vismodegib being an excellent option (HH signaling inhibitor)\textsuperscript{9,10}.

**CONCLUSION**

The reported case of BCC of the vulva in a 63-year-old postmenopausal woman of age with a history of vulvar injury following chronic inflammation of the non-photo exposed site. It is rare, and the treatment was carried out through surgical resection of the tumor with margins. In 14 months of post-surgery follow-up, the patient is without evidence of local or regional recurrence.

**COLLABORATIONS**

**MCAS** Conceptualization, Formal Analysis, Writing - Original Draft Preparation.

**REARC** Conception and design study, Conceptualization, Writing - Original Draft Preparation.

**ECXC** Conception and design study, Conceptualization, Writing - Original Draft Preparation.

**ESBC** Conception and design study, Conceptualization, Writing - Original Draft Preparation.

**AMLFV** Conception and design study, Conceptualization, Supervision, Writing - Original Draft Preparation, Writing - Review & Editing.

**SCV** Conception and design study, Conceptualization, Supervision, Writing - Original Draft Preparation, Writing - Review & Editing.

**REFERENCES**