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Case Report

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Basal cell carcinoma of the vulva: a case report

Abstract

Background: Basal cell carcinoma (BCC) is the most common malignant neoplasm in humans, accounted for at least 75% of non-melanoma skin cancers. The BCCs are directly related to intense exposure to ultraviolet radiation, being diagnosed preferentially in body areas with larger sunlight exposure. However, these tumors may appear in areas which are normally covered from ultraviolet light. Vulvar BCC is a rare tumor, accounting for less than 1% of all BCCs, with an unclear etiology, often delayed diagnosis and typically asymptomatic.

Methods and findings: The aim of this article is to report a case, due to its unusual location, of a patient with basal cell carcinoma diagnosed in the vulvar region, contributing to existing literature reports.

Conclusion: It was concluded that due to the characteristics of the vulva it is difficult an early diagnosis, favoring growth and local invasion of the tumor. The knowledge about vulvar BCC is important in the approach of vulvar lesions.

Keywords: Basal cell carcinoma, Vulva, Vulvar neoplasms

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Introduction

The Basal Cell Carcinoma (BCC) is the most malignant neoplasm in humans.[1,2] It is the most common cancer in Brazil, accounting for 25% of all malignant tumors registered in country.[3] BCC is the most common malignant tumor of skin in Caucasians.[4] The disease is responsible for 70% of non-melanoma cancers diagnosed in the country, with an estimated 175,760 new cases, being 80,850 in men and 94,910 in women in a period of two years (beginning of 2014 until January of 2016).[3]

In the United States represents 75% of all types of non-melanoma skin cancers,[2,9] with about 1,000,000 new cases diagnosed annually[1,2,8] with an approximate rate of 407 patients with BCC per 100,000 white men and 212 cases 100,000 per white women.[10] The body areas most affected by BCC are those with longer exposure to the sun, with an approximate incidence of 83% in the head and the neck.[11,12] However, these tumors can be found in areas which are normally covered from ultraviolet rays, such as genital and perianal areas, with an incidence of 0.44 to 2%.[1,9,11] The vulvar injury is uncommon, with an incidence less than the 1% of all BCC,[13] with represents less than 5% of all cancers in the vulvar region.[5,9]

In 1926, Temesvary was the first to describe a case of vulvar BCC,[15] since then have been reported about 250 new cases until 2006[5] and about another 20 new cases of BCC in the vulva until January of 2015.[7,8,14]

Case description

The patient was female, Caucasian and was 60 years old. She came to the gynecology service for gynecological routine. The patient was postmenopausal, with the last menstrual period dated at the age of 48. Obstetrical history: Gestation: IV To: III by caesarian Abortions: I. During consultation she reported vulvar spot of longstanding evolution, with itching. She expressed the wish to remove it. The examination showed hypochromic vulvar lesions in the order of 3 to 4 cm long and 1.0 cm wide at the large left labia (Figure 1). Due to the harmless appearance and the small size of the vulvar lesions it was made a biopsy, showing histopathologic diagnosis of basal cell epithelioma with compromised margins. The tissue underwent a total resection of the lesion and the histopathology report was Basal Cell epithelioma with expansive pattern, occupying the dermis media and with free margins (Figure 2).

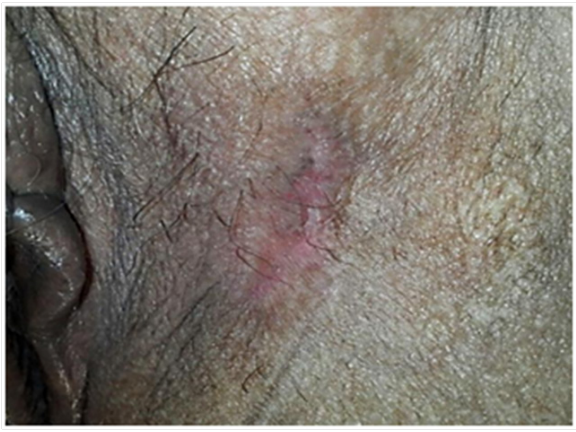
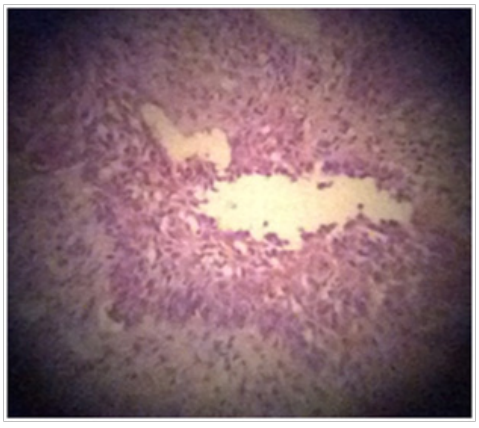


Figure 1 Hypochromic lesions in large left labia.



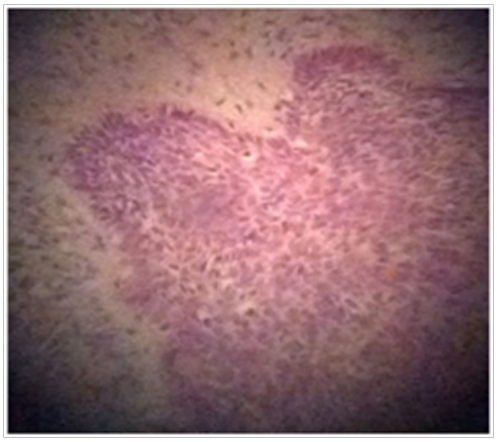


Figure 2 Image of histopathology of the specimen, showing expansive pattern, occupying media dermis.

Discussion

The most important risk factor of the incidence of BCC is directly related to intense exposure to ultraviolet radiation.[10] It has not been elucidated the reasons for the appearance of BCC in the vulva. Syphilis, chronic irritation, chronic infection, trauma, arsenic, age, immune deficiency, certain genetic diseases such as Gorlin syndrome, xeroderma pigmentosum, human papilloma virus (the most relevant in the squamous cells of the genital etiology), mutations in the p53 gene and ionizing radiation have been implicated as potential causes.[1,6,15,16] Usually, vulvar BCC affects Caucasian and postmenopausal women,[15] being relatively rare in childrens and black women.[3] It is found mainly between the ages of 34 to 96, with an average age of 70,[1,15] and there is one report of a patient with 20 years of age.[8]

The vulvar BCC is a disease with late diagnosis, characterized for being often asymptomatic, with non-specific physical signs and slow gradual growth.[4,6] When expressed, the symptoms are often present for a long period of time, the most common are itching, irritation and presence of palpable mass in the vulvar region.[15] The vulvar BCC is characterized by a lack of pigmentation and by showing similar clinical signs of other dermatological diseases such as infectious or inflammatory dermatoses, eczema or psoriasis. Therefore, the correct diagnosis is generally delayed by inadequate therapeutic measures, allowing tumor growth and increasing their invasiveness and discomfort to the patient. Vulvar BCCs, as well as in other body sites, can be presented as a rounded ulcer with jagged edges or as nodules or blotches. In the vulva most of the lesions are smaller than 2 cm in diameter and typically located in the labia.[1,7] Although they are usually slow growing tumors, they are locally invasive and destructive, with the risk of recurrence and metastasis. They have a tendency to grow over the anatomical path of least resistance, most commonly on the labia majora; this explains the delayed bone invasion, cartilage and muscle.[7]

Owing to the innocuous appearance BCC in these places, it is recommended the biopsy of all suspicious lesions[9] and the standard treatment consists of wide local excision with clear margins of approximately 1 cm histologically proven.[8,15,17] In cases where due to aesthetic or functional factors can not be obtained free margins, Benedet concluded that these patients can perform clinical follow-up safely due to the low propensity of tumor spread.[15]

Surgical resection of these lesions of the vulva should be larger than in other regions of the body due to the high number of recurrences. De Giorgi and colleagues[1] demonstrated commitment margins in 25% of cases of biopsies in the vulva,[1] which could explain the high rates of local recurrence, reaching 20% in some revisions,[9,15,16,18] the result of an underestimation of extension of tumor margins.[6,7]

In bulky or histologically aggressive types of BCC as morphea-like, or basosquamous (metatypical) carcinoma and perineural invasion of tissue involvement may be greater than clinically suggested. In the cases where the wide resection of the lesion is inadequate, with local recurrence risk and insufficient treatment, it is indicated the Mohs micrographic surgery.[6,9,18]

Mohs surgery involves excision of the tumor under controlled and careful microscopic monitoring, with the advantage of the study of deep and lateral margins at the time of resection, providing a relative certainty of complete tumor resection with minimum removal of healthy tissue.[6] This technique has been used particularly in the treatment of recurrent BCC, with cure rates greater than 97%.[6,18] It should also be considered in cases where it is necessary to preserve tissue in critical anatomical regions, such as the vulva and the clitoris.[6,8]

At first it was thought that the vulvar BCC did not generate metastasis, until in 1975 Jimenez reported the first known case of metastasis to the inguinal lymph nodes.[19] Despite cases of vulvar BCC metastasis are rare, with an incidence lower than 0.1%, and due to few reports in the literature these data are not representative.[6,14]

BCCs are moderately sensitive to radiation therapy. Although there are few cases reported for a long period of time, cure rates are similar with other approaches such as chemotherapy[6,7,20] associated with unpleasant clinical and aesthetic effects.[6,8,17]

The systematic chemotherapy is not useful for treatment of localized BCC. Some studies suggest that Cisplatin can present results alone or in combination with doxorubicin in patients with metastatic disease, however, with no definitive conclusions.[6,8] Adjuvant treatment of metastatic vulvar BCCs should be individualized to the type and degree of dissemination of the disease as well as the needs of the patient.[6]

Conclusion

Basal cell carcinoma in the vulvar area is diagnosed with low frequency, with a clinical presentation not characteristic, hindering its early diagnosis and promoting growth and local tumor invasion. Therefore, it is very important to keep in mind this disease in the approach of vulvar lesions, its insidious behavior and confusingly symptoms with infectious or inflammatory diseases leads to late consultation of patients.

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None.

Conflicts of interest

None.

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