PILOMATRIX CARCINOMA IN THE PRESTERNAL REGION. VERY RARE OR VERY OFTEN MISDIAGNOSED MALIGNANCY?

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ABSTRACT

Pilomatrix carcinoma is a very rarely diagnosed malignancy. There are around 150 cases reported in the literature. In the very past this type of carcinoma was considered as a low-grade, non-metastasizing tumor. Nowadays with its high potential of recurrence and metastasizing the approach to this type of neoplasm should always be careful and multidisciplinary. The first line of therapy is wide surgical excision, followed by adjuvant radiotherapy. If there is observation of lymph node metastases they should be surgically or radiotherapeutically treated. In the present case report, we present a 46-year-old patient with pilomatrix carcinoma in the presternal region.

Keywords: *pilomatrix carcinoma, recurrence, metastasizing, surgery, radiotherapy*

INTRODUCTION

Pilomatricoma is a rare benign tumor with differentiation towards hair matrix cells. It is mostly found in the head and neck region and occurs before the third decade. It was first described by A. Mahlerbe and J. Chenantais as „calcifying epithelioma" in 1880 (1). W. Dubreuilh and E. Cazenave reported the main histopathological features of the tumor in 1922 (2).

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The locally aggressive and infiltrating variant of pilomatricoma was described by S. Lopansri and M. C. Mihm in 1980, so it was named "pilomatrix carcinoma" or "calcifying epitheliocarcinoma of Mahlerbe" (3,4).

In the past this type of neoplasm was considered as one with low-grade malignancy rate and non-metastasizing characteristics (3,4). Today, pilomatricoma is classified as a very recurrent type of malignancy with high metastatic properties (5). Cases have been reported for both metastatic lymph nodes and systemic metastases (mainly pulmonary and bone ones) (6-11).

In this case report, we present a 46-year-old male with a slow-growing, erythematous, plaque-like lesion in the presternal region.

CASE REPORT

A 46-year-old male was examined in the Department of Dermatology and Venereology of the

Medical Institute of Ministry of Internal Affairs with a slow-growing, erythematous, plaque-like lesion in the presternal region (Fig. 1). The tumor showed an infiltrated, pearl-like border, islands of brown pigmentation and solitary telangiectasias, 40 mm/20 mm in size. The first clinical diagnosis was pigmented basal cell carcinoma. In the differential diagnosis, squamous cell carcinoma, Bowen's disease and trichoepithelioma were considered.

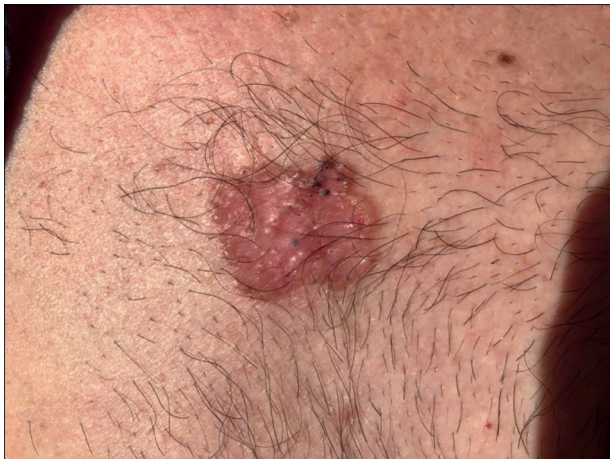

Fig. 1. Clinical presentation of the chest lesion

A biopsy specimen was taken for histopathological verification of the clinical diagnosis.

Microscopically the tumor showed the typical features of pilomatrix carcinoma (Fig. 2, 3, 4).

The lesions were composed of lobules of matrical cells, irregular foci of necrosis. The tumor cells were of basaloid type with medium degree of anaplasia and high mitotic rate. Pilar-type keratinization was observed in the center of several lobules. The epidermis appeared atrophic. The final dermatohistopathological diagnosis was pilomatrix carcinoma.

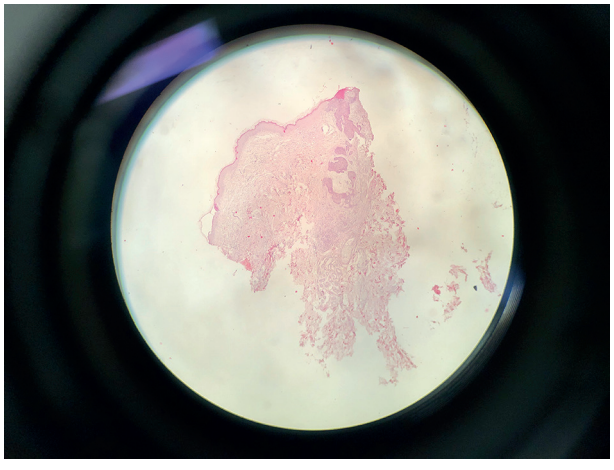

Fig. 2. Histological findings (x40)

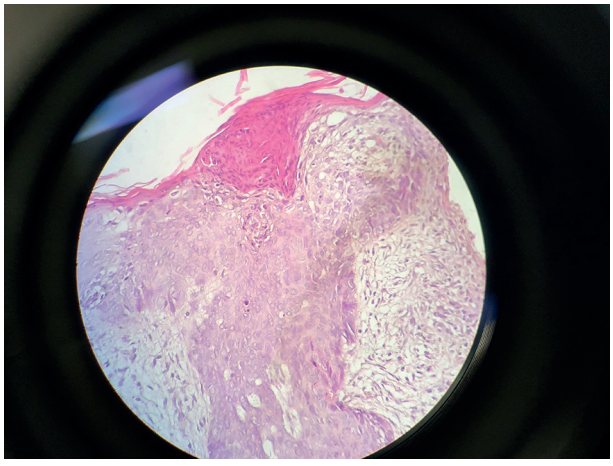

Fig. 3. Histological findings (x100)

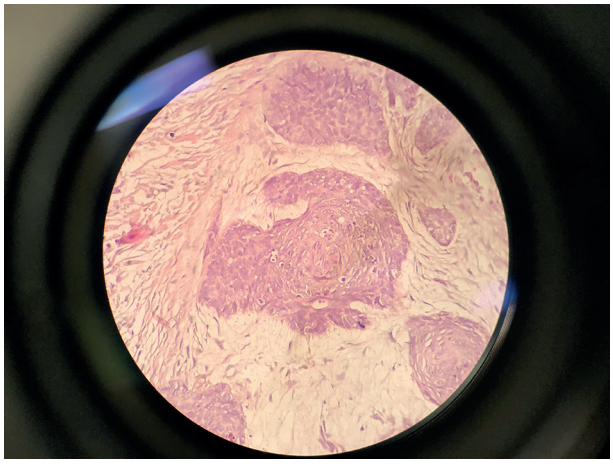

Fig. 4. Histological findings (x200)

Computed tomography scan was performed in order to stage the disease. There was no evidence of regional or distant metastases.

DISCUSSION

Pilomatrix carcinoma is a rarely diagnosed neoplasm (12). In contrast to its benign counterpart, pilomatrix carcinoma has a typical male predominance (male to female ratio of 4:1) and commonly occurs in the 5th to the 7th decade (12,13). It typically presents as a solitary, slow-growing, hard, indurated, painless and asymptomatic formation (14). The majority of these tumors are located in the head and neck region, mainly in the pre- and postauricular

area, scalp, posterior neck and upper back (15,16,17). Clinical differential diagnosis includes benign pilomatricoma, basal cell carcinoma, squamous cell carcinoma, Merkel cell carcinoma, and other pigmented and vascular lesions.

The histology of pilomatrix carcinoma reveals irregular nests of large anaplastic basaloid cells with prominent nucleoli and abundant mitotic figures. Shadow cells, transition to squamous ones, invasion of blood and lymphatic vessels, ulceration and infiltration are found in some cases (17,18).

It is still controversial, whether pilomatrix carcinoma arises de novo or represents malignant transformation of a pre-existing benign pilomatricoma (19,20). The first choice of treatment is surgical excision with wide free margins, although most of the reported cases show a high recurrence rate. There is still no consensus on optimal surgical excision margins. Some authors consider 5 mm as sufficient, while others insure themselves with 20 mm. Screening should also be performed, since cases of metastases in the regional lymph nodes, bones and lungs have been reported (16-20).

Recently many cases of recurrence of surgically removed pilomatrix carcinoma have been published. Consequently, an adequate adjuvant therapy should be performed. Nevertheless, the administration of chemotherapeutic agents has not proven to be effective (21,22). On the other hand, radiotherapy showed good tumor control in most of the recurrence cases.

Radiotherapy may also be an alternative to the surgical dissection of involved lymph nodes, and provide palliative treatment of systemic metastases (23,24).

In conclusion, in the present case a 46-year-old male with a presternal pilomatrix carcinoma has been reported. Pilomatrix carcinoma should always be considered in differential diagnosis of slow-growing, plaque-like, infiltrated border lesions, as high recurrence rate and metastasizing potential remain considerably high. Wide surgical excision of the lesion should be considered and adjuvant radiotherapy is commonly included in the therapeutic plan. Follow-up examinations should also be arranged for detection of any lymph node or systemic metastases.

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