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CASE REPORT
Reconstructive

Giant Pilomatrix Carcinoma of the Thorax: An Uncommon and Clinically Misdiagnosed Tumor

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Summary: Pilomatrix carcinoma is an unusual and aggressive malignant tumor deriving from follicular matrix cells and exhibiting a metastatic potential to lungs and regional lymph nodes in 10% of cases. We report the case of a 54-year-old male patient who presented with a biopsy-proven malignant pilomatrixoma of the thoracic region, which recurred multiple times after previous excisions. Due to the size of the tumor (28 by 22 cm), wide excision and axillary lymphadenectomy of levels I and II were performed, followed by reconstruction with a free deep inferior epigastric artery perforator flap and adjuvant radiotherapy. Owing to its rarity, this tumor can initially be misdiagnosed, resulting in delayed treatment and recurrences if inadequately excised. Also, large tumor size is correlated with a higher incidence of metastasis. High index of clinical suspicion and wide excision are recommended, along with the need of establishing oncological guidelines for better prognosis.
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Pilomatrix carcinoma, malignant pilomatrixoma, or calcifying epitheliocarcinoma of Malherbe is an unusual and aggressive malignant follicular tumor, described by Lopansri and Mihm in 1980, and represents the malignant form of calcifying epithelioma of Malherbe.[1,2] Clinically, it presents as a solitary, firm, painless, ulcerated, and fast-growing subcutaneous nodule, usually located in the head and neck,[1] most commonly affecting middle-aged men.[3] Histopathology shows enlarged anucleate epithelial cells called "ghost cells" and baseloid cells with central necrosis.[4] The tumor is locally aggressive, and metastasis can occur in 10% of cases, usually in the lungs and regional lymph nodes.[5] Up to date, only a few cases of pilomatrix carcinoma have been described, which, owing to their rarity, are usually initially misdiagnosed. In this report, we present the oncological management of a patient with a recurrent giant pilomatrix carcinoma of thorax, which required wide excision and reconstruction with a free fasciocutaneous flap, due to its large dimensions and subcutaneous extension of the tumor.

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CASE REPORT

A 54-year-old male patient without comorbidities or medical history was referred to our outpatient department with a recurrent enlarging mass over the lateral thorax (Fig. 1). It appeared four years ago as an asymptomatic, subcutaneous nodule, and it was excised by a general surgeon. Due to its clinical presentation, it was treated as a sebaceous cyst, and the specimen was not sent for histopathology. The nodule recurred, and it was reexcised three more times by the same surgeon. After the last excision, the specimen was sent for tissue biopsy, receiving the diagnosis of malignant pilomatrixoma. Clinically, the mass was located on the right anterolateral thorax with palpable subcutaneous expansion, measuring 28×22 cm. There were no palpable lymph nodes in the ipsilateral axilla. On MRI, the lesion was described as a well-defined soft tissue mass extending up to the fascia of pectoralis major muscle (Fig. 2). Staging with thorax and abdomen computed tomography was clear of metastatic disease. Due to the size, the recurrences, and the location of the tumor, wide surgical excision with 2-cm peripheral margins and down to pectoralis major fascia was performed (Fig. 3). Axillary lymphadenectomy of levels I and II was deemed necessary because of the proximity of the tumor. The defect was reconstructed with a free deep inferior epigastric artery perforator flap with end-to-end anastomoses to the thoracodorsal vessels. The immediate postoperative course was uncomplicated. Histopathology confirmed the diagnosis of pilomatrix carcinoma. The tumor was excised with clear margins, and lymph nodes were free of metastatic disease. Due to the size of the tumor and its malignant

Disclosure statements are at the end of this article, following the correspondence information.

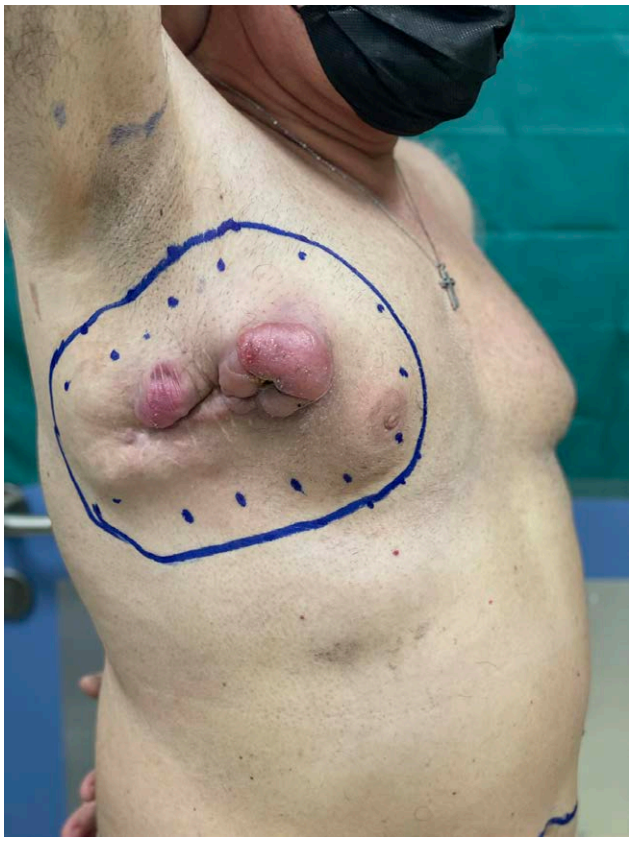

Fig. 1. Preoperative view of a recurrent enlarging tumor of the lateral thoracic region, with palpable subcutaneous expansion (dotted lines), measuring 28 × 22 cm.

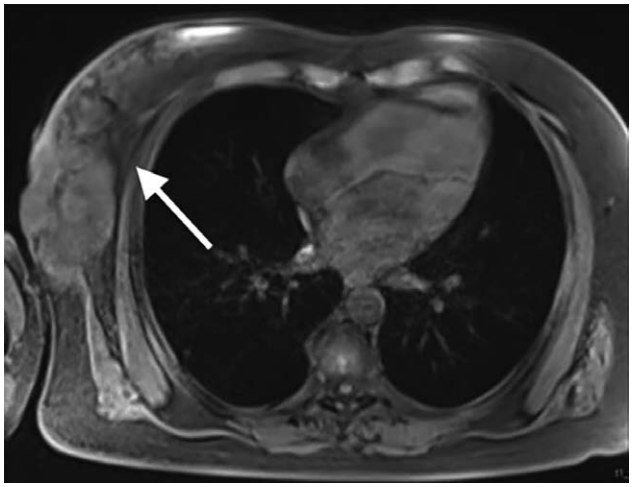

Fig. 2. MRI shows a well-defined soft tissue mass of the right anterolateral thorax (arrow) extending up to the fascia of pectoralis major muscle but without invading it.

behavior, adjuvant radiotherapy was recommended. At 2 years of follow-up and after one session of flap thinning, the patient is free of local and systemic disease and has a satisfactory aesthetic result (Fig. 4).

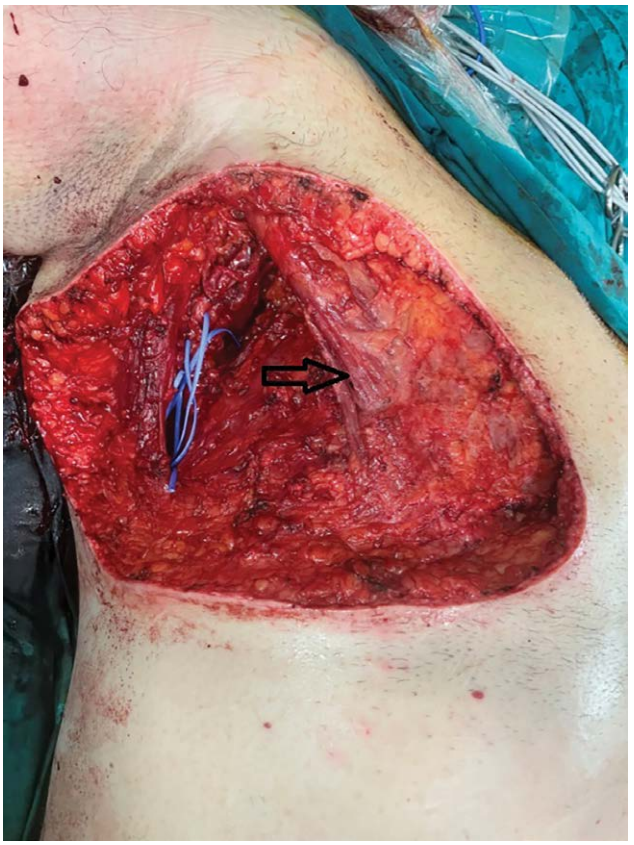

Fig. 3. Intraoperative view of the defect following wide surgical excision of the tumor and axillary lymphadenectomy of levels I and II. Pectoralis major muscle (arrow) and thoracodorsal vessels (blue vascular loops).

DISCUSSION

Pilomatrix carcinoma is an unusual follicular tumor with high recurrence rates. As pilomatrixomas commonly occur in the head and neck, they can easily be misdiagnosed as benign cysts, ossifying hematomas and lipomas.[6] On a recent literature review, out of 20 histologically confirmed malignant pilomatrixomas, none of them was clinically diagnosed preoperatively.[5] This fact emphasizes the reduced clinical suspicion that exists with this type of tumor. Subsequently, incorrect initial diagnosis may increase the likelihood of inadequate treatment.

Due to the rarity of this tumor, there is no consensus on its surgical management.[7] Surgical excision of malignant lesions with narrow margins has a 60% of recurrence rate,[5] and tumor recurrence has been associated with higher metastatic rates.[8] Due to the aggressive nature and the high incidence of recurrence, wide excision should be considered. The definition of "wide excision" varies considerably in the literature, with margins ranging from 4 mm to 3 cm.[5]In our case, excision with 2-cm peripheral margins and down to the muscle fascia was performed. Because of high metastatic incidence to the local lymph nodes and given the tumor's size, axillary lymphadenectomy was recommended by the oncological board.

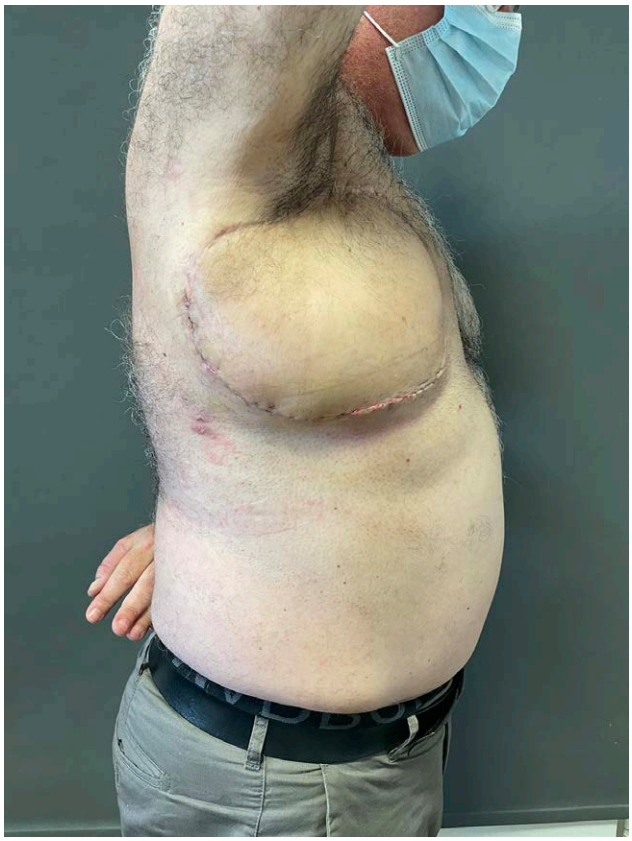

Fig. 4. Reconstruction with deep inferior epigastric artery perforator flap. Postoperative view, after flap thinning, at 2 years of follow-up.

Apart from recurrence, the size of the tumor has been considered as another aggravating factor for metastasis.[8] To date, approximately 140 cases of pilomatrixoma carcinoma have been described in the English-language literature, with a mean tumor size of 3.8 cm (ranging from 0.5 to 20 cm), and tumors with greater dimensions have been linked to an increased metastatic potential.[8] In our case, the measured dimensions of the tumor (22×28 cm) are the greatest ones reported in the literature. Despite the presence of two aggravating factors (size and recurrence), our patient, to date, remains free of metastatic disease.

The origin of this tumor, whether it arises de novo or degenerates from the benign epithelioma of Malherbe, has not been proved so far. Various reports refer to recurrent malignant tumors coming from previously excised lesions with benign characteristics. Sassmannshausan and Chaffins reported on a patient who underwent re-excision of a previously benign pilomatrixoma on the scalp, exhibiting histologically malignant degeneration.[9]In our patient, the lack of previous tissue biopsies made the origin of the tumor unclear.

Adjuvant radiotherapy following excision of primary and recurrent tumors has been reported to provide adequate local tumor control.[10] Radiotherapy is indicated for positive lymph nodes and for systematic metastasis as palliative treatment.[10] Despite our patient being free from metastatic disease, adjuvant radiotherapy was recommended due to the size of the tumor and the multiple local recurrences.

Reconstruction of the defect with a well-vascularized tissue was deemed necessary to offer a radioresistant tissue for the completion of radiotherapy. Because of its size, the defect could not be reconstructed with a locoregional flap. Given the patient's body type, a free fasciocutaneous flap, such as a deep inferior epigastric artery perforator flap, was a suitable option.

CONCLUSIONS

Malignant pilomatrixoma is an unusual and locally aggressive tumor that can initially be misdiagnosed. Surgeons usually remove pilomatrixomas, and only rarely have to deal with malignant transformations and recurrences of this typically benign lesion. However, if recurrence does occur, having knowledge that this could be a malignancy should always be suspected, so that further treatment would be justified. Limited surgical margins, local recurrences, and large tumor size are associated with higher metastasis incidence. A high index of suspicion is necessary, and oncological guidelines should be established for better prognosis.

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DISCLOSURE

The authors have no financial interest to declare in relation to the content of this article.

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