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Case Report

Pilomatrix carcinoma of the male breast: a case report

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Abstract: Pilomatrix carcinoma (PC) is a rare skin adnexal neoplasm derived from piliferous follicles, usually occurring in the head and neck region. An optimal treatment for PC has not yet been established, while surgery with wide margins is recommended, with radiation therapy (RT) and chemotherapy improving the curative effect. Location of this tumor in the breast is exceedingly rare, especially for a male. We now report an unusual case of a 53-year-old male who presented with 2 palpable masses in the right breast, which had been removed surgically about 4 and a half years prior, but recurred after 7 months. Ultrasonography revealed 2 irregular low-echo masses with a well-defined boundary, and computed tomography (CT) showed 2 soft tissue masses in subcutaneous tissue of the right breast. The tumors were completely removed without RT and chemotherapy. After about 4 years' follow-up, the patient remains free of local recurrence and metastasis. To the best of our knowledge, only 2 cases of PC in the breast region have been reported, but were female.

Keywords: Breast, male, pilomatrix carcinoma

Introduction

Pilomatrix carcinoma (PC) is a rare and malignant adnexal tumor of hair matrix origin, coming from pilomatrixoma [1, 2]. The tumor is a dermo-hypodermic in nature, with a low metastatic potential, but a high risk of recurrence following excision [2]. However, an optimal treatment regimen for the tumor has not yet been established. Pilomatrix carcinoma can have an intramammary location, but easily misdiagnosed as breast carcinoma on limited material. A high index of suspicion is required to arrive at an accurate diagnosis so as to obviate neoadjuvant chemotherapy. The present study reports the case of an older male who presented with a PC of the breast region and reviews the associated published literature.

Case report

A 53-year-old male first visited Gucheng County People's Hospital (Gucheng, Hubei, China) in 2015 due to a tenacious subcutaneous nodule that was about 2.0 cm in diameter on the right breast region. The nodule was surgically excised under local anesthesia with a pathologic diagnosis of pilomatrix carcinoma (**Figure 3**), but without follow-up treatment. About 7 months later, a tenacious mass with an unclear boundary reappeared at the previous site. A secondary excision was performed at the Department of Breast Surgery of Hubei Cancer Hospital (Wuhan, Hubei, China), and the histological evaluation revealed a pilomatrix carcinoma.

Upon physical examination, the tumors had the following characteristics: painless nodule, indurated, firm and immobile, clear border, with the sizes of 2.5×1.5 cm and 1.5×1.5 cm, respectively.

Breast ultrasound using high-resolution ultrasound scanners with high-frequency linear array transducer (LOGIQ-9, GE Medical Systems, USA) showed 2 irregular low-echo masses with a well-defined boundary, which were in the right breast at two o'clock (black arrow) and eight o'clock (white arrow) near the areola, respectively. The latter had minimal vascularity by power Doppler interrogation, while the former had relatively abundant blood flow signal (**Figure 1**).

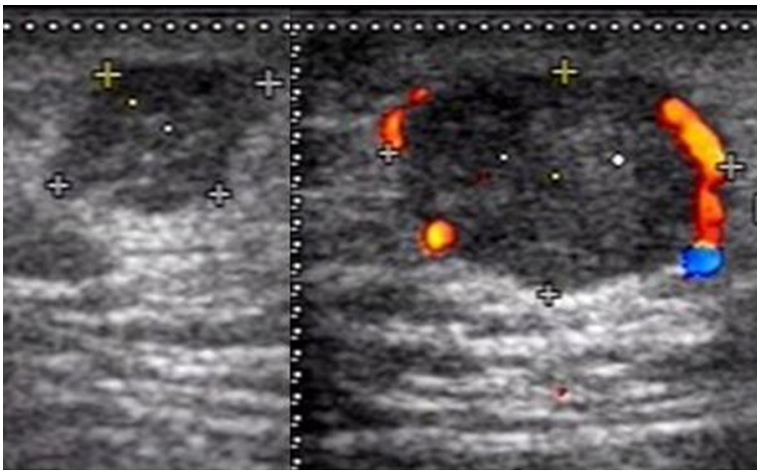

Figure 1. Ultrasound images showed 2 irregular low-echo masses with well-defined boundary, which were seen in the right breast at two o'clock (black arrow) and eight o'clock (white arrow) near the areola, respectively, while the former had relatively abundant blood flow signal.

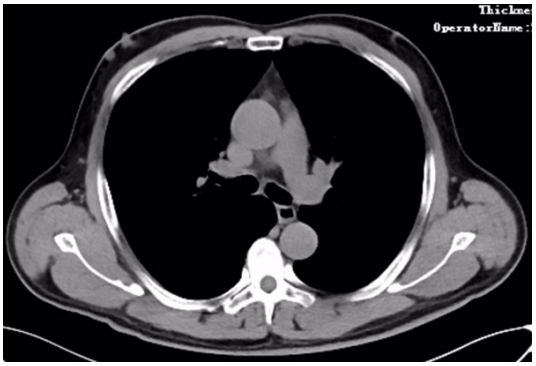

Figure 2. CT images revealed two soft tissue nodules (white arrow) in subcutaneous tissues of the right breast, which had a relatively clear boundary.

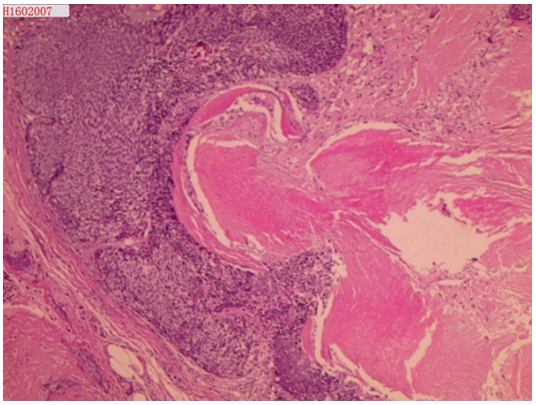

Figure 3. Pathologic image of pilomatrix carcinoma from Gucheng County People's Hospital. Eosinophilic shadow cells with a rim of peripheral basaloid cells infiltrating into the subcutaneous fat. Hematoxylin and eosin (H & E×100).

Chest CT (Lightspeed VCT-XT 64 Slice CT Scanner; GE Medical Systems, USA) was performed and revealed 2 soft tissue nodules in subcutaneous tissue of the right breast, with relatively clear boundary (**Figure 2**).

Clinical and imaging findings led us to the conclusion of local recurrence (Breast Imaging-Reporting And Data System category V). The differential diagnosis included pilomatricoma, invasive ductal carcinoma, basal cell carcinoma, or metastatic tumor.

The selected treatment strategy was resection of the whole tumor together with the right breast and lymph nodes in the ipsilateral axilla. The resected tumor was submitted for pathologic examination. Microscopic examination revealed that the tumor was composed of basaloid cells and paler shadow cells with prominent central necrosis and nuclear pleomorphism, with the positive expression of Ki-67 and P63 (**Figures 4**, **5**). A diagnosis of PC was therefore made.

In spite of the relapse, the patient received surgery, but refused radiation therapy to the right breast region. At about 4 years follow-up, the ultrasound and other clinical examination demonstrated that the patient remained free of local recurrence and metastasis.

The protocol for the present retrospective study was approved by the Ethics Committee of Hubei Cancer Hospital, and written informed consent was obtained from the study patient.

Discussion

Pilomatrixoma, first described as a "calcifying epithelioma" in 1880, is most frequently encountered in children and young patients, and is as a painless, slow-growing mass, usually located in the head or neck region, followed by the upper extremities, trunk, and lower extremities [3]. A malignant counterpart of pilomatrixoma, called "pilomatrix carcinoma" or "calcified epithelial carcinoma of Malherbe", comprised of actively proliferating, hyperchromatic, vesic-

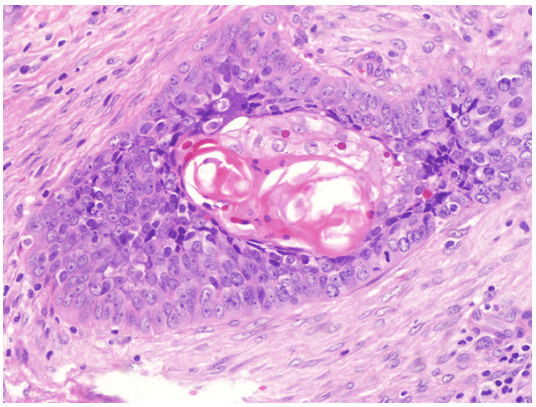

Figure 4. Postoperative pathologic result of pilomatrix carcinoma from Hubei Cancer Hospital. Higher magnification of the tumor showed the transition of basaloid cells into paler shadow cells (H & E×400).

ular basal cells with numerous mitoses, was first reported by Lopansri and Mihm in 1980 [4, 5]. Pilomatrix carcinoma has a bimodal distribution with most lesions occurring in the sixth and seventh decade [6-8], in which the male-to-female ratio is 1.8:1 [5]. PC is an exceedingly rare entity, while less than 200 cases of pilomatrix carcinoma have been described in literature to date, in which only an occasional case has been reported in the breast [1, 9, 10].

However, the origin of this tumor in the breast is still uncertain. Some studies mention its origin from the periareolar piliferous bulbs mimicking a breast malignancy [11]. The lesion might be caused by repeated skin trauma inciting an inflammatory response, whereas another hypothesis is that this tumor is hamartomatous [1]. The tumor may arise de novo as a solitary lesion, while many cases were arising in a pilomatrixoma or a previously excised pilomatrixoma, disclosing the malignant nature of the lesion, that was initially unsuspected [12].

To date, just two middle aged women with PC, which had an intramammary location, had been reported [1, 9]. The first one was a 57-year-old female patient who presented with a nodule measuring 2.5 cm in the left breast for a duration of 3 years, while the patient was alive without recurrence after nodulectomy [9]. The other woman, had a history of enlarging mass in the left breast for 2-year duration, measuring 15 cm, and the patient underwent modified radical mastectomy, followed by EBRT. That patient was also alive after the treatment completion without any history of recurrences [1]. The patient in our present study, had a history of nodulectomy in the right breast with a pathologic diagnosis of pilomatrix carcinoma, 4 and a half years prior. But 7 months later, the tumor recurred with 2 painless nodules, measuring 2.5×1.5 cm and 1.5×1.5 cm, respectively. The patient had modified radical mastectomy, but accompanied by no other therapy. Till now, the tumor has not recurred.

Clinically, PC typically manifests as a multicolored, asymptomatic cystic or solid mass, varying in size from 0.5-20.0 cm, presenting from several months to years prior to treatment [2]. The diagnosis of PC may be difficult, for a lack of clear histologic criteria to distinguish from other matrical tumors.

On histopathologic evaluation, features of pilomatrix carcinoma include sheets and islands of basaloid cells arranged in irregular nests throughout the tumor [13]. Asymmetric infiltrative expansion into subcutaneous tissue or muscle can be seen, as well as invasion into vascular or perineural structures [14]. PC tumors consist of pleomorphic basaloid cells with prominent nucleoli and frequent mitoses accompanied by central areas with keratotic material, shadow cells, and foci of necrosis [15]. The transition to squamous cells, clear cells, areas of necrosis, and frequent mitosis is associated with metrical cornification in PC [16]. The differential diagnoses of pilomatrix carcinoma include proliferating pilomatrixoma, aggressive pilomatrixoma, basal cell carcinoma with matrical differentiation, and squamous cell carcinoma.

Sonographically, PC presents as 2 well-defined, irregular low-echo superficial masses, as noted in our case (**Figure 1**), with few reports referring to ultrasound features of PC. Computed tomography (CT) scans show 2 soft tissue nodules in subcutaneous tissues of the right breast, as noted in our case [2, 17, 18] (**Figure 2**). PC in magnetic resonance imaging (MRI) may have some special features; however, the patient refused MR scan.

Although a standard treatment protocol for pilomatrix carcinoma is lacking, wide surgical excision of the primary lesion is the principal modality of treatment, and should be considered as the preferred option to limit tumor

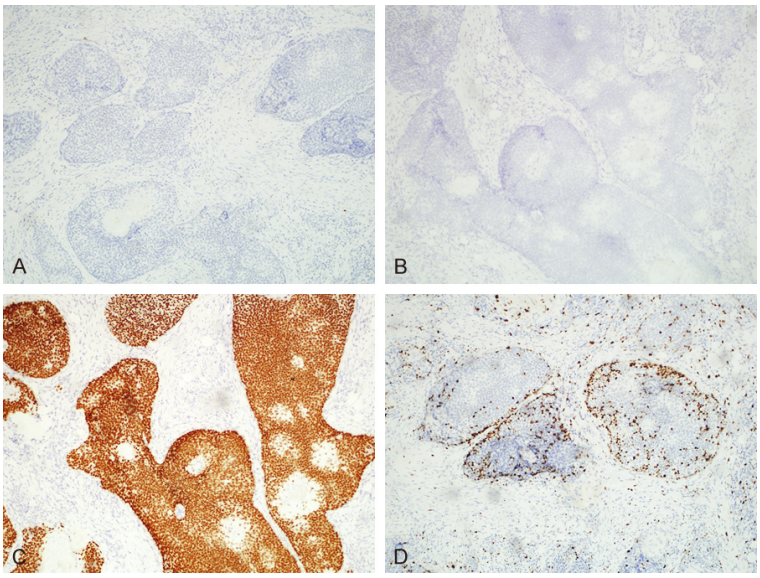

Figure 5. Immunohistochemical phenotype of pilomatrix carcinoma. A. Negative expression of CD15 in the tumor cells (SP, IHC×100). B. Negative expression of EMA in the tumor cells (SP, IHC×100). C. Positive expression of P63 in the tumor cells (SP, IHC×100). D. Ki-67 immunostain demonstrating high proliferation with Labeling Index of 20% (SP, IHC×100). IHC: immunohistochemistry.

recurrence. Radiation and chemotherapy have been used in cases of extensive local invasion or metastatic disease. However, the exact role of RT is unclear due to limited data. Systemic chemotherapy has also been largely unsuccessful [10]. Systemic disease is not responsive to chemotherapy, and is hence associated with a poor prognosis. The PC in the present case, after the patients underwent lumpectomy about 4 and a half years prior, recurred. Secondly he underwent radical mastectomy followed with no other therapy. At about 4 years' follow-up, the patient remained free of local recurrence and metastasis.

Conclusion

In conclusion, PC is extremely rare, especially presenting as an intramammary tumor in an old man. A high index of suspicion is required to arrive at an accurate diagnosis so as to determine an optimal treatment. Given that PC has high incidence of local recurrence, it is essential to have continued close followup of the patient.

Disclosure of conflict of interest

None.

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