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A case report and review of the literature

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Massimo Vergine*, Vittorio Pasta*, Adriano Redler*, Elisabetta Santucci*, Ilaria Vasselli*, Laura Ballesio**, Massimo Monti*

Università di Roma "Sapienza", Rome, Italy
**Dipartimento di Scienze Chirurgiche*
***Dipartimento di Scienze Radiologiche, Anatomopatologiche ed Oncologiche*

Cystosarcoma phylloides of the breast: a rare diagnosis. A case report and review of the literature

Sarcoma of the breast is a rare condition, with one possible meaning of relapse ad metastastasize, and behaves biologically as a stromal tumor.
Surgery is the first choise and mastectomy is the best procedure adopted in these tumors.
The aim of our study was to assess diagnostic and treatment options for PT, based on a review of the literature and our experience with a case of breast sarcoma arising from a phyllodes tumor.

KEY WORDS: Breast lesion, Cystosarcoma phyllodes, Phyllodes tumor.

Introduction

Cystosarcoma phyllodes (CP), otherwise known as phyllodes tumor (PT) of the breast, is a very rare but locally aggresssive fibroepithelial tumor in its malignant form and accounts for 0,3 – 1% of all breast neoplasis [1,2].
Phyllodes tumors (PT) were originally classified as benign breast lesions, but the current classification divides them into benign, borderline and malignant tumors (with the latter constituting 25-30% of PT). This classification is based on the degree of stromal cellularity and nuclear pleomorphism, the type of tumor margin (pushing or infiltrating) and the frequency of mitosis [3-4].
Histologically PT is made up of epithelial cells and connective tissue with greater stromal proliferation than a fibroadenoma, often with atypical cells, areas of necrosis, and hemorrhage. Metastases are rare, mostly hematogenous, and usually found in the lungs or bones. The local recurrence rate of PT is 20% [5]. Local excision is advisable for small lesions with a surgical margin of >1 cm, while for larger lesions, with multiple infiltrating foci, either mastectomy or wide quadrantectomy [6,7] should be performed. It is important to take into consideration the relationship between breast volume and tumor size.
It is doubtful whether radio- or chemotherapy is effective. In the literature the 5-year disease-free survival rate of patients with benign, borderline or malignant PT is reported to be 96%, 74%, and 66% respectively [8].
The aim of our study was to assess diagnostic and treatment options for PT, based on a review of the literature and our experience with a case of breast sarcoma, which, due to its histological characteristics could be considered one of the most malignant types of PT.

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Correspondence to: Dott. Massimo Vergine (e-mail: massimo.vergine@uniroma1.it)

Case report

A 27-year-old woman came to our attention a few months after an uncomplicated pregnancy and delivery, because of a swelling in her right breast, 8-10 cm in diameter, which developed when she began breast feeding.
Ultrasound examination (Fig. 1) had revealed a hypoechogenic highly vascularized lesion approximately 12 cm in diameter, with fluid-filled areas and unclear borders. As a result the patient underwent tru-cut biopsy followed by magnetic resonance imaging (MRI) (Fig. 2). A large high- intensity lesion with multiple cysts was observed on MRI. There were no other suspect areas in the parenchyma. Histological examination of the biopsy tissue suggested PT but the surgical specimen was required in order for a definitive diagnosis to be made. The patient therefore underwent wide surgical excision, and histological analysis revealed a high-grade sarcoma

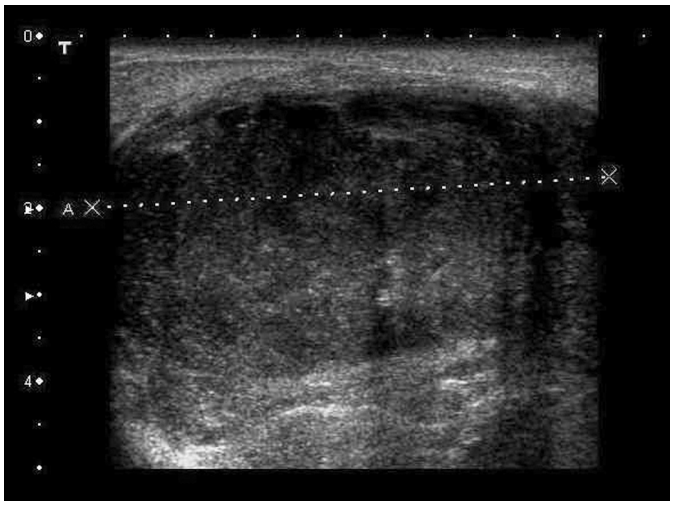

Fig. 1: Ultrasonography reveals a mass of 12 cm. with irregular border.

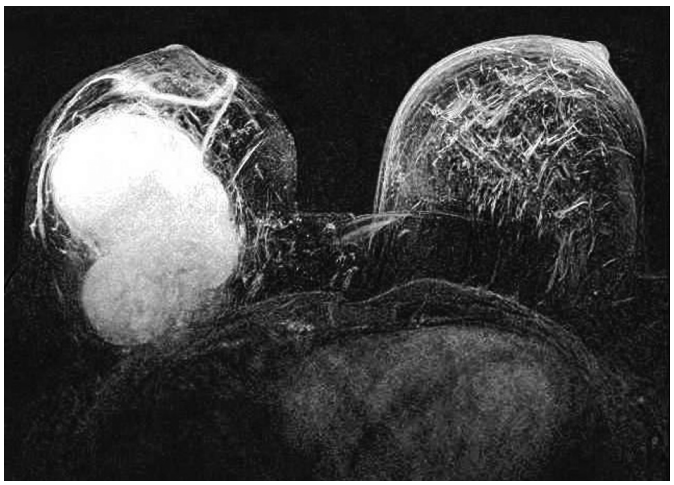

Fig. 2: MRI showing a 12 cm mass in the upper quadrants of the breast.

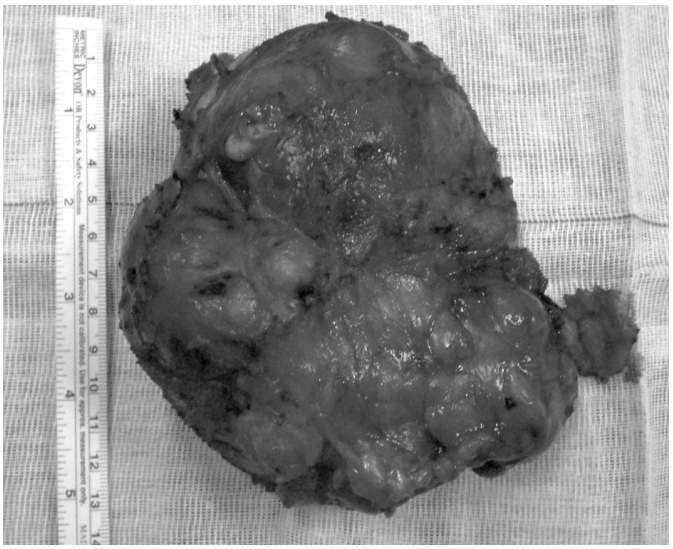

Fig. 3: Macroscopic view of the tumor.

that had arisen from PT with a high degree of proliferation (Ki-67:50%), and a high mitotic index (about 15 mitotic figures per high power field).
The lesion was encapsulated (Fig. 3) but there were some small satellite nodules close to the capsule. Because of the histology results the patient underwent compartmental mastectomy without axillary lymph node dissection, followed by radiotherapy.
The patient is disease-free one year after surgery.

Discussion

Since 1838 when Johannes Müller [9] first described cystosarcoma phyllodes (CP), it was found that the disease often pursues a benign course. Cystosarcoma phyllodes continues to be a challenge for both pathologists and surgeons due to difficulties regarding histologic diagnosis, classification, and clinical correlations which are closely linked to disease prognosis. According to some Authors, grading and adequate surgical excision with clear margins are the most important preventions of recurrence as well as of long-term prognosis since recurrent CP usually has a higher grade than the primary neoplasia [10]. Surgery is the best weapon to use against CP. Pharmacological treatment and radiotherapy have not been valuable effects [11]. The surgical approach consists of simple enucleation in cases of benign CP. There is no agreement regarding the management of borderline forms and malignant neoplasia, and both radical and conservative treatment have been recommended [12].
It has been shown that after simple excision the incidence of local recurrence in benign lesions, intermediate forms and malignant tumors is 21%, 46%, and 65%. respectively. However these numbers are 8%, 29% and 36% respectively after excision with wide margins [13]. Therefore some Authors feel that excision with 1-2 cm

clear margins is the ideal conservative operation for reducing the risk of local recurrence [14].
Axillary lymphadenopathy due to metastatic disease occurs in < 10% of patients [15]. In these cases lymphadenectomy can be considered but otherwise it is not routinely performed. Distant metastases are found in 25% of cases, mainly in the lungs (70-80%), pleura (60-70%), and bones (25-30%) [16-18].
There is uncertainty not only regarding treatment options for CP, due to intrinsic tumor variability and the relatively small number of cases in the literature, but also regarding prognosis given the low tumor sensitivity to chemo- or radiotherapy. To date, the only factors unanimously considered to have a prognostic value are recurrence and histological features: atypical nuclei, stromal cellularity, mitotic index, and anaplasia [19]. The available data does not clearly define the relationship between local recurrence and metastases. Some Authors claim to have demonstrated that there is a close relationship between recurrence and metastases while others maintain that the development of distant metastases can only be linked to tumor histology [20], especially to the ability of the tumor to invade the fibrous capsule by which it is often enveloped.

Conclusions

First-line treatment of breast sarcoma is surgical excision with adequate clear margins. Postoperative radiotherapy is associated with a small increase in survival. Routine axillary lymphadenectomy cannot be recommended because in cases of breast sarcoma there are almost never axillary metastases. Tumor response to chemotherapy is very limited. Patients with malignant CP often have local recurrence.
Histopathologic classification and the correct surgical approach seem to be the most important prognostic factors in CP of the breast.

Riassunto

Il cistosarcoma filloide è un raro tumore della mammella, con possibilità di recidivare e metastatizzare e si comporta biologicamente come una neoplasia stromale.
La chirurgia è la scelta di elezione e la mastectomia è la procedura migliore adottata in questi tumori.
Si presenta il caso di una giovane donna con un sarcoma della mammella ad alta malignità insorto su un tumore filloide, si descrivono le differenti ipotesi diagnostiche e le varie opzioni di trattamento di questo tumore, infine si fa una revisione della letteratura a riguardo.

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