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A rare case of true carcinosarcoma of the breast



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ABSTRACT

BACKGROUND: True carcinosarcoma of the breast is an extremely rare condition, accounting for 0.08–0.2% of all breast malignancies.

The correct definition of this tumor requires both a carcinomatous component and a malignant non-epithelial component of mesenchymal origin, without evidence of a transition zone between the two elements.

CASE PRESENTATION: We present a case of a 49-year-old woman presenting with a 4 cm mass at the level of her left breast upper-outer quadrant with a histologic diagnosis of true carcinosarcoma of the breast.

DISCUSSION: The most appropriate therapeutic regimens for breast carcinosarcoma are still unclear because of the rarity of this condition, but Breast Conserving Treatment (BCT) followed by adjuvant chemotherapy seems to provide a prognosis equalling that of usual Invasive Ductal Carcinoma of the breast.

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1. Background

True carcinosarcoma of the breast is an extremely rare breast condition, accounting for 0.08–0.2% of all breast malignancies [1].

The correct definition of this tumor requires both a carcinomatous component and a malignant non-epithelial component of mesenchymal origin, without evidence of a transition zone between the two elements [2,3].

There is controversy about the cells of origin for this neoplasm, but most research leads to believe the cells are of myoepithelial origin, a cell with potential biphasic differentiation [4].

True carcinosarcoma of the breast should be distinguished from metaplastic carcinoma, including spindle cell carcinoma, carcinoma with cartilaginous or osseous metaplasia, matrix producing carcinoma, malignant phyllodes tumor and other types of sarcoma [5,6].

The most important finding to differentiate metaplastic carcinoma from carcinosarcoma is whether a transition zone exists.

Carcinoma of the breast can undergo spindle-cell and other metaplasia, such as fibroblastic, chondroid, osseous or osteoblas-

tic. Although these metaplastic and infiltrative cancer cells form pseudosarcomatous stroma, as if carcinomatous components are mixed with sarcomatous components, a transition zone is always seen between these two components [7].

Most true carcinosarcoma of the breast shows no expression of estrogen and progesterone receptors and HER2-neu, with a so called “triple negative” pattern [8].

The 5-year survival rate of true carcinosarcoma is 49%, worst than other metaplastic breast cancers. Treatment strategies for true breast carcinosarcoma resemble those for usual breast cancer [9–11]. Carcinosarcoma metastasize by the lymphogenous route [3], therefore axillary examination with sentinel node biopsy and/or axillary dissection is suggested. This tumor also shows hematogenous metastases: pleural and pulmonary more commonly than skeletal, liver or brain metastases [10,11].

2. Case presentation

We report the case of a 49-year-old woman presenting a mass at the level of her left breast which rapidly grew in the period of 2 months.

Physical examination showed a firm mass measuring 4 cm in greatest dimension with irregular margins in the upper outer quad-

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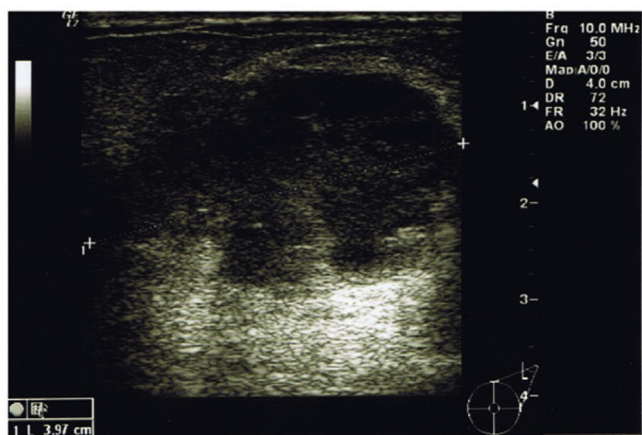


Fig. 1. Ultrasonography shows a 40 mm hypoechoic non homogenous mass with irregular margins.

rant of the left breast without enlarged palpable lymphnodes in the omolateral axilla.

Ultrasonography showed a solid hypoechoic 4 cm non homogenous mass with irregular margins (Fig. 1).

Mammography revealed a radiopaque lesion with irregular boundaries at the level of the left upper-outer quadrant (Fig. 2).

We performed a pre-operative Ultrasound-guided Fine-Needle Aspiration Cytology (US-FNAC) that showed hystiocytes and epithelial cells with moderate nuclear atypia surrounded by blood and necrotic cells, suggesting a surgical biopsy with intraoperative histological examination.

We performed an excisional biopsy of the mass with the histological diagnosis of carcinosarcoma (Fig. 3), followed by a wide excision (in relation to the favourable tumor/breast ratio) and sentinel lymphnode biopsy.

In gross description the mass measured 4 × 3 cm, revealing areas with epithelial and mesenchymal characteristics on histopathological examination.

Immunohistochemistry showed Ki67 proliferation index of 70–80% in both epithelial and mesenchimal component.

All tumor cells were negative for estrogen, progesterone and c-erb-B2.

Carcinomatous component was positive for low and high molecular weight cytocheratines (pancytocheratine AE1/AE3, CK 7, CK 8/18, CK 34 beta E 12).

Sarcomatous component was negative for cytocheratines and positive for vimentine, muscle-specific actine, p63 and focally for S-100; Vimentine and p63 were positive in the epithelial component as well. CD34 reveals a particular angiogenesis in the neoplasia.

No residual tumor was found in the excision area in the quadrantectomy specimen.

Histopathological examination of the sentinel lymph node was consistent with 'reactive hyperplasia'.

No evidence of distant disease was found at systemic imaging investigations.

The patient received adjuvant radiotherapy (total dose of 50 Gy) and chemotherapy with Anthracyclines and Taxanes. She is now in her 4th year of follow-up with no evidence of disease.

3. Discussion

The clinical and pathologic features of true carcinosarcoma of the breast are important to distinguish this rare condition from other types of uncommon breast malignancies such as spindle cell carcinoma, matrix producing carcinoma, fibrosarcoma, osteosarcoma, malignant fibrous histiocytooma, phylloides tumor

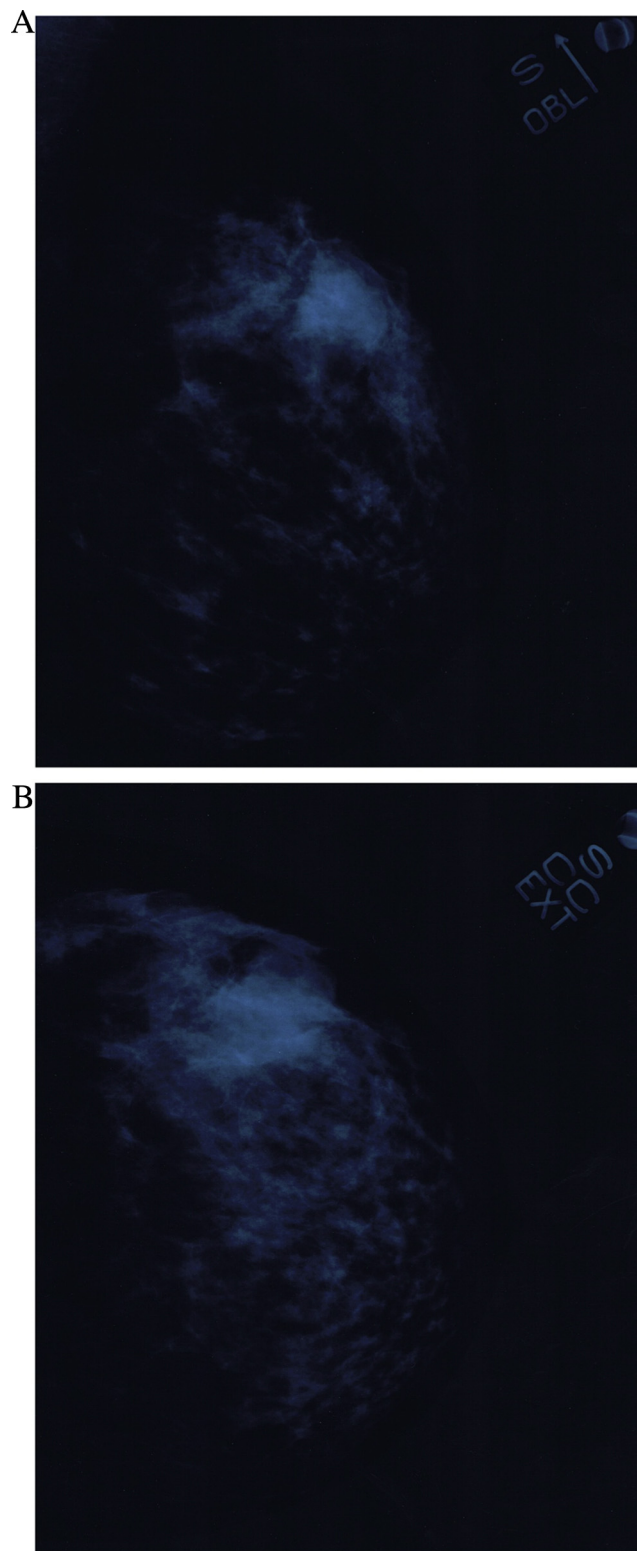


Fig. 2. (A and B) A high-density irregular mass in the left upper outer quadrant can be seen at mammography. (A) Oblique view; (B) Craniocaudal view.

and stromal sarcoma as their behavior, response to treatment and survival rates differ greatly [9].

Clinical features of breast carcinosarcoma are similar to those of invasive ductal carcinoma [10–12]. Recurrence can be rapid as the primary tumor is aggressive, thus mandating close interval follow-up after resection. Pulmonary metastasis is more common than

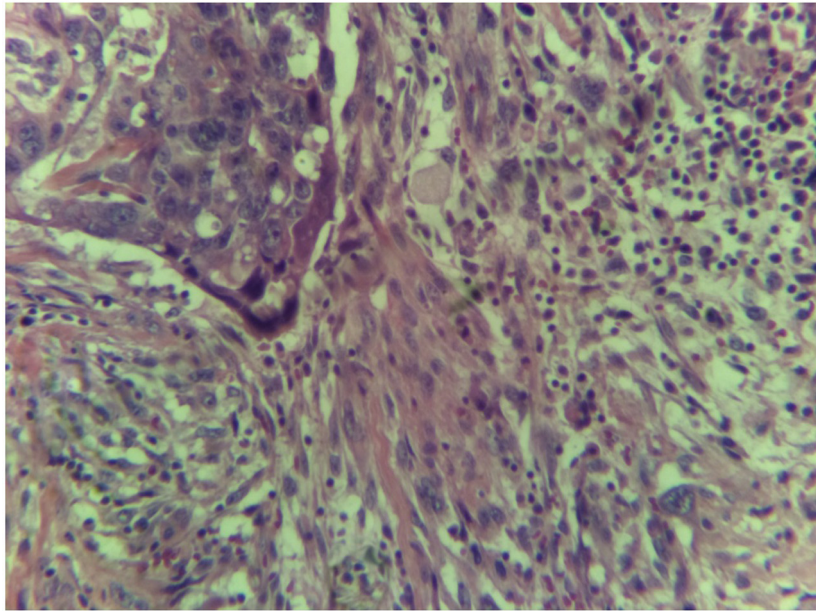


Fig. 3. Histopathology. The image shows large atypical epithelial cells and spindle atypical cells with storiform pattern.

brain, skeletal or hepatic metastasis, and the prognosis for these metastatic patients is poor.

Outcomes for local recurrences are somewhat improved when surgical resection is achievable. In general, the recommended treatment options have followed the established NCCN guidelines for patients with invasive breast cancer. In the majority of the reported cases, surgery with sentinel lymphnode biopsy or axillary node dissection was performed, followed by post-operative chemotherapy and radiation therapy in various combinations. Evaluation of patients with breast carcinosarcomas includes analysis of the expression of various receptors on the primary tumor.

The HER1/EGFR receptor is reported to be over-expressed in the majority of carcinosarcomas of the breast. The results may then be utilized to tailor the adjuvant therapy based upon these findings.

New treatment opportunities may exist with the development of agents targeting the EGFR receptor such as Gefitinib (ZD1839, Iressa) and Cetuximab (Erbitux) [13,14].

Obtaining an accurate diagnosis of true carcinosarcoma is essential in order to optimally tailor adjuvant therapy towards this aggressive breast cancer subtype. This requires an interdisciplinary approach to treatment, with the involvement of the entire oncology team comprised of the primary care physician, the pathologists, surgical oncologist, radiation oncologist, medical oncologist, and the supporting nursing and research staff. Adjuvant chemotherapy is generally recommended utilizing established NCCN guidelines for the more common types of breast cancer.

By improving our understanding of true carcinosarcoma of the breast, we may provide such patients with novel and potentially effective treatment options that will ultimately translate into improved overall outcomes.

Conflict of interest

No conflicts of interest to declare.

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Ethical approval

Ethical approval no needed.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author contribution

All authors contributed significantly to the present research and reviewed the entire manuscript.

AA: Participated substantially in conception, design, and execution of the study and in the analysis and interpretation of data; also participated substantially in the drafting and editing of the manuscript.

GC: Participated substantially in the drafting and editing of the manuscript. GADC: Participated substantially in conception and design of the study.

PR: Participated substantially in conception and design of the study.

GA: Participated substantially in collecting data. CS: Participated substantially in design of the study.

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GF: Participated substantially in design of the study.

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Guarantor

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