

A Rare Case of Breast Carcinosarcoma with Lymphatic Metastasis

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Introduction and Patient Profile: *Carcinosarcoma of the breast is a rare malignancy composed of two cell lines described as a ductal-type carcinoma with a sarcoma-like component. It is an aggressive neoplasm that is usually larger in size than epithelial breast cancers and characterized by a rapid increase in size. A 32-year-old woman presented with a palpable lump in the left upper outer breast. Imaging findings and an ultrasound-guided biopsy demonstrated a malignant neoplasm with chondroid differentiation.*

Interventions and Outcomes: *The patient underwent a modified radical left breast mastectomy with sentinel node biopsy. Pathology report from the mastectomy demonstrated an infiltrating metaplastic carcinoma (MPC) with positive lymph nodes.*

Discussion: *The most unusual feature of this case is the lymph node positivity, as lymphatic spread is uncommonly associated with carcinosarcoma or any subtype of metaplastic carcinoma of the breast. This case is important because it illustrates the potential future need for treatment guidelines for this uncommon tumor.*

Keywords: *breast cancer; carcinosarcoma; clinical protocols; treatment protocols; lymphatic metastasis.*

INTRODUCTION AND PATIENT PROFILE

Carcinosarcoma of the breast, one of five subtypes of metaplastic carcinoma (MPC), is an aggressive neoplasm composed of two cell lines described as a ductal-type carcinoma with a sarcoma-like component. It is an aggressive neoplasm that is usually larger in size than epithelial breast cancers and characterized by a rapid increase in size.¹ It has been reported to account for 0.08–0.2% of all breast malignancies.² Breast malignancies affect 12.3% of women at some point during their lifetime, with an estimated incidence of 232,340 new cases diagnosed in 2013.³ MPC of the breast most often presents in women more than 50 years of age as a unilateral, well-defined, large, and painless mass within the breast. Typically, MPCs of the breast do not express the estrogen or progesterone receptors and do not over-express the HER2/neu oncogene.⁴ This 'triple-negative' phenotype tends toward more aggressive tumors that are unlikely to respond to targeted therapy with drugs such as trastuzumab (Herceptin) or estrogen/progesterone receptor antagonists. Prognosis for MPC is less favorable compared with invasive ductal or invasive lobular carcinoma.¹ Predictors of poor outcome include age younger than 39 years at presentation, skin invasion, and size greater than 5 cm, while the subtype of MPC has not been shown to affect outcome.⁵ Five-year

overall survival for carcinosarcoma of the breast is 49–68%.²

A 32-year-old woman with a medical history of congenital heart disease with double outlet right ventricle status post-repair presented with a palpable lump in the left upper outer breast. The patient had been breastfeeding for the past 2 years and presumed that the lump was related to lactation. No palpable lymphadenopathy was noted on clinical exam.

The patient was sent to radiology by a breast surgeon for initial imaging, which included a bilateral mammogram and a left breast ultrasound examination. An approximately 6 cm mildly lobulated mass was present in the upper outer quadrant left breast posterior depth (Fig. 1). No additional abnormalities were noted on the mammogram. A focused ultrasound examination was performed at the area of concern. On the ultrasound examination, a large lobulated cystic and solid mass was visualized with associated vascularity, corresponding to the patient's palpable concern. Imaging findings were reported as suspicious for carcinoma and an ultrasound-guided biopsy was recommended.

Left breast core biopsy was obtained, which demonstrated a malignant neoplasm with chondroid differentiation (Fig. 2). The differential diagnosis was reported as phyllodes tumor, MPC, and chondrosarcoma.

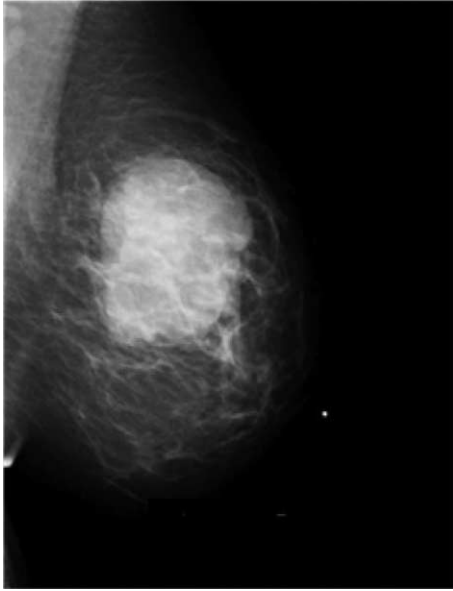


Figure 1. Left mediolateral oblique (MLO) mammogram image: 6-cm mildly lobulated mass in the upper outer quadrant left breast posterior depth.

A bilateral breast MRI examination was performed for treatment planning. The MRI exam demonstrated a large predominantly cystic complex mass with thick

enhancing septations and a thick enhancing peripheral rim corresponding to the patient's known carcinoma (Fig. 3). The mass was noted to involve a large portion of the lateral aspect of the left breast. Edema and inflammatory changes associated with this mass were noted to extend to the chest wall with no definite chest wall involvement. No additional abnormally enhancing lesions were noted in either breast that would be suggestive of malignancy. No abnormally enlarged lymph nodes were noted on the MRI exam.

A PET CT was performed for staging. A large irregularly contoured mass was noted with intense fluorodeoxyglucose (FDG) avidity peripherally with a standard uptake value (SUV) of 13.7 in the left breast. Additionally, an FDG avid left internal mammary lymph node was noted measuring 13 × 7 mm with an SUV of 4.2. No additional foci of FDG avidity were noted to suggest metastasis.

INTERVENTIONS AND OUTCOMES

The patient underwent a modified radical left breast mastectomy with sentinel lymph node biopsy. Pathology report from the mastectomy demonstrated an infiltrating MPC with predominant cartilaginous differentiation. Foci of high-grade ductal carcinoma *in*

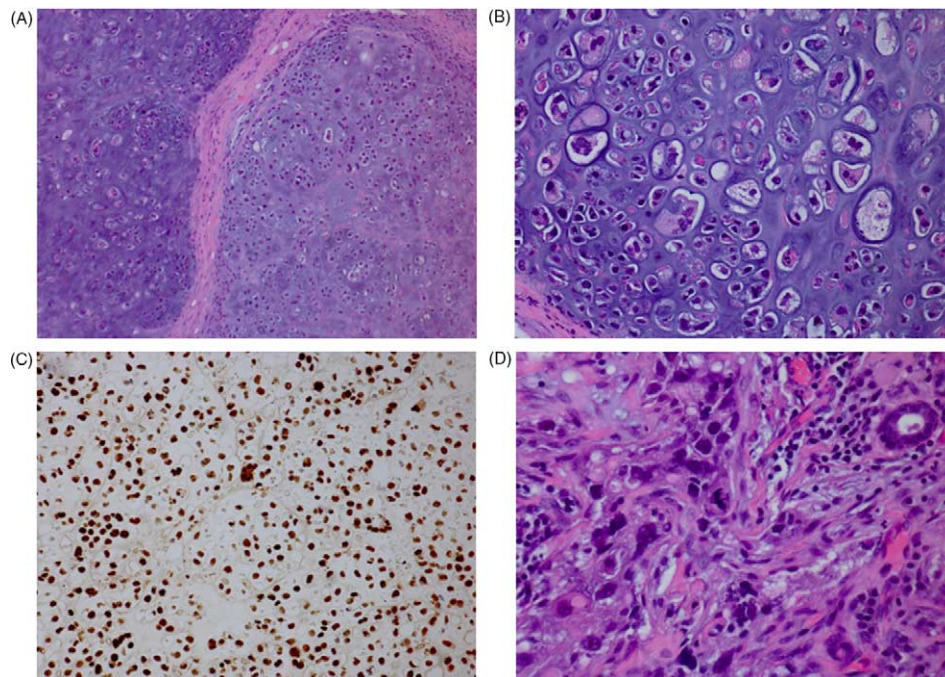


Figure 2. (A) Cartilaginous differentiation in the tumor (hematoxylin and eosin (HE) × 100x). (B) Markedly atypical neoplastic cells lying in lacunae (HE × 200x). (C) The atypical cells express S100, consistent with cartilaginous differentiation (3,3'-diaminobenzidine × 100x). (D) Background neoplastic cells with severe atypia are identified infiltrating the stroma as individual cells (HE × 200x).

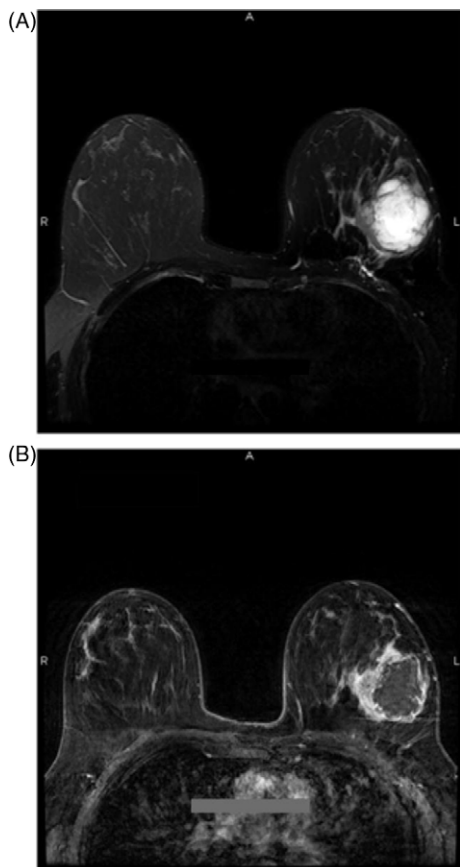


Figure 3. (A) MRI T2-weighted image shows a large circumscribed mass with internal high-signal intensity and edema and inflammatory changes noted to extend to the chest wall. (B) MRI post contrast T1-weighted image shows a large predominantly cystic complex mass with thick enhancing septations and thick enhancing peripheral rim.

situ with comedonecrosis and dense chronic inflammation were seen surrounding the tumor. Tumor size was noted to be 10.5 cm in maximum gross dimension on pathology. The tumor was described as a mixed epithelial and mesenchymal MPC with 70% chondrosarcoma and 30% ductal carcinoma. Overall Scarf Bloom Richardson histologic grade was 9/9 with a grade of 3 in tubule formation, nuclear grade, and mitotic count. No lymphatic, vascular or perineural invasion was identified. The tumor was weakly estrogen receptor positive and progesterone receptor negative. Expression of the HER2/neu oncogene was negative.

A sentinel lymph node biopsy was done which showed two out of five lymph nodes positive for metastatic carcinoma with metastatic foci measuring 1.2 and 2.6 mm. A completion axillary dissection revealed

12 additional lymph nodes, which did not harbor any metastatic carcinoma. The tumor was staged as p T3, p N1, M0, stage IIIA.

The patient received chemotherapy for 6 months after diagnosis and did not receive radiation therapy. A subsequent PET scan demonstrated no evidence of FDG avid malignancy.

DISCUSSION

Metaplastic breast cancer is characterized as an unusual, uncommon tumor that is comprised of malignant epithelial tissue (carcinoma) mixed with malignant mesenchymal cells (sarcoma). Most metaplastic tumors of the breast are poorly differentiated, high grade, and of triple-negative receptor status.² The patient presented with a large, rapidly-dividing tumor that was found to be of high histologic grade as well as weakly estrogen receptor positive, in contrast to the triple-negative phenotype of most MPCs. The most unusual feature of this case is the lymph node positivity, as lymphatic spread is uncommonly associated with carcinosarcoma or any subtype of MPC of the breast. However, there is a high hematogenous metastatic potential to extranodal sites such as lung and bone.⁴

Clinical features of metaplastic breast cancer and carcinosarcoma are similar to those seen in patients with invasive ductal carcinoma. Obtaining an accurate diagnosis is imperative in order to optimally tailor adjuvant therapy and improve survival and patient outcomes.⁴

Imaging features of MPC have been reported as benign on mammograms with round or oval shapes and circumscribed margins. The lesions are often noncalcified with a high rate of architectural distortion.⁴ Our patient presented with an oval-shaped 6 cm mildly lobulated mass in the upper outer quadrant left breast posterior depth, consistent with a poor prognosis based on size greater than 5 cm. On sonogram, MPCs are generally described as a heterogeneous or hypoechoic solid mass. They may also be described as a mixed cystic and solid mass with a round, oval or lobular shape, and most frequently demonstrate posterior acoustic enhancement (compared with posterior shadowing).⁶ The patient's mass was described as a large lobulated cystic and solid mass with associated vascularity, also consistent with known imaging features of MPC. The MRI exam findings were also found to be characteristic of typical MPC findings, which include T2 hyperintensity and lesions containing ring-like, homogenous, heterogeneous, or nonenhancing internal components.⁴

These nonenhancing T2 high-signal-intensity internal components correlate with necrosis, cyst, and chondroid matrix on pathology exam findings.⁶

Evaluation of MPC includes analysis of receptor expression on the primary tumor, as well as close interval follow-up after resection for metastasis due to a high rate of extranodal spread. Currently, there are no standard guidelines for the treatment of MPC. The recommended treatment has followed the NCCN guidelines for patients with invasive breast cancer. Most patients undergo modified radical or radical mastectomy followed by adjuvant chemotherapy and/or radiation therapy.⁵ Mastectomy without axillary lymph node dissection is considered standard treatment for women with breast carcinosarcoma because lymphatic spread is uncommon in this type of malignancy.⁷ This case is important because it identifies the potential for lymphatic spread of MPC in a young patient and illustrates the potential future need for treatment guidelines for this uncommon tumor.

LEARNING POINTS

1. Most metaplastic tumors of the breast are poorly differentiated, high grade, and of triple-negative receptor status.
2. Clinical features of metaplastic breast cancer and carcinosarcoma are similar to those seen in patients with invasive ductal carcinoma.
3. The most unusual feature of this case is the lymph node positivity, as lymphatic spread is uncommonly associated with carcinosarcoma or any subtype of MPC of the breast.

4. Mastectomy without axillary lymph node dissection is considered a standard treatment for women with breast carcinosarcoma because lymphatic spread is uncommon.

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