



## Rare Primary Sarcoma of the Breast in a 54-year-old Female: A Case Report

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### Abstract

Primary breast sarcoma is a rare breast malignancy that can present as a palpable nodule with rapid increase in size. Given its rarity and similar presentation to that of the ominous metaplastic carcinoma, diagnosis may not be made until thorough histopathologic evaluation is performed. Here we present the case of a 54-year-old female who was found to have primary breast sarcoma at the time of tissue diagnosis.

### Introduction

Breast sarcomas are rare tumors that can occur as primary tumors of the breast, post-radiation therapy or from lymphedema of the arm or breast following treatment of another malignancy. They account for less than 1% of all breast malignancies and less than 5% of all soft tissue sarcomas [1,2]. Typical symptoms at time of presentation include a unilateral, well-defined, large, painless, firm mass within the breast, with rapid increases in size. Imaging findings can be non-specific and differentiating between epithelial and non-epithelial origin tumors can be difficult, posing a challenge for management sans tissue diagnosis [3].

### Case Presentation

A 54-year-old woman presented to the oncology clinic with a chief complaint of a nodule in her left breast. Upon review of her history, she had initially felt a small nodule in her left breast four months prior to presentation, however had waited initial evaluation until a month prior with her scheduled well woman visit. She had noted that the nodule was increasing in size and pain over the span of several weeks. A diagnostic mammogram demonstrated a 4.6 cm lobulated left lower inner quadrant mass anteriorly with a 1.8 cm left axillary lymph node (Figure 1). Core needle biopsy was performed of the mass and revealed a poorly differentiated malignant neoplasm with osteoclast-like giant cells, CD 68 positive, E-Cadherin negative, GATA negative, keratin negative, Ki 67 high proliferative index, and vimentin positive. These findings were suspicious for metaplastic carcinoma, warranting additional work-up. Upon presentation to our clinic, exam had demonstrated a palpable left lower outer quadrant mass, firm to hard in consistency with questionable left axillary and left supraclavicular lymphadenopathy. Left axillary lymph node core needle biopsy was performed and negative for tumor. Following this, bilateral skin-sparing mastectomy was implemented and left breast pathology was significant for a 5.9 cm high grade sarcoma with giant cells that was negative for tumor within the lymph nodes (Figure 2). The findings were discussed with the patient and she was provided treatment options in regards to observations vs. radiation therapy for prevention of recurrence.

### Discussion

The patient's definitive diagnosis was made with biopsy of the breast mass. With similar presentation of that of an epithelial cell tumor and no previous history of malignancy or radiation therapy the suspicion for metaplastic carcinoma was present and diagnosis of primary sarcoma was unexpected. Primary breast sarcomas are rare occurring in 4.6 cases per million women annually [4]. Risk factors include ionizing radiation exposure, chronic lymphedema, in addition to inherited causes such as Li Fraumeni, neurofibromatosis 1, and familial adenomatous polyposis. Various environmental factors can play a role in development including exposure to HHV-8, HIV, herbicides, arsenic compounds and vinyl chloride [5,6]. The patient did not have any identifiable risk factors suggesting this was a primary sarcoma in nature. Clinically, breast sarcomas are larger than carcinomas with median size of 5 cm to 6 cm, unilateral, firm and painless and generally have a rapid increase in size [7]. Imaging is non-diagnostic; however mammography can demonstrate

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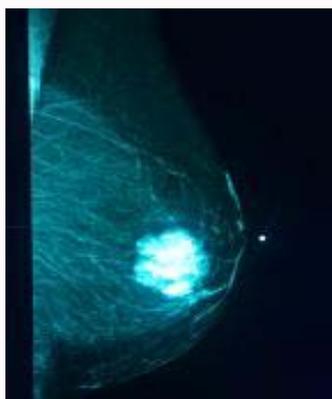
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**Figure 1:** Mammogram of left breast demonstrating 4.6 cm lobulated mass of the anterior left lower inner quadrant.

a non-calcified oval mass with indistinct margins, similar to that of metaplastic carcinoma. Axillary lymph node involvement is typically seen with metaplastic carcinoma, but is unusual for breast sarcomas. Breast sarcomas have been shown to metastasize to lungs and pleura, liver, bone and superficial soft tissues [7,8]. Laboratory studies have not been found to be helpful with diagnosis. Biopsy is recommended for diagnosis, whether this be performed *via* incisional, excisional or core-needle, however fine needle aspiration is not recommended as it has a low diagnostic accuracy for sarcoma, and histologic subtype or grade are unable to be determined [7]. Treatment is based on staging present at the time of initial diagnosis, and given that sarcomas are spread hematogenously, lymph node involvement does not play a role in treatment or staging [9]. Surgical management without need for local axillary node dissection using wide local excision with negative margins or mastectomy are primary treatment modalities [10-12]. Adjuvant radiation therapy is subsequently used to treat micrometastases, however there is a limited role for chemotherapy. Local recurrence is not uncommon and typically is associated with tumors greater than 5 cm [3,12].

## Summary Points

Carcinoma, especially metaplastic carcinoma, must be ruled out prior to the diagnosis of primary breast sarcoma. A breast nodule that is rapidly increasing in size should prompt consideration of possible primary breast sarcoma. Surgery is a valuable treatment and diagnostic method that patients should undergo to insure reliable management. Ideally, negative breast margins should be obtained and radiation therapy should be considered, especially in patients with axillary lymph node involvement. The potential for local recurrence should be considered when discussing treatment options with patients, especially with larger tumors.



**Figure 2:** Histopathologic specimen of left breast mass demonstrating high grade sarcoma with the presence of giant cells.

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