

Case Report

Primary Breast Sarcoma: A Case Report

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Abstract

Introduction: Breast sarcomas are very rare, accounting for less than 1% of all breast malignancies and less than 5% of all soft tissue sarcomas [1,2]. They can develop as primary malignancies or secondary to radiation therapy or after treatment of another malignancy [3,4]. Primary breast sarcomas are particularly rare, accounting for only 44.8 new cases per 10 million women [5]. Because breast sarcomas are so rare, clinical research is difficult and there is little data to guide management. Most published data on breast sarcomas is limited to small retrospective case reviews and case reports.

Case presentation: A 65 year-old woman presented with a three week history of swelling and bleeding of her left breast. On initial exam, there was an ulcerated, exophytic multinodular mass of the inferior, medial aspect of the right breast measuring approximately 18 cm. Ultrasound guided biopsy was performed which revealed the mass was a high grade pleomorphic breast sarcoma. The patient underwent a right mastectomy. Three months later, the patient re-presented with a bleeding lesion on her scalp. Fine needle aspiration was performed, which revealed a high grade, poorly differentiated malignancy.

Conclusion: This case represents a very rare form of breast sarcoma and could potentially contribute to the body of scientific evidence on breast sarcomas. A centralized registry might be able to compile this and other evidence for further analysis to better understand breast sarcomas and guide management.

Keywords: Breast cancer; breast sarcoma

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Consent: Consent was taken from the patient for publication of this case report.

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Introduction

Breast sarcomas are very rare, accounting for less than 1% of all breast malignancies and less than 5% of all soft tissue sarcomas [1,2]. They can develop as primary malignancies or secondary to radiation therapy or after treatment of another malignancy [3,4]. Primary breast sarcomas are particularly rare, accounting for only 44.8 new cases per 10 million women [5]. Because breast sarcomas are so rare, clinical research is difficult and there is little data to guide management. Most published data on breast sarcomas is limited to small retrospective case reviews and case reports. Reported here is a case of primary breast sarcoma in a 65 year-old woman with metastatic disease.

Case presentation

A 65 year-old female was seen at the outpatient clinic complaining of bleeding and swelling of her right breast for about three weeks. The patient had fallen and injured her right breast and side in December 2014, but did not note any bleeding until three weeks prior to presenting to the clinic. On physical exam, there was an ulcerated, exophytic multinodular mass of the inferior, medial aspect of the right breast measuring approximately 18 cm (Figure 1).



Figure 1 Right breast mass on presentation.

The mass was noted to be largely necrotic and malodorous with exudation. Bilateral diagnostic mammogram performed in March 2015 demonstrated a 15 x 9 cm irregular high-density mass with a macrolobulated margin and heterogeneous calcifications. Targeted ultrasound showed an irregular mass comprised of multiple nodules with increased vascularity demonstrated on color flow imaging. Ultrasound guided biopsy was performed which revealed the mass was a high grade pleomorphic breast sarcoma. Metastatic work up was deferred due to significant renal impairment which did not respond to resuscitation, owing to the large necrotic focus. The patient underwent a right mastectomy. The mass was 14.5 x 11 cm on gross examination. Microscopic examination showed high-grade pleomorphic sarcoma arising from phyllodes tumor resected with clear margins; with the closest margin being 3mm (Figure 2).

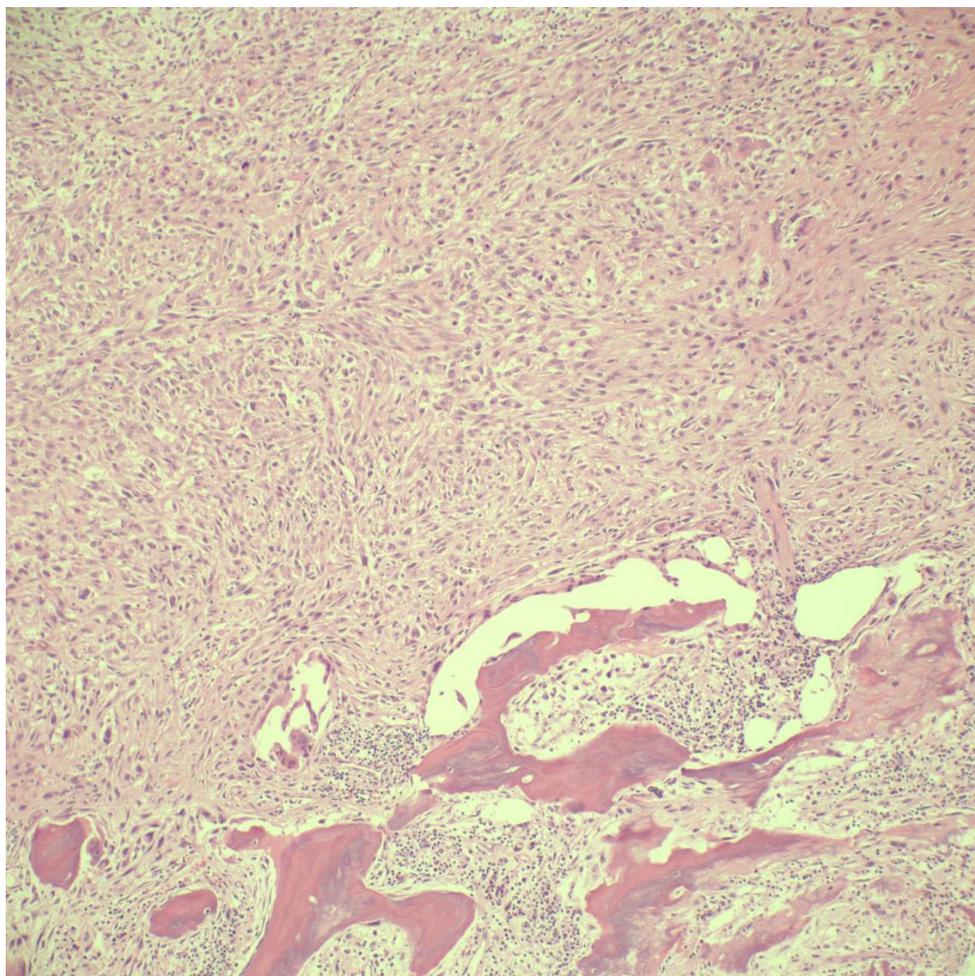


Figure 2 High grade pleomorphic sarcoma arising from phyllodes tumor.

After surgery, her renal function improved and she underwent a full body CT scan, under acetylcysteine for nephroprotection. The CT scan revealed two pulmonary nodules, the largest measuring 1.3 cm on the left side. The patient was discharged on post-operative day three and followed up in clinic on post-operative day 12. Three months later, the patient re-presented with a palpable right chest wall mass, as well as a bleeding lesion on her scalp. Fine needle aspiration was performed on both, which revealed a high grade, poorly differentiated malignancy from the scalp lesion; the chest wall lesion was consistent with fat necrosis. She then received adjuvant radiation and chemotherapy.

Discussion

Breast sarcomas are rare malignancies that arise from the connective tissue within the breast [6]. These sarcomas can be primary, or they can develop as secondary, therapy-related malignancies. Breast sarcomas usually present as unilateral, well-defined, large, painless masses. They tend to be larger in

size and are often characterized by a rapid increase in size [4,7]. Primary sarcomas also tend to arise in younger women (average age under 40), while secondary sarcomas are more often seen in older patients (average age 64) [5,8].

Overall, the annual incidence of breast sarcomas is 4.6 cases per million women, according to data compiled from the Surveillance, Epidemiology, and End Results (SEER) Program at the National Cancer Institute [7]. The exact incidence of secondary, therapy-related breast sarcoma is difficult to distinguish. Secondary breast sarcomas have been shown to be associated with prior radiation therapy and conditions causing chronic lymphedema [3,4,9]. Although the absolute risk is small, secondary breast sarcomas most often arise following radiation therapy for a previous breast cancer [3,10]. However, breast sarcomas have been shown to develop after radiation therapy for other malignancies when the chest wall or breast is included in the radiation field [11]. Chronic lymphedema of the chest, axilla, or upper extremity, typically following treatment for breast cancer, is another risk factor for the development of secondary breast sarcoma, particularly angiosarcoma [12]. Primary breast sarcomas are exceedingly rare, accounting for less than 50 cases per 10 million women [5]. In contrast to therapy-related breast sarcomas, risk factors for developing a primary breast sarcoma are largely unknown, and a specific causative factor is difficult to detect in many patients [13]. Breast sarcomas most commonly metastasize by direct local invasion. They can also spread hematogenously, and, rarely, through regional lymph nodes [7].

Due to the rarity of breast sarcomas, there is little research to guide management. Surgery is the mainstay of treatment of breast sarcomas, with adequate resection margin being the single most important determinant of long-term survival [7,14]. The use of adjuvant radiation therapy for primary breast sarcomas postoperatively is controversial. Radiation therapy has been shown to reduce the rate of local recurrences, however, the effect on long-term survival is unclear [1,7,13].

Conclusion

This case represents a very rare form of breast sarcoma. Because breast sarcomas are so uncommon, there is little data to direct management. This case potentially adds information to the scientific evidence that currently exists on breast sarcomas. A centralized registry might be beneficial to compile this and other scientific evidence for further analysis to better understand and guide management of this rare form of breast cancer.

Consent

This patient has given informed consent for the case report to be published.

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