

# Primary breast sarcoma: A rare case report

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## Abstract

**Background:** Primary breast sarcoma (PBS) is an extremely rare breast malignancy, constituting less than 1% of total breast malignancies. There is no definitive consensus regarding PBS treatment and current recommendations derived from retrospective case reviews and extrapolated from non breast soft tissue sarcoma studies. Complete surgical resection with negative margin (R0) is recommended for curative intent. However there is debate between breast conservation surgery and mastectomy. Role of radiotherapy and chemotherapy in metastatic PBS is unclear. Primary objective of this study is to determine clinico-pathological characteristics of primary breast sarcoma and to identify patient, pathological and treatment characteristics that predict survival outcome. **Case history:** A 35yr woman presented with complain of palpable lump in her right breast. FNAC from the mass suggested malignancy. Metastatic work up showed no evidence of metastasis. She underwent modified radical mastectomy (MRM) and postoperative histopathology suggested primary sarcoma breast. This was followed by EBRT to the chest wall. Now she is asymptomatic and on regular follow up since last 5 years. **Conclusion:** optimal treatment methodology in PBS needs to established

**Key Word:** FNAC, surgery, radiotherapy

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because of the low incidence of primary breast sarcomas with few cases being reported.

## CASE REPORT

A 35 year old woman presented in our hospital with complain of palpable lump in her right breast. Her personal history did not feature other important details. The routine blood investigation revealed Hb- 13g/dl, TLC- 7100, Platelets-390,000 lacs, Urea- 23, Cr- 0.6, serum bilirubine-0.4mg/dl. Tumour marker CA 15.3 was 6U/ml. The examination revealed a 7×7 cm hard lump in right outer upper quadrant, mobile over chest wall and overlying skin was free with no axillary or supraclavicular lymphnode. FNAC from mass was suggestive of neoplasm. The CECT chest (figure.1) showed 8×6 cm mass with heterogenous enhancement with intravenous contrast with no signs of necrosis and haemorrhage in it. There were no evidence of nodal involvement or distant metastasis. Due to the local extent of the disease we proceeded for modified radical mastectomy with axillary clearance 10th august 2013. The postoperative histopathology was suggestive of sarcoma breast(nos)(figure. 2), skin and margins were free and none out of 10 lymph nodes were involved. The pathological stage was pT3pn0. Immunohistochemistry showed positivity for vimentin (figure.3). The Patient was

## INTRODUCTION

breast sarcomas which develops from mesenchymal tissue are rare and their annual incidence is approximately 4.6 cases / 1,000,000 women, representing less than 1% of breast malignancies. They can appear as primary or secondary to chronic lymphedema or radiation therapy to chest wall, with the two forms presenting different features. The primary form appears histologically as heterogenous subtypes and their mean of diagnosis is around 40yrs. In contrast the secondary form typically presents later at around 40-45yrs. Lymph nodal metastasis is uncommon and surgery is the only They can share some clinical features with breast carcinoma but therapy and prognosis differ substantially. We present this case

given 50Gy/25# EBRT to chest wall completed in march 2014. Patient is on regular follow up since 2014 with

regular clinical check up, ultrasound abdomen and chest X-rayshowing normal findings (figure. 4).

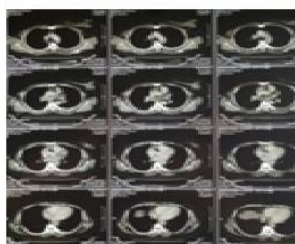


Figure 1

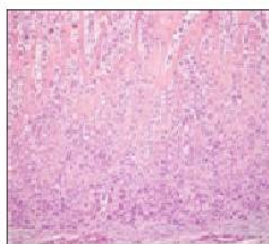


Figure 2

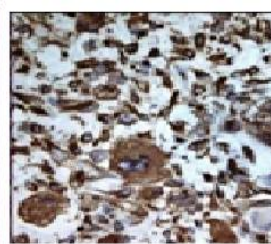


Figure 3



Figure 4

**Figure 1:** CECT thorax showing 8x6 cm heterogenous enhancing mass lesion in right breast, **Figure 2:** Histopathology showing multiple spindle shaped tumour cells with variable size round to oval nuclei with eosinophilic cytoplasm, **Figure 3:** tumour cells showing vimentin positivity in IHC, **Figure 4:** Patient with healthy post op scar

## DISCUSSION

Breast sarcoma is rare but aggressive entity. It is comparable to other soft tissue sarcoma and is totally independent of epithelial cancers. Due to its rarity, there is no sufficient knowledge to support consensus regarding best management. Core biopsy is the procedure of choice for diagnosis. The most commonly used staging system for breast sarcoma is American Joint Committee on Cancer staging system for soft tissue sarcoma chest. Lymphatic spread is uncommon so nodal status in breast sarcoma is less informative. Computed tomography is helpful since lungs are the predominant metastatic sites. The first line treatment is complete surgical excision of the tumour with adequate margins, regardless of histological subtype. Lymph node involvement is rare thus axillary lymph node dissection is recommended in cases of enlarged node, suspicious node on imaging. Routine lymphadenectomy does not seem to improve outcome. Adjuvant radiotherapy and chemotherapy for PBS is not consensual and depends mainly on the risk of tumour recurrence. There is a role of adjuvant radiotherapy for large (>5cm), high grade tumours or in positive surgical margins in reducing rates of locoregional recurrence. The impact on overall survival remains uncertain. Response rate to chemotherapy ranges from 20-40%. Adjuvant chemotherapy is an option for patients with good functional reserve, high risk sarcoma or in recurrent cases. Breast sarcomas have high recurrence rate and poor prognosis. Size of the tumour (5cm cut point) seems to be the most reliable prognostic factor. Studies have shown that a 5 year disease free survival ranges from 44 to 66% and 5 year overall survival ranges from 49 to 67%. Detrimental events such as local or distant failure and death, occur most frequently during the first year after diagnosis.

## CONCLUSION

Primary breast sarcoma is a rare entity and core biopsy is required for diagnosis. Unlike epithelial breast carcinoma lymphatic spread is uncommon and hence the nodal involvement. Computed tomography is helpful since lungs are the predominant metastatic sites. Surgery is the potential curative therapy. Tumour size and adequate resection margin are the important prognostic factors. Controversy still exists about the use of radiotherapy or chemotherapy. Currently, use of adjuvant therapy is limited with cases with high risk of recurrence due to positive margin, tumour >5cm or high grade sarcoma. A close follow up is recommended during the first two years when approximately 80% recurrences appear.

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