

Breast sarcoma: A rarity

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ABSTRACT

Delays in breast cancer diagnosis are associated with negative clinical effects. The main causes are protocol violations, clinical errors, radiological errors, pathological errors and patient fault. Although rare, breast lump that is clinically benign, but has undergone rapid growth should alert the clinician of the possibility of underlying sarcoma. We report a case of a 42-year-old female patient with breast sarcoma who developed recurrence of sarcoma along with ipsilateral pleural effusion. Owing to rarity of the disease and controversies involved in the management, there is no protocol for management of these patients.

Key words: Breast sarcoma, delayed diagnosis, metastasis, rapid growth

INTRODUCTION

Sarcoma of breast was first described by Chelius in 1828.^[1] Primary breast sarcoma accounts for <1% of women with breast malignancy and <5% of all the soft-tissue sarcomas.^[2] These tumors are important as they have high rate of recurrence and poor prognosis. Sarcomas are a heterogenous group of mesenchymal tumors that occur in extremities. When they occur in the specialized stroma of breast it is known as stromal sarcoma. Here, we report a case of a patient who developed recurrence within 6 months of surgery along with ipsilateral pleural effusion.

CASE REPORT

Our patient is a 42-year-old married perimenopausal woman who presented a year ago with a painless lump in right breast for 2 months for which she underwent simple mastectomy at a center. Histopathology report then was suggestive of mesenchymal stromal sarcoma. She did not receive adjuvant therapy. At 6 months later, she presented to us with recurrence of lump on the operated site which was rapidly progressive and painful. She also complained

of breathlessness and pain with fullness of right upper abdomen. On clinical examination, the patient was pale and tachypneic. There was a single lump approximately 7 cm × 6 cm in the right breast involving the whole breast with previous surgical scar mark beneath the lump. The right axillary lymph nodes were hard, fixed and palpable approximately 1.5 cm × 1 cm [Figure 1]. Radiography of the chest revealed massive pleural effusion on the right side with shift of trachea [Figure 2]. She underwent wide local excision of the recurrent lump along with axillary dissection. She also had drainage of the malignant pleural effusion. Histopathology report was indicative of stromal sarcoma of breast with angiosarcomatous and liposarcomatous areas [Figure 3]. At our center, we do not have facility for IHC hence it could not be performed. Inadequate treatment resulted in recurrence as in our case. Post-operatively, patient was advised to undergo adjuvant radiotherapy (RT). At 2 months later, patient came in emergency ward with dyspnea at rest and on chest X-ray patient had massive pleural effusion. In spite of all resuscitative measures however she expired.

DISCUSSION

Breast sarcomas arise from the connective tissue of the breast and are non-epithelial in origin.^[3] They can be primary in origin or arise as a complication of RT for the treatment of carcinoma of the breast (secondary).^[4]

The cause for primary sarcoma of the breast cannot be ascertained in most of the patients. Secondary or treatment related sarcoma of the breast can be traced to RT of the breast

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Figure 1: Clinical picture depicting scar with recurrence with axillary mass

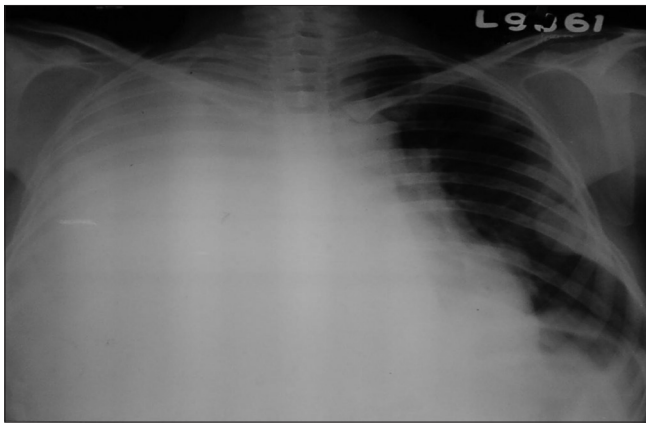


Figure 2: Chest X-ray depicting massive pleural effusion on right side

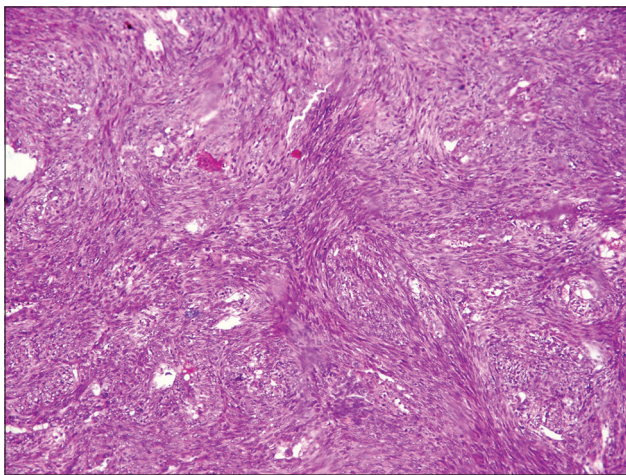


Figure 3: Microphotograph: Stromal sarcoma of breast with angiosarcomatous and liposarcomatous areas

either for carcinoma of the breast or for other malignancies like lymphoma, where either the breast or chest wall or both are included in the field of radiation.^[5] The risk of sarcoma increases with higher doses of radiation. A sarcoma is said to be secondary to ionizing radiation if:^[5]

- The initial tumor treated by RT is of different histology than the assumed radiation-induced sarcoma.
- Sarcoma develops in the previously irradiated field.
- There is a prolonged latent period (usually >4 years) between the two malignancies.

Chronic lymphedema of the breast following surgery or RT, where the axillary lymph nodes are sclerosed increases the risk of sarcoma, especially lymphangiosarcoma. This syndrome is called Stewart-Treves syndrome.^[6]

Other than ionizing radiation, some environmental factors like exposure to phenoxy herbicides,^[7] vinyl chloride, arsenic compounds have been linked to sarcomas.

As the sarcomas of the breast are histologically heterogeneous, the histology must be reviewed by an experienced pathologist for making the diagnosis and determining the sub type.

Stromal sarcoma of the breast is a distinct and rare entity. First described by Berg *et al.* in 1962, this subtype encompasses a group of breast sarcomas other than angiosarcoma, malignant cystosarcoma and lymphoma.^[8] Unlike the more common cystosarcoma phylloides, which arises from the intra lobular component, stromal sarcoma arises from the periductal stromal tissue and the lobular structure of the breast is not distorted. Apart from this Burga and Tavassoli at the Armed Forces Institute of Pathology have laid down the following criteria:^[9]

- A predominant spindle cell proliferation of variable cellularity and atypia around open tubules and ducts devoid of a phyllodes pattern
- One or multiple nodules separated by adipose tissue
- Stromal mitotic activity of three or greater in 10 high-power fields and
- Stromal infiltration into surrounding breast tissue.

The grade of differentiation of these tumors depends upon the extent of differentiation, necrosis, mitotic count and pleomorphism. As with the sarcomas in the rest of the body, the grade of the tumor determines the prognosis.

Sarcomas of the breast present as a painless, progressive lump in the breast which are usually unilateral and are often characterized by a rapid increase in size. On examination, the sarcoma is usually firm, non-tender, well-defined large mass in the breast. With the exception of angiosarcomas, the nipple areola complex and the skin are uninvolved.^[10] Regional lymph nodes are usually uninvolved. Staging is similar to the sarcomas arising from other sites. The most common site for metastasis of this tumor is the lung.

The diagnosis is made based on the clinical findings, imaging and biopsy. A mammogram is non-specific in

this setting and sometimes may even be negative or show a benign lesion. Fine-needle aspiration cytology has low accuracy in the diagnosis of this disease and should be avoided if a sarcoma is suspected. Core needle biopsy is the procedure of choice as it is possible to grade the tumor and determine the histological sub type. Incisional and excisional biopsy may also be attempted in suitable clinical scenarios.

Surgery is the standard treatment for sarcomas of the breast. It is critical to achieve an adequate negative margin during surgery as this increase the long term survival of the patient.^[11] The extent of surgery would depend on the size of the tumor. For tumors greater than 5 cm; mastectomy with or without reconstruction is ideal. Smaller tumors are managed by lumpectomy as sarcomas are usually not multicentric. Achieving negative margin is more important than the extent of surgery in these cases. If the axilla is clinically negative, axillary lymph node dissection is not indicated as sarcomas rarely spread through the lymphatics. The use of adjuvant RT remains unproven due to lack of level 1 evidence in this group of patients. However, there is support for RT in breast sarcomas because randomized controlled trials (RCTs) have proven an improved local control with RT in sarcomas of other sites.

As with RT, the role of chemotherapy is also not well-defined. There is some evidence for improved survival using doxorubicin and ifosfamide based chemotherapy.^[12] However, there are no RCTs that prove the effectiveness of chemotherapy in breast sarcoma.

As this disease is rare there are no fixed protocols for treatment. It is preferred that the treatment be tailor made for each patient by a multidisciplinary team consisting of an experienced surgeon, medical and radiation oncologist.

Proper psychological counseling and understanding of these patients is essential for them to continue with the treatment.

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