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Case Report

Primary Recurrent Stromal Sarcoma of Breast: A Rare Case

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A B S T R A C T

Sarcoma of the breast is an extremely rare tumour, usually of the spindle-cell variety, accounting for 0.5 % of malignant breast tumours and <5 % of all sarcomas. Fibrosarcoma is notorious for its rapid growth and rarely can be caught on FNAC, mammography, though histopathological diagnosis with immunohistology is for diagnosis is the gold standard. Here we report a 35 years old female, presenting with a huge, hard, painless left breast lump since, 10 years. Core needle biopsy revealed stromal tumour suggestive of phyllodes hence a left mastectomy was done and histopathology reported it to be a Stromal Sarcoma. Post which she was lost to follow-up and only returned after 1 year with a recurrent left chest wall hard lump. The patient then underwent a wide local excision with left latissimus dorsi rotational flap for the defect. Histopathology thus reported Stromal sarcoma of the breast. After uneventful post-operative hospital stay patient was further given External beam radiotherapy with photons. The purpose of this presentation is to give an insight on diagnosis and management of primary breast Stromal sarcoma.

Keywords: Recurrent stromal sarcoma; Histopathology; Radiotherapy

1. Introduction

Sarcomas of breast are extremely rare breast tumours with an incidence of <1% of all the primary malignancies of Breast and < 5% of all the sarcomas[1]. Estimated annual incidence is 17 new cases per 1,000,000 women[2]. They arise from mesenchymal tissues of the breast. A 90-year search of the mayo Clinic database revealed that primary breast sarcoma accounted for 0.06% of all breast cancers[3]. Tumour size was the most valuable prognostic factor, with 91% overall survival for women with sarcomas less than or equal to 5 cm and 50% for those with sarcomas greater than 5 cm and unlike carcinoma of any type of mammary sarcoma rarely metastasizes to lymph nodes[4].

Poland et al conducted a study in which, during a period of 80 years, only 4 cases of fibrosarcoma were reported of the 25 cases of primary breast sarcoma[5]. Terrier et al, reviewed of 33 cases of breast sarcoma of which only 2 cases of fibrosarcoma were reported[6]. Blanchard et al, reported only 2 cases of fibrosarcoma from his study consisting of 55 sarcoma cases[7].

Thorough tissue sampling is required to exclude a sarcoma arising in malignant phyllodes tumour, and immunohistology is necessary to exclude a high-grade sarcoma showing specific mesenchymal differentiation[8].

The most common presentation of fibrosarcoma of breast is rapidly progressing painless lump in breast which attains a large size. Not much literature is available on breast sarcomas, due to its rarity in incidence and reporting.

Occurrence of Breast sarcoma is more common in younger age group of 30-40 years age. Treatment is simple mastectomy with negative margins followed by Radiotherapy. The prognosis is dependent on staging and the histological type of the primary sarcoma[9].

2. Case Presentation

A 35year old lady, came to the surgical OPD with a complaint of a huge left breast lump, gradually increasing in size over 10 years, but she noticed rapid increase in the size seen over the

past few months. The lump was painless, not associated with nipple discharge. There was no history of any hormonal therapy, radiation exposure or trauma. Married since15 years, breastfed both children till 2 years; menstrual & obstetrics history were within normal limits. There were no positive findings in the past, personal or family history either.

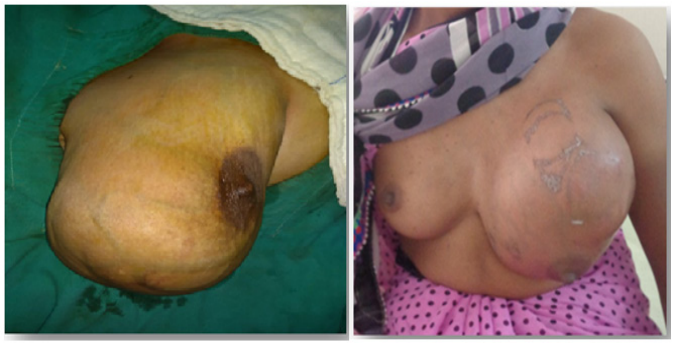

Figure 1: (a) & (b) Clinical photo of Left Breast Lump.

Clinically (**Figure 1**), there was single, large lump involving the entire left breast of 22x16cm dimensions, hard in consistency with ill-defined margins and bosselated surface. Left nipple-areolar complex was at a lower level compared to the right due to the lump, no nipple discharge seen. The lump was mobile; not fixed to the underlying chest wall, skin over the lump was pinchable, dilated veins visible, no dimpling or puckering seen. On Investigating further, the sonomammography reported 'A large ill defined heterogenous predominantly hypoechoic lesion note involving the entire left breast parenchyma with flecks of calcification within and showing vascularity on colour doppler s/o Neoplastic aetiology BIRADS 5 with bilateral axillary lymphadenopathy.' TRUCUT biopsy revealed 'Left breast Stromal tumour suggestive of phyllodes'. Hence, a simple mastectomy was done, (**Figure 2**) post which the histopathology reported 'Left breast stromal sarcoma.

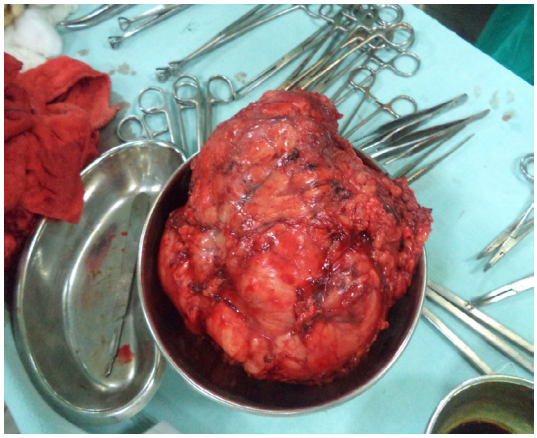

Figure 2: Mastectomy specimen.

The patient did not follow up for further treatment advised by the Oncologist, thus landing up with a local recurrence, left chest wall and axillary tumour (**Figure 3**). Two lumps 9x8cm over the left chest wall and 3x3 cm in left axilla, both hard in consistency, with irregular surface and rounded margins, mobile, not fixed to the underlying structures, skin above was shiny, not pinchable with previous mastectomy scar visible. CT Thorax reported 'A well-defined, round lobulated soft tissue lesion 7x4.6x8.6cm over the anterior chest wall, underlying muscle and ribs appear normal however, the lesion has infiltrated into the subcutaneous plane. Similar small lesion 1.6x1.2x1.7cm noted medial to the previous lesion'. A wide local excision with left Latissimus dorsi, pedicled rotational flap was done as a definitive surgical treatment.

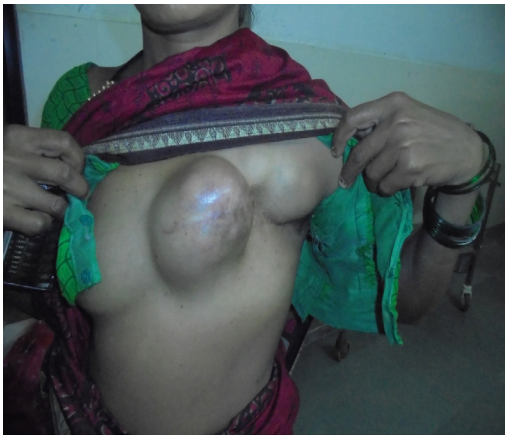

Figure 3: Recurrent chest wall Tumour.

3. Histopathology

Gross examination showed tumour near the base of the specimen with skin infiltration, four separate masses identified, no lymph node identified. Microscopic examination of the mass showed interlacing fascicles of spindle shaped cells with spindle shaped nuclei with tapering cytoplasmic processes. Frequent mitotic activity is noted (>10/10HPF) suggestive of left breast 'Stromal Sarcoma.' IHC was negative for Cytokeratin and EMA. The patient was further referred to Radiation Oncologist whereby she received 50Gy/25# in total and follows up regularly, no recurrence or new lesion found elsewhere (**Figures 4,5**).

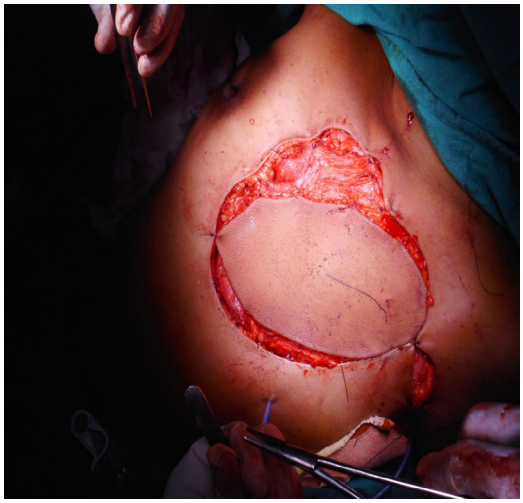

Figure 4: Left Latissimus dorsi flap to cover the defect.

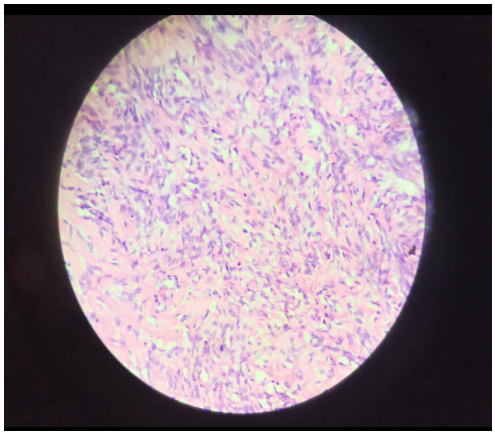

Figure 5: Microscopy of the Stromal sarcoma of the breast.

4. Discussion

Primary stromal sarcoma of the breast can be of various types like, fibrosarcoma, malignant fibrous histiocytoma, liposarcoma, rhabdomyosarcoma, leiomyosarcoma, hemangiosarcoma,

osteogenic sarcoma, chondrosarcoma and malignant schwannoma[9]. Fibrosarcoma usually occur primarily in the extremities, rarely does it occur in the breast. The size of primary breast sarcomas is variable and it ranges from less than 1 cm to larger than 40 cm[10]. Primary breast sarcomas may metastasize by hematogenous metastasis or direct invasion. Rarely axillary lymph node involvement is seen[1]. And the incidence of actual node metastasis also appears to be very low[11]. The prognosis is dependent on the tumour size and the histopathological grade[3].

Terrier et al[6]. reviewed 33 cases of primary breast sarcoma retrospectively and assessment of primary breast sarcoma prognostic factors was done. Of the total, 17 cases were Cysto-sarcoma phyllodes and Stromal Sarcomas were 16. The classification of stromal sarco-mas were done as follows-Malignant Fibrous Histiocytomas (11 cases), Leiomyosarcomas (2 cases), Fibrosarcoma (2 cases) and Liposarcoma (1 case).

In that study, only the histological grade, consisting of the presence of tumour necrosis, tumour differentiation, and the mitotic activity was significantly correlated with the metastasis-free survival rate. Both Cysto-sarcoma and stromal groups had identical clinical courses and survival, thus making the clinical value of this pathologic distinction questionable. All the local recurrence, metastasis or death occurred within 30 months, although the follow-up was much longer. Performing immunohistochemistry has not been very beneficial for identifying the specific histologic sub-types.

Post-operative radiotherapy helps to prevent local recurrence. Adjuvant chemotherapy is of some help in patients with highly malignant sarcoma, with positive surgical margins or post-operative recurrence[12], although the role of chemotherapy for breast sarcomas remains still unclear. The most effective regimen for chemotherapy is Adriamycin (ADM) + Ifosfamide (IFO).

5. Conclusion

Primary breast Stromal sarcomas are rare soft tissue tumours diagnosed mainly on histopathology. Surgical management is effective, followed by radiotherapy to avoid local recurrence and chemotherapy in cases of highly malignant tumours, thus demanding a multidisciplinary team approach for the betterment of the patients and efficient management.

6. Conflict of Interest

None declared.

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