

*Case Report***Stromal Sarcoma of Breast - An Unforeseen Rarity**Dr Evith Pereira¹, Dr Shilpi Sahu², Dr Reeta Dhar³, Dr Urshlla Kaul¹, Dr Kalyani Mahore¹¹2nd year Resident, ²Associate Professor, ³HOD & Professor,
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ABSTRACT

Soft tissue sarcomas of the breast being rare malignancy their early diagnosis is imperative as treatment modalities vary. Primary sarcomas of breast are extremely rare malignancies as compared to their counterpart epithelial tumors. We report a case of 36-year-old female who presented with a huge, fungating, irregular mass involving the entire left breast. The tumor was diagnosed histologically as Primary stromal sarcoma of breast with no axillary nodal involvement.

Key words: Stromal, Sarcoma, Breast, immunohistochemistry, CD 10.

INTRODUCTION

Malignant mesenchymal tumors of the breast are extremely rare neoplasms of breast with an annual worldwide incidence being 44.8 new cases per 10 million women. Breast sarcomas arising as primary lesion without any prior association with other malignancies or previous irradiation therapy are extremely rare. ^[1,2] Primary breast sarcomas constitutes around less than 1% of all the breast malignancies in women and less than 5% of all the sarcomas. ^[2-5]

CASE REPORT

Our patient is a 36 year old female who presented with a huge, fungating, irregular mass involving the entire left breast measuring 27x18x8 cm over a period of 6 months. There was no history of previous breast trauma, surgery or any positive family history of any breast malignancy. It was painless, foul smelling with areas of hemorrhage and necrosis (figure 1). There were multiple discrete lymph nodes palpable in the left axilla, largest measuring 1.4x1.0cm. The

contralateral breast was normal on examination. CT scan revealed a large ulcero-proliferative mass involving entire left breast with metastatic appearing left axillary and retropectoral lymph nodes. Bone scan showed no evidence of osteoblastic skeletal metastases. Ultrasonography of the abdomen and pelvis was non-contributory. Digital mammography of right breast showed no dominant mass, architectural distortion or any suspicious calcification. Few prominent axillary lymph nodes were detected which on USG correlation, these nodes show maintained fatty hilum which were suggestive of reactive etiology. The patient underwent left breast radical mastectomy.

Histopathological Examination:

The specimen was sent for histopathological examination which measured 28x19x8.5cm, with overlying flap measuring 25x20cm. The nipple and areola could not be identified. On serial sectioning, a large fungating tumor with a glistening lobulated cut surface measuring 27.5x18.5x8cm, involving the entire breast

was seen. It was seen ulcerating the skin and 0.7cm from the deep resection margin. Areas of hemorrhage and necrosis were seen in the cut section of the specimen. Two lymph nodes were sent measuring 1x0.8x0.6 cm. Microscopic examination revealed tumor cells with predominantly a perivascular growth pattern and fascicles separated by large areas of hemorrhage, necrosis and myxoid degeneration. Tumor cells are spindle to epithelioid with increased nuclear to cytoplasmic ratio, marked nuclear pleomorphism, hyperchromatic nuclei and vacuolated eosinophilic cytoplasm. Also seen are tumor giant cells with typical and atypical mitotic figures with mitotic rate of 10-15/high

power field, with areas of necrosis (Figure 2A & 2B). No epithelial differentiation was seen, with no areas resembling phylloides tumor. The base was free of tumor and no perineurial invasion or lymphovascular emboli were noted. The axillary lymph nodes showed features suggestive of reactive lymphadenitis. On immunohistochemistry, the tumor showed positivity for Smooth muscle actin, CD-10 and Ki-67 >40% proliferative index, whereas the tumor was negative for CK 5/6, CK 14, AE1/AE3, Desmin, CD-34, CD-31(Figure 3-A, B, C&D), thus suggesting high grade stromal sarcoma of breast.



Figure 1- Showing huge, fungating irregular tumor with areas of necrosis and hemorrhage measuring 27x18x8 cm

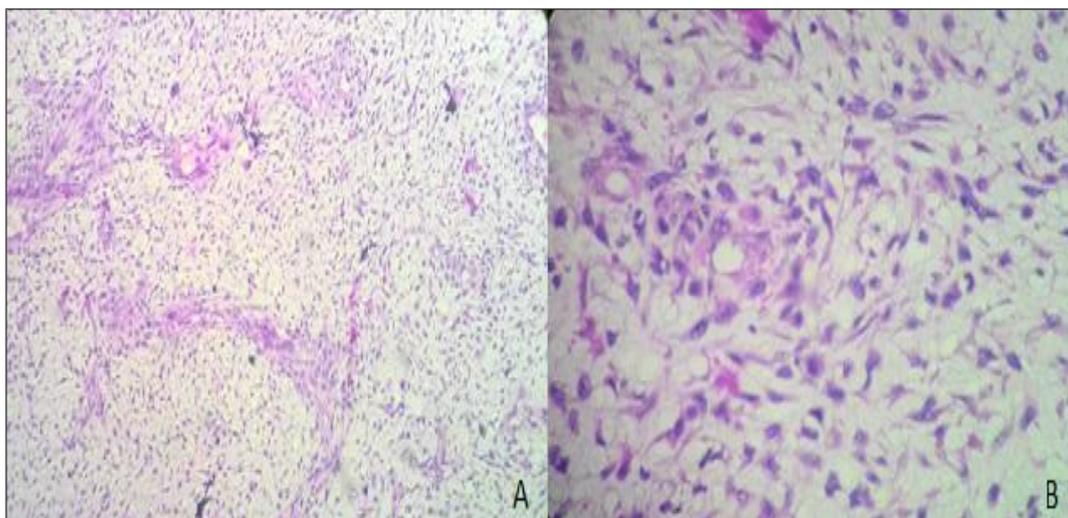


Figure 2 (A) – H& E 10x – Showing Perivascular growth pattern with myxoid degeneration.

Figure 2 (B) – H&E 40x – Showing Spindle shaped cells with nuclear pleomorphism, increased N: C ratio, vacuolated eosinophilic cytoplasm with mitotic figures.

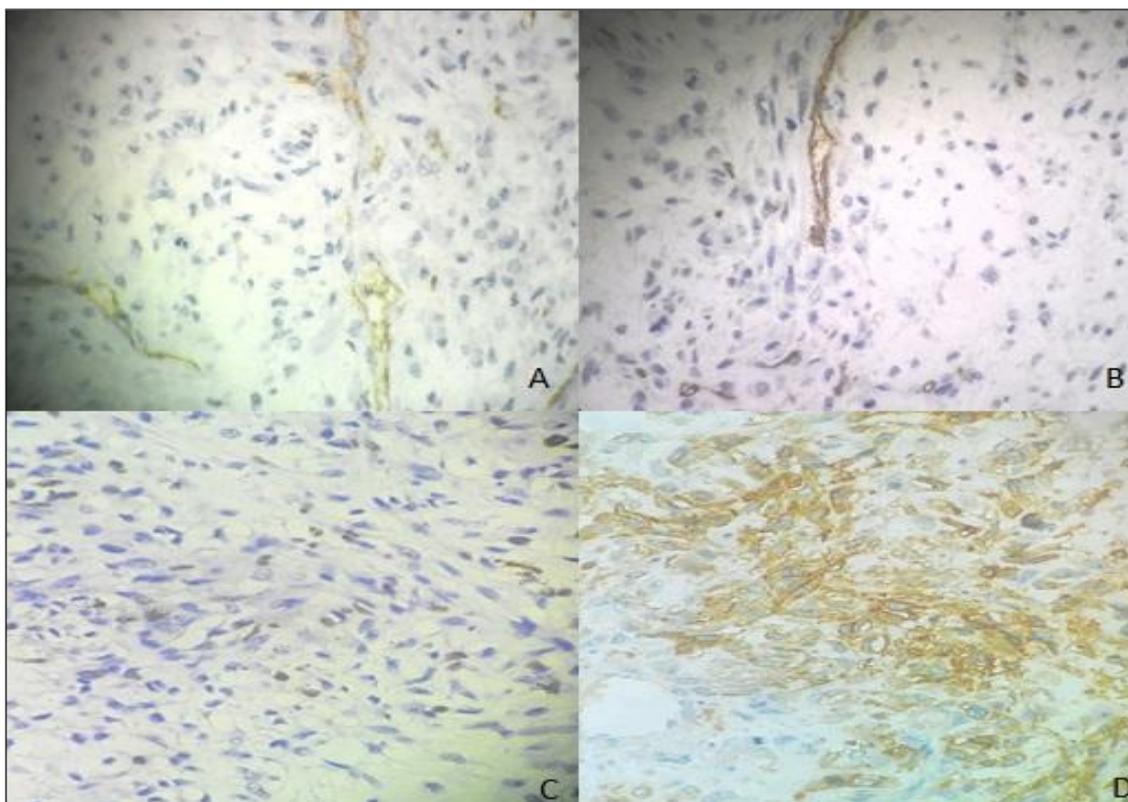


Figure 3 - Immunohistochemistry stains 40x – (A) CD 31- negative, (B) CD 34 – negative, (C) Ki67 >40% (D) CD 10 – Positive.

DISCUSSION

Primary stromal sarcomas are extremely rare malignant mesenchymal tumors of breast. [1,4] These tumors being rare, few hundred cases are reported in the English literature. [5] The clinical course, incidence of sarcomas and histological types of breast sarcomas are not yet well established which is basically due to rarity of these tumors. [2] Berg et al. was the first to define Stromal sarcoma. They defined it as a group of mesenchymal malignant tumors comprising of fibrous, myxoid and adipose components excluding malignant phylloides, lymphomas and angio-sarcomas of breast. [1-5] These tumors were named as stromal sarcomas because they were considered as the normal variants of mammary duct stroma. [2] Unlike the common variants like cystosarcomaphylloides, which are intra-lobular in origin, the stromal sarcoma arises from the peri-ductal stromal tissue leaving the lobular architecture of breast undistorted. [4] Etiology of primary breast sarcoma includes patient previously treated with radiotherapy, lymphangio-sarcoma

following lymph edema of the breast, neurofibromatosis and hereditary diseases. [3] It has also been suggested about the possible association between silicon prosthesis implantation and that of breast sarcomas but this has not been proved yet and angiosarcomas being radiation induced. [2]

The prognosis in these tumors is directly proportional to the grade of tumor, size and extent of metastasis. These tumors generally metastasize through the hematogenous route to lung, bone and exceptionally to the lymphnodes, with nodal metastasis indicating end stage disease. [1,2] In our case the patient had no distant metastasis of tumor; despite multiple discrete lymphnodes being palpable in the left axilla they showed features of reactive lymphadenitis on histological examination. Nodal involvement along with high mitotic rate is the poor prognostic markers, with size of the tumor and infiltrating borders having lesser values comparatively. Immunohistochemistry markers prove to be an important guide to differentiate stromal sarcomas from other varied sarcomas of the

breast. Stromal sarcomas arise from the specialized stroma of breast lacking epithelial component. Many of these sarcomas do not match with the precise appearance of the tumors arising from the other usual sarcomas of soft tissue locations, owing to the specialized stroma, thus CD 10 positivity are examples of this phenomenon. In our case, on immunochemistry proved CD 10 positivity thus supporting the diagnosis of Stromal Sarcoma.^[6]

Adjuvant radiotherapy may be indicated when the primary tumor is bulky or when there is no satisfactory local clearance of tumor cells. Role of chemotherapy has not yet been defined in stromal sarcomas, these tumors show ineffectiveness towards adjuvant hormonal therapy as breast sarcomas lack hormonal receptors.

CONCLUSION

Stromal sarcomas of the breast should be kept in the differentials of all malignant mesenchymal and undifferentiated breast lesions which can present with or without any nodal metastasis. In large breast lesions early tru-cut biopsies and immunohistochemistry

should be considered to differentiate epithelial origin tumors from mesenchymal origin ones. For early detection and typification of breast malignancies an effort should be made to raise awareness and emphasize the use of immunohistochemistry for the diagnosis of such rare entity.

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