Primary Leiomyosarcoma of Breast

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ABSTRACT

Primary leiomyosarcoma of breast is an extremely rare form of breast cancer with an unpredictable biological behavior, and usually presents as a slow growing mass in a middle to old age women. Surgical resection with wide margins is the main stay treatment followed by chemo radiotherapy. Long term follow up is done to look for local recurrence or distant metastasis. We are presenting a case of 73 year female diagnosed with primary leiomyosarcoma of the breast.

Keywords: Leiomyosarcoma, Breast carcinoma, sarcoma breast

INTRODUCTION

Stromal sarcomas of the breast constitute about 1% of all malignant tumors of the breast and less than 5% of all soft tissue sarcomas[1]. Primary leiomyosarcoma of the breast comprises a rare type of stromal sarcoma of the breast [2]. In a study conducted at the Mayo Clinic, the prevalence of primary breast sarcomas among breast cancers was found to be 0.0006 % [3]. Surgery represents the only potentially curative therapy in these cases, and tumor size and adequate resection margin are the most important prognostic factors.

We are presenting a case report of a 73 year old female with high grade leiomyosarcoma of breast.

CASE REPORT

A73 year old female came with complaints of painless lump in the left breast since 1 year, which was gradually progressive in size. Patient had no history of previous breast disease or radiation to breast.On examination her vitals were normal. Examination of the left breast revealed a lump of 4x3 cm in the upper and outer quadrant of left breast near the anterior axillary fold . The lump was well defined with a smooth surface[Figure 1]. It was non tender and variable in consistency. There was no retraction of the nipple, no fixity to underlying structures and no palpable lymphadenopathy.

Her hematological and biochemical parameters were within normal limits. FNAC of the lump showed features of high grade malignancy with a possible differential diagnosis of poorly differentiated carcinoma or high grade sarcoma. For further evaluation core biopsy from the left breast lump was done which showed large tumor cells with high N: C ratio, highly pleomorphic hyperchromatic nucleus,thin walled blood vessels and few tumor cells surrounding blood vessels suggestive of angiosarcoma. USG of the left axilla showed no significant lymph nodes.

In view of diagnosis of angiosarcoma of breast with no clinical and radiological evidence of lymph nodes, the patient underwent simple mastectomy. Post-operative period was uneventful. Patient was discharged on 7thpost-operative day after drain removal.

Histopathology showed tumor size 6.5x 6x 4cms in upper outer quadrant with infiltrating tumor composed of spindle cells with pleomorphic nuclei and coarse chromatin suggestive of high grade sarcoma[Figure 2]. IHC confirmed grade 3 leiomyosarcoma with no lymphovascular invasion ER, PR, HER2neu negative, KI antigen 80-85% positive and anti positive SMA. Pathological staging: Grade 3 pT2NxMx was given.

Patient then underwent adjuvant radiotherapy of 50Gy in 25 fractions and 5 fractions per week, for 5 weeks duration by IMRT technique by 6MV photon linear accelerator followed by six cycles of adjuvant chemotherapy with Gemcitabine. Patient is being followed up regularly.

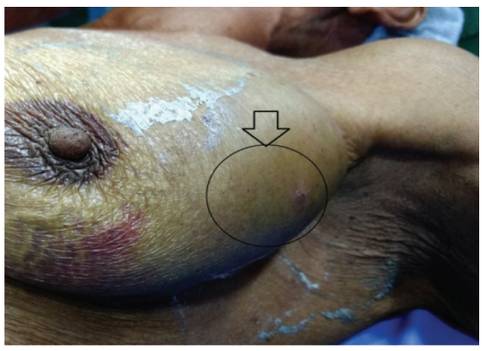

Figure 1: Showing the lump in the upper outer quadrant

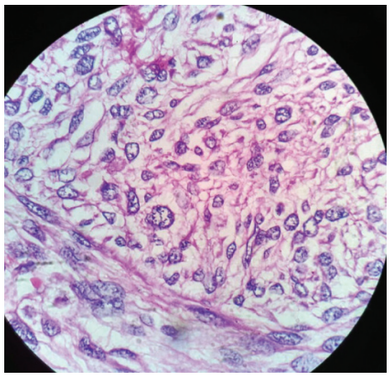

Figure 1- Showing spindle shaped cells with hyperchromatic nuclei, H&E x100

DISCUSSION

Breast sarcoma is a malignant neoplasm that arises from mesenchymal tissue and is part of a wide spectrum of connective tissue tumors, accounting for less than 1% of malignant breast lesions. The incidence, histological type and clinical course of sarcomas of the breast are not well established. This is due to the rarity of these tumors and a lack of definition in various reports.

The most commonly described breast sarcomas are fibrosarcoma, liposarcoma and undifferentiated high-grade sarcoma. Other sarcomatous lesions such as leiomyosarcoma and sarcoma with bone and cartilage are rare. The etiology of breast sarcoma is unknown. Angiosarcoma may be induced by radiation. Leiomyosarcoma probably originates from blood vessels, the smooth muscle of the nipple- areola complex or the myofibroblast[4]. They can appear as primary forms (denovo) or secondary to chronic lymph edema or radiation therapy on the breast or chest wall, with the two forms presenting different features.

The primary forms appear histologically as heterogeneous subtypes and their mean age of diagnosis is around 40 years. In contrast, the secondary forms typically present later at around 45–50 years and the most common histological subtype is angiosarcoma.

Leiomyosarcoma is characterized by spindle-shaped cells with pleomorphic, hyperchromatic and elongated nuclei, eosinophilic cytoplasm,large nucleoli and significant mitoses. Definitive diagnosis is established through histological examination, in which positive staining is observed immunohistochemically with desmin, vimentin, and muscle-specific actin, whereas negative staining is seen with cytokeratin, myoglobin, and S-100[5].

Lymph node metastases are rare in breast sarcomas and spread of the disease usually occurs haematogenously. The principal organs to be metastasized are lungs, bones and liver. Hence sentinel lymph node biopsy does not seem to offer sufficient benefit to the patient or to prompt a change in clinical management. Surgery is the standard treatment, tumor size and adequate resection margin are the most important prognostic factors and an adequate resection margin is the most important determinant of long-term survival [6].

The prognosis of leiomyosarcoma is better than that of other breast sarcomas [7]. The benefits of chemotherapy and radiotherapy have not yet been confirmed [7,8]. Al Beena et al. in his review article advised to consider chemotherapy and radiotherapy to improve local recurrence especially in cases where the tumour is larger than 5 cm and also where histology shows positive surgical margins[9]. Some studies showed uncertain benefit of chemotherapy in breast sarcoma [10]. Breast sarcomas are also non-responsive to the hormonal treatment as they lack hormonal receptors [11].

CONCLUSION

Breast sarcoma is a rare but aggressive entity and core biopsy is required for diagnosis. Unlike with epithelial breast carcinoma, lymphatic spread is uncommon and staging studies differ from other breast tumors due to the fact that nodal status in breast sarcoma is less informative. Surgery represents the only potentially curative therapy in these cases, and tumor size and adequate resection margin are the most important prognostic factors. They require long-term follow-up because local recurrence and distant metastasis can occur long after the operation. The benefits of chemotherapy, radiotherapy, and hormonal therapy are still controversial. It has a better prognosis than other breast sarcomas. However, there is a need for further studies to determine the prognostic factors.

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