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BREAST LEIOMYOSARCOMA, A RARE CASE REPORT

Radiation Oncology

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ABSTRACT

Breast sarcomas accounts only 1% of all malignant tumors of the breast, among which only 5% - 10% are leiomyosarcomas. LMS commonly noticed in 45-55 year age group people with almost similar presentation like other stromal tumors of breast.

It is difficult to diagnose leiomyosarcoma preoperatively as excision biopsy and immunohistochemistry are essential for the final diagnosis. Due to the rarity of the disease the treatment for LMS is not standardized. However excision of the tumor either by lumpectomy or simple mastectomy with wide clear margins and adjuvant radio therapy to the chest wall is considered as the optimal treatment.

KEYWORDS

Leiomyosarcoma, Biopsy, Rare

INTRODUCTION:

Sarcomas of breast are rare which accounts only less than 1% of all the malignant tumors of breast [1, 2].

Among sarcomas of breast only 5%-10% that is (0.0006%) [3, 4] are leiomyosarcomas, and only less than 50 cases are being reported till date in English literature which signifies the rarity of the disease [5].

The myofibroblasts found in the nipple areolar complex believed to be the origin for the neoplasm [6]. The 45-50 years age group are commonly effected with almost similar presentation like other stromal tumors of breast [7-9]. It is difficult to diagnose primary leiomyosarcoma of breast preoperatively, as histopathology and immunohistochemistry are essential for the final diagnosis.

Excision of the tumor with clear margins is generally considered as the optimal treatment as most of them are low grade tumors [10, 11]. Lymph node dissection is not routinely done as the tumor spreads through hematogenous route [12].

Due to the rarity of the disease the role of chemo-radiation is not well established. Here we report a case of primary leiomyosarcoma of breast with optimal treatment regimen

Case report:

a) Clinical presentation and examination:

A 52 year old postmenopausal female presented with complaints of painful lump over the right breast. She has noted the lump accidentally while having bath 9 months earlier which was of marble size. Since than the lump increased progressively to a size of cricket ball. On clinical examination 6*6 centimeter large, well defined, firm, mobile pass was palpable in her right upper inner quadrant with overlying skin and nipple areolar complex normal. No axillary or supraclavicular lymph nodes were palpable. Left breast examination was unremarkable. The patient did not have any family history or other comorbidities in her first degree relatives nor previous history of radiation to chest wall. Mammography of right breast revealed large well defined lesion with microlobulations in upper inner quadrant of right breast with mild increased density in surrounding breast tissue. Left breast was unremarkable. Other investigations like chest scan and ultrasound abdomen and pelvis were normal.

b) Pathological findings:

The patient underwent core needle biopsy from the breast lump revealed high grade stromal tumor. Multidisciplinary opinion sorted and simple mastectomy of right breast done.

Microscopic examination of the MRM specimen revealed cellular tumor composed of interlacing bundles of moderate to highly pleomorphic oval to spindle shaped cells with hardly any intervening stroma. Tumor cells show oval to elongated nuclei, irregular nuclear margins, coarsely clumped chromatin , variable to prominent nucleoli and moderate amount of fusiform eosinophilic cytoplasm with many mitosis.

Immunohistochemical staining for Vimentin, SMA, h- caldesmon, calponin was strongly positive. cytokeratin, desmin and S-100 staining was neagative, the axillary nodes were negative for metastasis and by which confirmed the diagnosis of primary leiomyosarcoma of the breast.

Post multidisciplinary opinion the patient was than advised with adjuvant external beam radiotherapy of 50Gy to whole breast and a dose of 60Gy to the tumor bed for a period of 5 weeks. Post radiation the patient is on follow up since 1 year and no local recurrence or distant metastasis noticed.

DISCUSSION:

Breast cancer is the most common cancer in females worldwide [13]. Generally epithelial tumors dominate in breast and mesenchymal tumors are rarely reported [14]. Less than 1% of malignant breast tumors are sarcomas and leiomyosarcomas among them are even rare [1]. Till date only around fifty cases are being reported in the literature. Leiomyosarcomas are generally encountered in fifth or sixth decade of life. They are believed to originate from muscular wall of the blood vessels or from the smooth muscles of nipple areola complex [15].

Sarcomas can occur any where in the body however commonly seen sites are uterus, subcutaneous tissues, retroperitoneum, and gastro intestinal tract [16]. Leiomyosarcomas are extremely rare in breast either primary or from metastasis.

Following predisposing factors may be associated with breast sarcomas 1) Prior radiation to breast or chest wall 2) preexisting fibro adenomas 3) chronic lymphedema of the breast or arm 4) Neurofibromatosis or Li-fraumeni like syndromes [13].

Breast sarcomas typically presents with lump which is well circumscribed , dense , lobulated , fibro adenoma like mass growing slowly over 2yrs – 5yrs , which signifies it as a long standing mass. The average tumor size is about 4.5 cm [17] with high chance of local recurrence even after 15 – 20 years [18].

Leiomyosarcomas generally metastize through hematogenous route and has no significant correlation with tumor size for metastasis [19].

Breast skin and nipple areolar complex are not generally involved and findings on ultrasonography and mammography are not specific [20]. So it is difficult to diagnose the disease preoperatively without biopsy or immunohistochemistry.

The tumor microscopically suspected with features like high cellularity, fusiform spindle cells with blunt end nuclei and atypia with frequent mitosis. In immunohistochemistry staining, tumors cells are

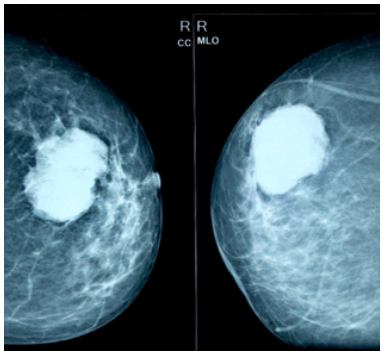
positive for SMA (smooth muscle actin), desmin, calponin. Negative for S-100, cytokeratins, and other epithelial markers [21, 22].

Till date surgical excision either simple mastectomy or modified radical mastectomy with wide clear margins is considered as the optimal treatment . However according to ESMO guide lines for soft tissue and visceral sarcoma and some studies favour for adjuvant radiotherapy which improve in loco regional control [23, 24]. In our current case also simple mastectomy done with adjuvant postoperative radiotherapy done to improve loco regional control. The patient is on follow up since 8 months with no local recurrence or distant metastasis. The prognosis is better in patients with leiomyosarcoma when compared to other sarcomas of the breast [19]. There is a need for long term follow up, to study the prognostic factor and overall survival.

CONCLUSION:

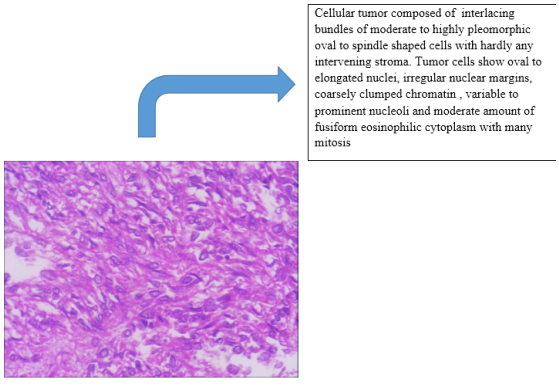
Mesenchymal tumors are rare when compared to epithelial tumors of breast. Biopsy and immunohistochemistry (desmin+,SMA+,vimentin +, calponin +) can confirm the diagnosis of leiomyosarcomas from other breast sarcomas. Surgical excision with wide clear margins and adjuvant Radiotherapy for locoregional control is considered as the optimal treatment. Lymph node dissection is generally not considered as the tumor spreads via hematogenous route. The prognosis of leiomyosarcomas is good compared to other breast sarcomas. Large studies are necessary for the proposal of standard treatment regimen and outcomes.

Fig 1: Mammography right breast:

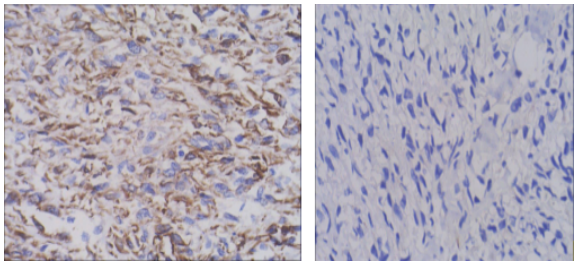


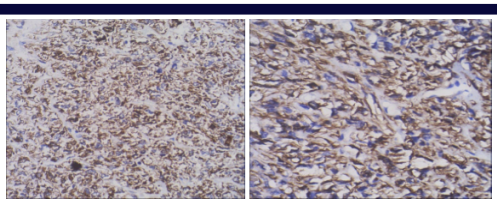
Large well defined lesion with microlobulations in upper inner quadrant of right breast with mild increased density in surrounding breast tissue.

Fig 2: Histopathology examination



a)VIMENTIN positive **d) DESMIN negative**





b) CALDESMON positive **c) CALPONIN positive**

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