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Myxofibrosarcoma of the Breast: A Rare Case Report

Benjamin Malsawmtluanga[1*], Rahul Mahawar[2], Yumkhaibam Sobita Devi[3], Robert Zirsangzela Thangluah[1]

[1]Junior Resident, [2]Senior Resident, [3]Professor

Department of Radiation Oncology, Regional Institute of Medical Sciences(RIMS), Imphal, Manipur, India

***Corresponding Author:**
Dr. Benjamin Malsawmtluanga
Junior Resident, Department of Radiation Oncology, Regional Institute of Medical Sciences(RIMS), Imphal, Manipur, India
E-mail: bmchhangte@gmail.com

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Abstract

Myxofibrosarcoma are rare malignant neoplasms. It generally develops in the extremities with high risk of local recurrence. We are reporting a case of primary myxofibrosarcoma of the left breast in a 37 years old woman treated with multimodality approach including surgery, chemotherapy and radiotherapy.

Keywords: myxofibrosarcoma, soft tissue sarcoma, breast cancer, radiotherapy

Introduction

Myxofibrosarcoma of breast is a subtype of soft tissue sarcoma (STS) and is extremely rare. They account for less than 1% of all malignant breast tumours.[1] There are no specific immunohistochemical marker and genetic profiles for myxofibrosarcoma, so diagnosis is based on histology.[2] The key features of myxofibrosarcoma includes atypia, myxoid stroma, multinodular growth pattern and curvilinear vascular structures.[3]

Case Report

A 37 years old female patient attended the Surgery OPD at Regional Institute of Medical Sciences (RIMS) Imphal with complaint of pain and swelling in the left breast for last 3 months. On physical examination a single well defined globular mass 6 cm x 5 cm in size, mobile, non-tender, with smooth surface and margin was palpated in the outer lower quadrant of left breast. No other lymph node or mass were clinically palpable.

The mammogram showed a large lobulated soft tissue opacity in the left breast 4 0'clock para-areolar location measuring 7 x 5 x 4 cm in size, extending into the retromammary fat, likely a large fibroadenoma/ phylloides tumor, BI-RADS II. On sonogram a heterogenous lobulated mass lesion 7.7 x 7.3 cm with no vascularity on Doppler was seen in the left outer lower quadrant. Incisional biopsy was performed and the histopathological examination (HPE) showed a malignant tumour composed of spindloid to epitheloid cells arranged in sheets and in fascicles. Individual cells have moderate to abundant amount of cytoplasm and highly pleomorphic vesicular nuclei with open chromatin and distinct nucleoli. Extensive areas of necrosis and infarction are seen and many abnormal mitotic figures and atypical cells are identified with overall features suggestive of a high grade sarcoma (Figure 1). On immunohistochemistry (IHC), the tumour cells are negative for CK (AE1/AE3), ER, PR, Her-2/neu, CD 34 and S100 but positive for SMA and Desmin with a Ki 67 index of 50% (Figure 2).

Contrast enhanced computed tomography (CECT) thorax showed a heterogeneously contrast enhancing lesion with central necrosis seen in lower half of the breast measuring 8.5 X 6 cm in size. A contrast

enhancing nodes was seen in left axilla measuring 2.5 X 0.7 cm. Other baseline investigation were within normal limits with no evidence of metastasis on metastatic work up. She received Neoadjuvent chemotherapy with Inj. Epirubicin (day1&day 2) and Inj. Ifosfamide (day 1-day 5) 3 weekly for a total of 4 cycles.

The patient underwent left modified radical mastectomy one month after the completion of 4th cycle of chemotherapy and the excised specimen was sent for HPE. Macroscopically, serial cut sections showed a tumour involving almost the entire specimen measuring 19x12x11 cm. Serial cut section were soft to firm, friable, grey white and glistening with extensive myxoid areas. Areas of necrosis were noted. Microscopically, multiple sections showed breast tissue with a highly malignant tumour displaying hypocellular and hypercellular areas. The hypercellular areas display spindloid to epitheloid cell arranged in short fascicles and in multinodular haphazard growth pattern. Individual tumour cells have abundant amount of cytoplasm and highly pleomorphic vesicular nuclei with open chromatin and distinct multiple nucleoli in a collagenised stromal background. The hypocellular areas shows tumour cells arranged in loose singles in a myxoid stromal background. Many scattered tumour giant cells with bizarre nuclei and atypical cells are seen. Numerous abnormal mitotic figures are identified (>30/10 hpfs). Extensive areas of necrosis (>50%) are seen and many foci display moderate to dense mixed inflammatory cell infiltration composed predominantly of neutrophils with the tumour invading the stroma with irregular infiltrating margins with feature suggestive of myxofibrosarcoma of the breast tissue.

Four weeks after the surgery, the patient attended the OPD at the department of radiation Oncology, RIMS with the HPE report. The general condition of the patient was good with the KPS of 90% and BSA of 1.5 m^2. Routine baseline investigation were within normal limit, and the patient was planned for external beam radiotherapy.

Patient underwent adjuvant external beam radiotherapy to the chest wall, axilla and supraclavicular fossa with a total tumoricidal dose of 50 Gy in 25 fractions(2 Gy /fraction) , 5 days a week for 5 weeks (Figure 3) Two weeks following the completion of EBRT, the patient came to the RIMS casualty with complain of respiratory distress with chest X ray (PA view) showing multiple bilateral pulmonary nodules and bilateral pleural effusion (Figure 4). The patient was kept on supportive care and was planned for palliative chemotherapy, but the patient succumbed to the disease after 5 days.

Discussion

Malignant Fibrous Histiocytoma (MFH) is a subtype of STS and Myxofibrosarcoma is a variant of MFH, characterized by nodular growth, a myxoid matrix, a coarse, plexiform capillary pattern, and spindle or stellate tumour cells with hyperchromatic, atypical nuclei.[1] Breast Sarcomas presents as breast lumps which was clinically and radiological mimic breast fibroadenomas. These tumors carry a high recurrence rate with worst prognosis than adenocarcinoma of the breast.[4]

The diagnosis of Myxofibrosarcoma is based on histology in most cases, and various IHC markers were examined for differential diagnosis.[5] Some breast tumours consists of pleomorphic spindle-shaped tumor cells that often show a mucinous matrix and are difficult to differentiate when attempting to obtain a diagnosis of myxofibrosarcoma. These includes Phyllodes tumor, myxofibroblastoma, and metaplastic carcinoma.[6]

In phyllodes tumor, positivity for Progesteron receptor (PgR) and Androgen receptor (AR) have been reported in 100%,[7] and 98%[8] of examined stromal cells respectively. In the present tumor, PgR was negative. Therefore, the possibility of a malignant phyllodes tumor could be excluded.

A component of spindle cells in metaplastic carcinoma is immunoreactive for cytokeratins though variably, while the present tumor did not express cytokeratins at all. But myofibroblastoma is a benign tumor, and the tumor cells in that tumor usually express CD34, ER, PR,[9] and; these were not expressed in the tumor in question.. Therefore, both myxofibroblastoma and metaplastic carcinoma could be excluded from the differential diagnosis of the present tumor.[2]

Complete surgical resection with negative margin is the recommended treatment.[4] Surgical approach can be achieved with breast conservative surgery (BCS) versus simple mastectomy. Lymph nodes dissection is not recommended as standard practice, since the

Volume 7, Issue 3; May-June 2024; Page No 284-287

incidence of lymph nodes metastasis in soft tissue sarcomas are extremely rare. Adjuvant radiotherapy should be recommended to improve local control. The role of chemotherapy in non-metastatic disease is not well documented.[4]

Patient with myxofibrosarcoma had five years survival rate of 30% to 35%.[10] They can metastasize to lung, bone, and lymph node and risk of local recurrence is high, careful follow up is considered necessary

Conclusion

The present report provide details of the case in which myxofibrosarcoma of breast was diagnosed based on histology and immunohistochemical examination results. The first step of treatment is surgery with adequate safe margins. Dissection of axillary lymph nodes is debatable. Adjuvant radiotherapy must be considered. We are reporting a case of myxofibrosarcoma of breast which was treated by multimodality approach consisting of neoadjuvent chemotherapy followed by surgery and adjuvant local radiotherapy.

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Volume 7, Issue 3; May-June 2024; Page No 284-287

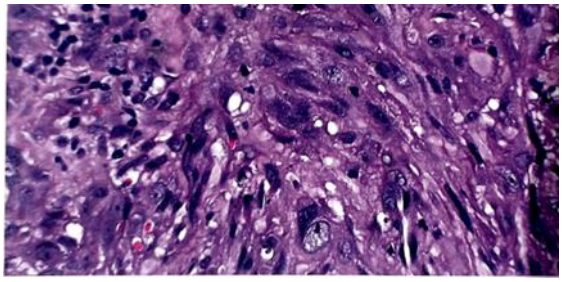

Figure 1: HPE finding of the biopsy specimen showing high grade sarcoma

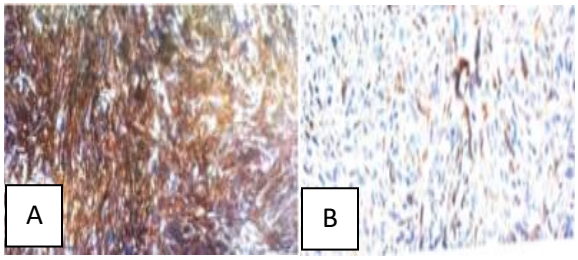

Figure 2: A) Positive immunoreactivity of tumour cells with SMA. B) Positive immunoreactivity of tumour cell with Desmin.

Figure 3: Picture showing patient receiving External beam radition therapy

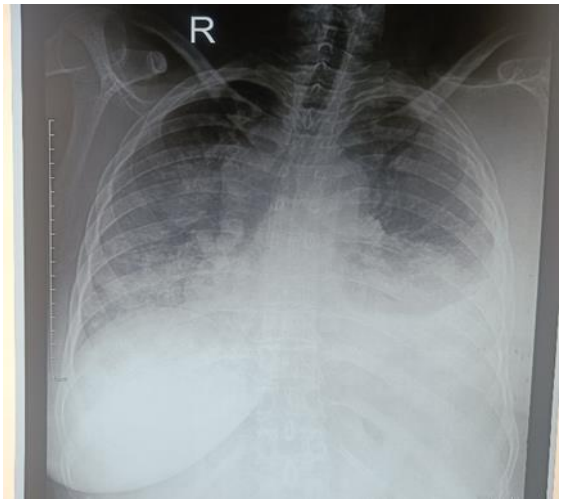

Figure 4: Chest X-Ray (PA View) showing bilateral pleural effusion and bilateral lung nodules

Volume 7, Issue 3; May-June 2024; Page No 284-287