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Malignant Fibrous Histiocytoma of the Breast: A Rare Case Report

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Abstract
Background: *Primary malignancies of the breast stromal elements and sarcomas of mesenchymal elements constitute less than 1 % of all breast neoplasms and malignant fibrous histiocytoma (MFH) of the breast is exceptionally rare. Histopathologically it is characterized by tumor pleomorphism with varying proportion of fibroblast and histiocyte like cells which stain positive for vimentin. We report our experience of a young female with MFH of the breast treated with mastectomy and adjuvant chemotherapy.*
Case: *A 27 years old female presented with a progressively increasing lump in her right breast. On examination the lump was in the upper outer quadrant of the right breast,8 X 8cm in size, firm, mobile and nontender. The overlying skin was tense and glossy; neck or axillary nodes were not palpable. FNAC showed round to spindle shaped cells arranged in clusters; trucut biopsy revealed the cells in a typical storiform pattern suggestive of MFH.A mastectomy was done and patient received 3 cycles of adjuvant chemotherapy. 2 months after completion of chemotherapy she was asymptomatic.*
Conclusion: *MFH of the breast is an extremely uncommon entity and a complete clinicopathological correlation with histopathological profile is requisite to arrive at an accurate diagnosis. The present case stresses upon the fact that MFH of breast must be kept as a differential diagnosis of any breast lump showing spindle cells and pleomorphism on cytology.*
Key Words: *MFH, breast, tumor pleomorphism*

Introduction

Malignant mesenchymal tumors constitute <1 % of all malignant neoplasms of the breast.[1] These tumors may arise de novo from the breast parenchyma itself or following adjuvant irradiation for carcinoma of the breast. Among these tumors, primary malignant fibrous histiocytoma (MFH) of the breast is very rare because of which optimal treatment guidelines are still not clear.MFH is also known as malignant fibrous xanthoma, pleomorphic fibrous histiocytoma and pleomorphic fibrous xanthoma.[2] We present here an uncommon case of primary malignant fibrous histiocytoma of the right breast.

Case

A 27 years old female presented in the Department of Radiation Oncology, Regional Cancer Centre, Cuttack with a lump in her right breast, which was progressively increasing in size for the last 6 months. She had no history of fever, pain, weight loss, trauma or discharge from the nipple. On examination, there was a lump in the upper outer quadrant of her right breast, about 8X8 cm in size, firm, mobile, nontender. The lump was free from the underlying structures and the overlying skin appeared tense, glossy and erythematous. There were no lymph nodes palpable in the neck or bilateral axillae. The contralateral breast and axilla were normal. Abdomen was soft, nontender without any ascites or hepatosplenomegaly. Routine laboratory parameters and biochemical profile were within normal limits except for mild anemia.USG abdomen pelvis and radiograph of the chest were both normal. Bilateral mammogram was done and mediolateral oblique view of the right breast showed large lobulated well defined retroareolar mass with indistinct margins and absence of microcalcifications or spiculation.(Fig 1) FNAC from the lump showed large, round to spindle shaped cells arranged in loose clusters or dispersed singly with necrosis and haemorrhage in the background. A trucut needle biopsy from the lesion was consistent with MFH of breast and she was referred to the department of surgical oncology for further evaluation. Mastectomy of the right breast was done, gross examination of the specimen showed a retroareolar growth about 8X 7X5 cm involving the whole nipple areola complex.(Fig 2) Cut surface was grayish yellow, fleshy with typical fish flesh appearance, abundant hemorrhagic areas and necrosis. (Fig 3).Postoperative histopathology revealed diffuse proliferations of round to spindle shaped cells arranged in storiform or cartwheel pattern with round to oval vesicular nuclei, plump eosinophilic or foamy cytoplasm .Numerous atypical mitoses were seen along with binucleated or multinucleated giant cells and prominent necrosis suggestive of MFH of breast.(Fig 4,5) She was started on combination chemotherapy of injection vincristine, adriamycin and cyclophosphamide alternating with injection ifosfamide and etoposide and received 3 cycles each. 2 months after completion of chemotherapy she was asymptomatic and was thereafter lost to follow up.

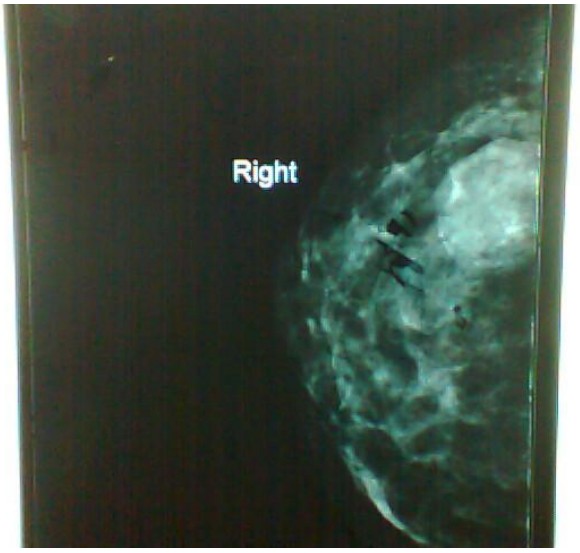

Fig 1: Mammogram of right breast showing large lobulated well defined retroareolar mass with indistinct margins and absence of microcalcifications or spiculation.

Fig 2: Mastectomy specimen showing a large retroareolar growth about 8X 7X5 cm involving the whole nipple areola complex

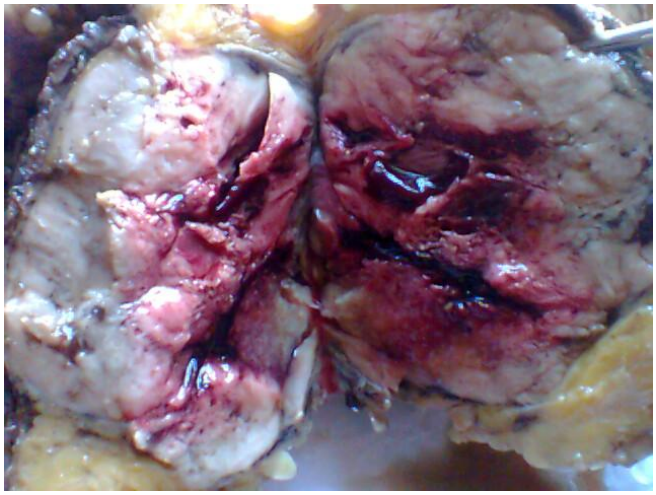

Fig 3: Cut section of the specimen showing pale myxoid with typical fish flesh appearance with areas of hemorrhage and necrosis.

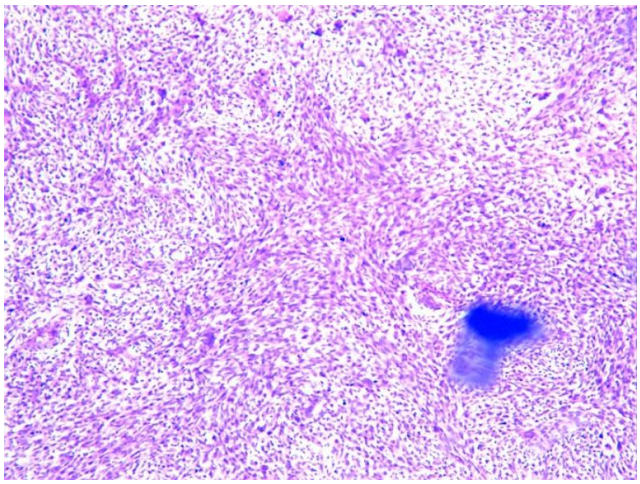

Fig 4: Histopath (low power) showing spindle shaped cells cells arranged in clusters or dispersed in a storiform pattern.

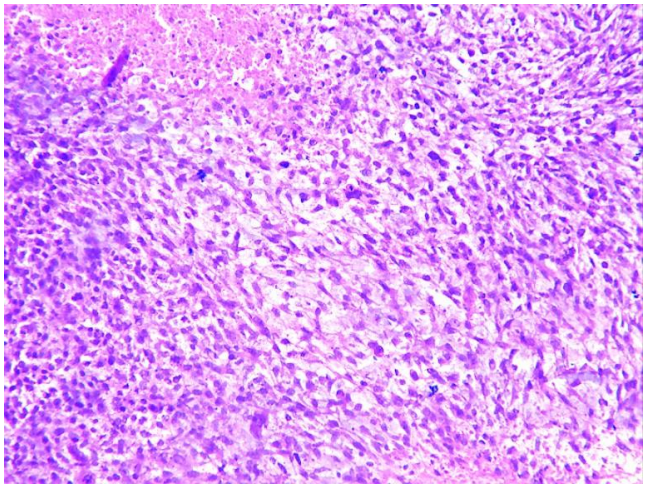

Fig 5: Histopath (high power)showing round to spindle shaped cells with oval vesicular nuclei, plump eosinophilic or foamy cytoplasm arranged in a typical storiform or cartwheel pattern.

Discussion

The term MFH(Malignant fibrous histiocytoma) was first introduced in 1963 by Ozzello et al to describe a malignant soft tissue tumor morphologically characterized by a storiform or cartwheel like growth pattern.[3] Enzinger and Weiss described it to be arising from primitive mesenchymal cells with partial histiocytic and fibroblastic differentiation.[4]MFH is essentially a tumor arising from the skeletal muscle, dermis and retroperitoneum but has also been described in other rare sites like larynx, conjunctiva, vulva, bone and breast. Primary malignancies of the breast stromal elements and sarcomas of mesenchymal origin account for less than 1% of all breast neoplasms and MFH arising from the breast is exceptionally rare.[5]

MFH of the breast can be of various types: (i)primary MFH of the breast where it arises de novo from the connective tissue of the gland,(ii)MFH of breast associated with cystosarcoma phylloides,(iii)post radiation MFH of breast.[6] Radiation induced MFH is seen several years after treatment in the irradiated field of breast carcinoma, retinoblastoma, Hodgkin's disease and multiple myeloma.MFH of breast in post irradiated patients is increasing because of the growing trend of breast sparing surgery followed by radiation in the treatment of carcinoma breast.[7]

Clinically these patients present with enlarged, painless slow growing mass in the breast with tense, glossy appearance of the superficial skin. It may also present as a large hematoma because of intratumoral hemorrhage. Cut surface shows pale, myxoid, fleshy appearance with hemorrhage and necrosis.

Histopathologically, the cardinal feature of MFH is tumor pleomorphism which contains two types of cells:-fibroblast like cells and histiocyte like cells in varying proportions to display a wide spectrum of histological patterns. The four principal variants of MFH are:-(i)fibrous type,(ii)giant cell type or pleomorhic,(iii)myxoid type and(iv) inflammatory type.[8] For proper management, MFH of breast needs to be excluded from other carcinoma or

sarcoma of the breast. Immunohistochemically these cells stain positive for vimentin and negative for cytokeratin. Some authors have also described these cells to stain positive with alpha1antitrypsin and lysozyme. The differential diagnosis of MFH includes dermatofibrosarcoma protuberans, cystosarcoma phylloides and pleomorphic rhabdomyosarcoma. Dermatofibrosarcoma protuberans also has a storiform growth pattern, is locally invasive and can infiltrate the breast parenchyma thereby leading to confusion with MFH of breast. However primary MFH of breast as described by Lunde et al are characterized by absence of an epithelial component and skin involvement.[9] MFH of breast occurring in patients with existing cystosarcoma phylloides are to be categorized and treated as the latter.Cystosarcoma phylloides is characterized by a combination of glandular and stromal components; as the tumor becomes more aggressive the stromal components become more predominant. Multinucleated giant cells are strikingly absent in cystosarcoma while multiple sections in a specimen of MFH shows many bizarre tumor giant cells and mitotic figures 8 to 10/ HPF without any epithelial or dermal component. Pleomporhic rhabdomyosarcoma of the breast on histopathology shows multiple cross striations and rhabdomyoblasts. Like cystosarcoma, MFH also recurs locally and distant metastases is via hematogeneous route to lungs and lymph nodes.

In general prognosis of this tumor is poor due to local recurrence and the most important prognostic factors include depth of location and size.[10] Langharn et al reviewed 4 patients of primary MFH of breast whose lesions were treated by simple or radical mastectomy which showed local recurrence but no distant metastases or deaths within a follow up period of 11 to 24 months. [11]

Although treatment guidelines are unclear, surgery is by far the primary modality of treatment. There is no literature to recommend modified radical mastectomy over simple mastectomy in MFH but surgical margins of more than 3 cm is considered safe.[12]Review of literature suggests that axillary lymph node metastases in MFH are exceedingly uncommon and therefore routine axillary dissection is not recommended in these patients. The role of adjuvant treatment is not defined and both radiotherapy and chemotherapy have been used with some benefit. We treated this patient with adriamycin based combination chemotherapeutic regimen which has shown success in some cases.

Conclusion

Primary MFH is a rare malignant neoplasm of the breast but the possibility should not be ignored in a rapidly growing painless mass of the breast. A careful histopathological evaluation should be correlated with the clinicopathological profile to reach a definitive diagnosis. Surgery remains the primary option followed by adjuvant treatment with either radiotherapy or adriamycin based chemotherapy. Long term follow up of these patients is essential to define treatment sequelae and outcome.

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