CASE REPORT

Primary pleomorphic rhabdomyosarcoma of breast: Report of a rare neoplasm

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Primary sarcoma of the breast is very rare and constitutes less than 1% of all breast cancers. Herein, we report a case of pleomorphic rhabdomyosarcoma (PRMS) of the right breast in a 49-year-old female patient presented with a mass (7 cm × 6.5 cm). Mammography and ultrasonography suspected a malignant lesion and a diagnosis of poorly differentiated carcinoma was made on fine needle aspiration cytology. Modified radical mastectomy was carried out. Histopathological examination revealed a high grade stromal sarcoma with rhabdoid morphology and multinucleated tumor giant cells. The tumor cells were strongly positive for desmin, vimentin and Myo D1 focally. The tumor cells were immunonegative for cytokeratin, epithelial membrane antigen (EMA), CD34, CD45, SMA, S100, CD68 and HMB45. A final diagnosis of PRMS was rendered. Surgical margins were free and no metastasis was seen in axillary lymph nodes. Neither post-operative radiotherapy nor adjuvant chemotherapy was given and the patient has remained disease free 12 months post-operatively.

Key words: Breast, pleomorphic, primary sarcoma, rhabdomyosarcoma

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INTRODUCTION

Primary breast sarcomas are rare and comprise <1% of all malignant breast neoplasm.[1] In a study, at Mayo clinic of America primary sarcomas of breast accounted for 0.0006% (18 out of 27,881 malignancies).[2] These malignancies arise from the connective tissue of the mammary gland, however, cases of cystosarcoma phyllodes are believed by Pollard to be excluded from this list as it is a distinct clinicopathological study.[2] Primary sarcomas of the breast are a highly heterogeneous group, which include malignant fibrous histiocytoma (MFH), liposarcoma, leiomyosarcoma, fibrosarcoma, angiosarcoma, rhabdomyosarcoma and other sarcomas.[3]

CASE REPORT

A 49-year-old postmenopausal woman presented with a right breast lump for 7 months and attended medical college and hospital, Kolkata, India for medical treatment in February, 2012. On physical examination, a hard mass (7 cm × 6.5 cm) was palpable in the upper outer quadrant of the right breast.

Mammography showed a well-circumscribed, lobulated mass of soft-tissue density, which is denser than adjacent parenchyma. No calcification was found within the breast lump. Ultrasonography revealed a heterogeneous mass with relatively high internal echogenicity. Fine needle aspiration cytology (FNAC) showed discretely arranged large polygonal pleomorphic cells and few multinucleated giant cells. A provisional diagnosis of poorly differentiated carcinoma was given on cytologic smear examination. Modified radical mastectomy along with axillary lymph node dissection was performed. The tumor was stage III, T3N0M0.

Grossly, a right mastectomy specimen (12 cm × 9 cm) was received. Cut section revealed a well-circumscribed, non-encapsulated tumor (7 cm × 6.5 cm) with hemorrhage, necrosis and internal cystic changes. Deep resection margin and overlying nipple-areola complex were grossly free of tumor. A total of 17 lymph nodes were sampled from axillary clearance. All lymph nodes were small and grossly unremarkable. Histologic sections from tumor showed large polygonal cells arranged in diffuse sheets. The cells have rhabdoid morphology having eccentric nuclei and eosinophilic cytoplasm. The cells have high nucleocytoplasmic ratio and prominent nucleoli. Mitotic figures were high. Multinucleated tumor giant cells were present. Entrapped ducts were unremarkable and no atypia was noted in these ductal epithelial cells [Figure 1]. Based on morphology a list of differential diagnosis including pleomorphic rhabdomyosarcoma (PRMS), epithelioid sarcoma, malignant melanoma and anaplastic

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lymphoma were included and immunohistochemistry was suggested for confirmatory diagnosis. The tumor cells were diffusely and strongly positive for desmin and vimentin. Myo D1 was focally positive. The tumor cells were immunonegative for cytokeratin (CK), epithelial membrane antigen (EMA), CD34, CD45, SMA, S100, CD68 and HMB 45 [Figure 2]. Only entrapped benign ductal epithelial cells showed CK positivity. Thus, we excluded the possibilities of malignant melanoma (HMB45$^-$ S100$^-$), anaplastic lymphoma (CD30$^-$, EMA$^-$, CD45$^-$), epithelioid sarcoma (CK$^-$, EMA$^-$, CD34$^-$). However, tumor cells were positive for desmin, vimentin and MyoD1 (focal). Hence; a final diagnosis of PRMS was established. All 17 lymph nodes were negative. In our case, there was no previous history of radiotherapy or chemotherapy.

Neither adjuvant chemotherapy nor post-operative radiotherapy was given. The 12 months follow-up period was uneventful.

DISCUSSION

Primary breast sarcomas or true "stromal sarcomas" are distinctly rare and are a highly heterogeneous group. They should be differentiated from the two main differential entities, metaplastic (sarcomatoid) carcinoma and malignant phyllodes tumor.[4] The metaplastic carcinoma is recognized by the presence of a carcinomatous component and immunoreactivity for CK and EMA in neoplastic spindle cells. Malignant phyllodes shows stromal overgrowth with spindle cell pattern in predominate area, leaf such as a pattern and invasion of the surrounding tissues. All these features were absent in our case and positivity for desmin, vimentin and Myo D1 help to rule out malignant phyllodes in the present case. Moreover, in the present case the tumor cells were polygonal with rhabdoid morphology rather than spindle cell pattern seen in the above two differential diagnoses.

PRMS is a high grade sarcoma occurring almost exclusively in adults, are more common in men and present at a median age in the 6th decade.[4] These tumors usually occur in the deep soft-tissue of the lower extremities.[5] PRMS arises in areas where myofome derived skeletal muscle occurs and is therefore usually occurs in extremities, especially in the thigh. However how does it occur in breast parenchyma, remains unclear as in our case. Sailer S Opined that the rhabdomyoblastic tumor of the breast arises from a misplaced mesenchymal "rest". However, others believe that it is not wise to explain it by aberrant differentiation for misplaced cellular "rests" especially in an organ such as breast where mixed tumors occur and mesenchymal elements show great plasticity in producing bone, cartilage, adipose tissue and other heterologous elements.[5] So, it is more reasonable to consider that rhabdomyoblastic elements are products of differentiation occurring in a tumor of mesenchymal origin.[6] Stout AP believed that rhabdomyoblastic cells can "assume the guise and function of fibroblasts," but the presence of many monster giant cells is evidence that the tumor is not a fibrosarcoma.[7].

In a study of 25 cases of primary breast sarcoma, it was found that tumor size very important prognostic factor. 5 years overall survival and cause specific survival were both 91% for tumor <5 cm and 50% for tumors >5 cm. Mean tumor size with recurrence or metastasis was 7.7 cm, compared with 4.9 and 4.3 cm respectively for patients without recurrence or metastasis.[2] The predisposing factors for breast sarcomas are prior radiotherapy, genetic syndrome like Li-Fraumeni and Gardner's syndrome, environmental associations such as arsenic, vinyl chloride, chemotherapeutic agents, certain viruses such as herpes and human immunodeficiency virus.

The standard therapy for breast sarcoma is mastectomy, which, according to Pollard, should include the excision

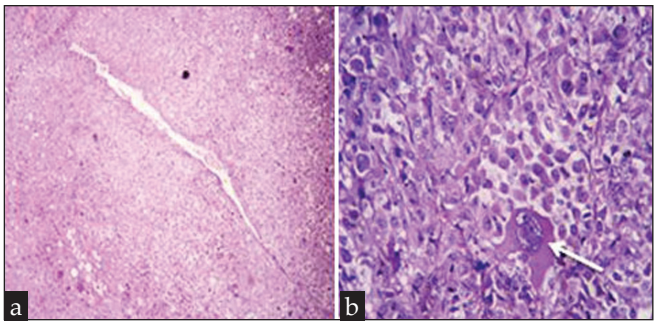

Figure 1: (a) Photomicrograph showing tumor cells with rhabdoid morphology and arranged in diffuse sheets (H and E, ×100). (b) Photomicrograph of tumor cells showing eosinophilic cytoplasm, eccentric nuclei, prominent nucleoli and occasional giant cells (arrow) (H and E, ×400)

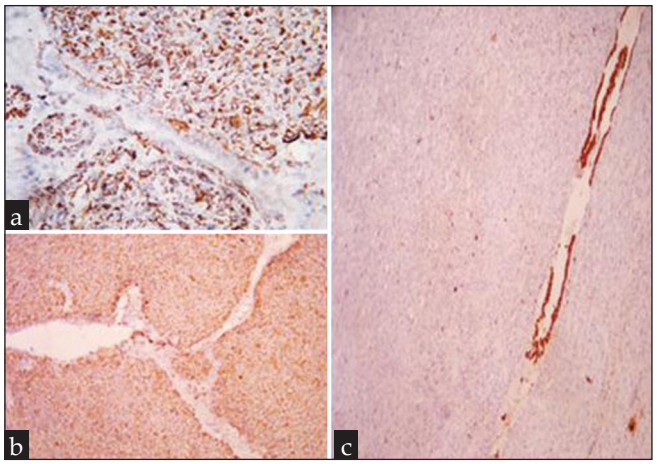

Figure 2: (a) Photomicrograph of vimentin positivity among tumor cells, but entrapped duct epithelial showing negative expression (×200). (b) Photomicrograph of desmin positivity among tumor cells, but entrapped duct epithelial showing negative expression (×100). (c) Photomicrograph of cytokeratin negativity, but positivity seen in entrapped un-remarkable duct epithelial cells (×100)

of both underlying pectoral muscles, in under to reduce the recurrence rate.[3] Other approaches include a breast preserving wide local excision (WLE) with adequate negative margins, particularly for small sarcomas (<5 cm). The staging or therapeutic role of routine axillary lymph node dissection has not been defined. As lymph node involvement is low, many believed that lymph node dissection is unjustified.[8] Poynter *et al*. in 2006 observed that routine lymphadenectomy does not confer a survival benefit. Rather than performing axillary sampling for these patients, a consideration may be the routine use of sentinel lymph node biopsy.[9] However, Pollard *et al*. reported high rates of local recurrence after WLE and simple mastectomy (67% and 54% respectively).[3]

Role of adjuvant therapy in breast sarcomas is controversial as there is no prospective randomized trial. Chemotherapy is not very promising in these tumors. Radiotherapy could be tried in patients with inadequate surgical margins or microscopically involved.[3,8] In our case, the tumor size was >5 cm (7 cm × 6.5 cm) and modified radical mastectomy with axillary dissection was done, but no adjuvant therapy was given. To rule out possibility of secondary sarcoma of breast, patient was thoroughly checked and relevant investigations were done. However, no other primary tumor was detected. Besides, patient had no previous history of radiotherapy or chemotherapy especially for primary breast carcinoma. Hence, diagnosis of primary sarcoma of breast was established. Primary sarcoma of the breast is rare and primary PRMS is extremely uncommon.

AUTHOR'S CONTRIBUTION

All authors have contributed in designing and conducting the study. All authors have assisted in preparation of the first draft of the manuscript or revising it critically for important intellectual content. All authors have read and approved the content of the manuscript and confirmed the accuracy or integrity of any part of the work.

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