CASE REPORT

Primary Chondrosarcoma of Breast

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ABSTRACT

Mammary sarcomas are heterogeneous group of malignant neoplasms that arise from the mammary stroma. They are uncommon tumours and most of these occur as a component of other tumours. Of the malignant breast mesenchymal tumours, pure sarcomas which lack epithelial component are rarer as these comprise only 0.5% of the breast tumours. Of these, the most common are angiosarcomas, liposarcomas and osteosarcomas. Pure, primary and *De novo* chondrosarcomas features as one of the rarer types of sarcomas of breast and should be differentiated from Phylloides tumours with chondromatous areas by extensive sampling which also excludes ductal elements in the tumour. This case report describes very rare primary breast sarcoma i.e. chondrosarcoma in a female aged 40 years which was treated by simple mastectomy.

Key Words: *Chondrosarcoma. Malignant neoplasm. Sarcoma. Primary breast malignancy. Mastectomy.*

INTRODUCTION

Breast sarcomas are primary non-epithelial malignancies arising from the connective tissue within the breast.[1] They can arise *De novo* (primary) or secondary to radiation therapy or lymphedema after treatment of other malignancies (secondary).[2,3] Primary sarcomas of the breast are extremely rare ranging from 0.5% to less than 1% of all malignant tumours of breast. Of these, pure chondrosarcomas without any other area of epithelial or mesenchymal differentiation are even rarer. This excludes tumours like malignant cystosarcoma phyllodes and metaplastic carcinomas where malignant cartilaginous areas may be present. Further primary chondrosarcomas arise from breast stroma and not from the underlying bone or cartilage. Although their clinical features mimic breast carcinomas in some ways, therapy and prognosis differ dramatically.[4]

This case report describes very rare primary breast sarcoma i.e. chondrosarcoma in a female aged 40 years which was treated by simple mastectomy.

CASE REPORT

A 40-year-old female presented in the breast surgery out patient department (OPD) in Liaquat National Hospital with a lump in the right breast for the last 10 months. There was no history of nipple discharge or any other breast problem. Patient was married with two children. There was neither family history of breast cancer nor history of any exposure to radiation.

On examination, a hard lump of about 23 x 20 cm, occupying most of the breast was noted. It was mobile, not attached to the underlying structures and the overlying skin also appeared normal. The ipsilateral axillary nodes were not palpable. The contralateral breast and axilla were also normal on examination. Clinical suspicion of breast carcinoma was raised.

The mammographic study was reported as BIRADS IV. Her pre-operative metastatic workup including CT chest and abdomen showed clear lung fields and absence of liver metastasis. Subsequently simple mastectomy was done.

The mastectomy specimen of right side was received in formalin in one container in the Department of Histopathology, Liaquat National Hospital. It measured 26 x 22 x 17 cm. The elliptical skin flap measured 23 x 15 cm. The skin, nipple and areola appeared normal. On serial slicing, a huge grayish white tumour was seen measuring 21 x 19 x 11 cm. There was no gritty or necrotic area seen.

A malignant neoplasm with multinodular pattern of growth was noted. The individual tumour nodules consist of round to slightly elongated cells separated by myxoid / chondroid matrix. The individual cells have mostly vesicular nuclei with prominent nucleoli and deeply eosinophilic cytoplasm. Mitoses were appreciated readily. Skin, nipple and areola were tumour free. The tumour was 0.3 cm away from closest posterior resection margin. Benign skeletal muscle fibers were also seen. The specimen was further sampled extensively to look for any other malignant stromal or epithelial component. None was found (Figure 1). A panel of immunohistochemical antibodies were applied by DAKO envision method including CKAE1/AE3, Vimentin, S-100, EMA and CK-7. Out of these, Vimentin and S-100 were found strongly positive in tumour cells and the remaining were negative. A final diagnosis of primary chondrosarcoma of breast was made. Patient did not develop any postoperative complication. She received six cycles of radiotherapy and has been on regular follow-up till now.

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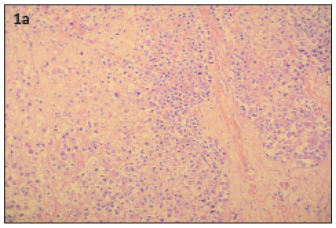

Figure 1a: Section of the tumour exhibiting chondroid areas only with no malignant epithelial component (Hematoxylin and Eosin x 20).

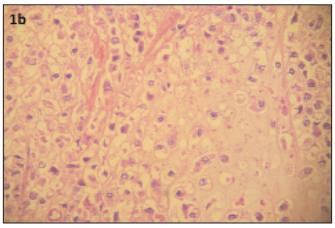

Figure 1b: High power view of Chondrosarcoma revealing chondrocytes showing marked cellular atypia. Mitotic figures are also appreciated (Hematoxylin and Eosin x 40).

DISCUSSION

Primary breast sarcomas are heterogeneous group of tumours. They can be primary or secondary. The prevalence, risk factors and clinical course of breast sarcomas and other primary non-epithelial malignancies are less well characterized than are the breast tumours arising from epithelial tissue. A causative factor is not identified in majority of cases. In contrast, secondary breast sarcomas are associated with primary radiation therapy or conditions causing chronic lymphedema.[5]

Majority of the primary breast sarcomas are malignant fibrous histiocytoma, fibrosarcoma, liposarcoma, angiosarcoma, rhabdosarcoma, dermatofibrosarcoma and desmoid tumours etc.[4]

Primary and pure chondrosarcoma occurring in the breast is extremely rare. It arises from the breast itself and contains chondromatous areas throughout the tumour. Very few cases of pure and primary chondrosarcomas of the breast have been reported so far.[1-10] Theses tumours typically present as freely mobile and circumscribed mass, tend to grow rapidly. Mammography reveals a dense mass. They are usually large in size and do not involve the overlying skin and lymph nodes.[4]

Microscopically, the tumour shows chondroid areas with cellular atypia. This has to be clearly differentiated from matrix producing metaplastic carcinoma and from malignant cystosarcoma pyllodes with chondroid differentiation. Immunohistochemistry for cytokeratin and myoepithelial markers is helpful for ruling out epithelial component and excluding metaplastic carcinoma. Further, the sarcomas like areas in metaplastic carcinoma though show vimentin positivity, but still retain epithelial marker positivity. Similarly, differentiation from malignant cystosarcoma pyllodes with dominant chondromatous areas requires benign ductal elements to be interspersed. Cystosarcoma phyllodes with chondromatous areas are still rare.[4,5]

Surgery remains the mainstay of treatment for most sarcomatoid tumours.[4,9] Multimodality treatment may decrease local and systemic recurrence rates of somatic sarcomas, but results are inconclusive in patients with breast sarcomas.[4,10] As only cases of primary chondrosarcoma of the breast have been reported so far, role of radiotherapy or chemotherapy in this particular breast sarcoma is difficult to assess. As against this, the present case showed partial response to chemotherapy. Favourable prognostic factors like low degree of cellular atypia and mitotic rate could have had a bearing on this response to chemotherapy.

The tumour was negative for any of the hormonal receptors. This supports the theory that adjuvant therapy with estrogen antagonists and other hormone manipulations have no role in the treatment of mammary sarcomas.[4,5]

Primary sarcomas of the breast are rare tumours and it is important that these be recognized as a separate entity from the more common breast carcinoma keeping in mind the difference in behaviour of the two tumours while planning therapy.

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