

Mammary Sarcoma - Not Otherwise Specified: A Case Report

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Abstract

Mammary sarcomas are a heterogeneous group of malignant neoplasms that arise from the mammary stroma. Primary sarcomas of the breast are extremely rare and comprise less than 0.1% of all malignant tumors of the breast. We present the case of a 56-year-old female diagnosed as stromal sarcoma, not otherwise specified - a very rare entity.

Keywords: Breast sarcoma, Undifferentiated

Introduction

Mammary sarcoma or stromal sarcoma arises from interlobular mesenchymal elements which comprise the supporting mammary stroma. Mammary sarcomas mostly show features of liposarcomas or angiosarcomas and rarely pleomorphic sarcoma or malignant fibrous histiocytoma. Undifferentiated mammary sarcoma or mammary sarcoma - not otherwise specified (NOS) is an extremely rare finding and diagnosis is made after exclusion of all other malignant spindle cell tumors in the breast.¹

Case Report

A middle aged female presented with a slowly growing mass in the left

breast of one year duration. On examination it was firm to hard in consistency in the inner quadrant and measured 6×5×3 cm with a skin nodule size of 3×2×1 cm. Biopsy showed slender to plump spindle cells arranged in sheets and fascicles with nuclear pleomorphism and abundant mitoses (4-6/HPF) with varying amounts of collagen (Figures 1, 2). A diagnosis of mammary sarcoma was made. She underwent a modified radical mastectomy and the specimen was solid, greyish white in color. The specimen was sectioned widely to search for a glandular epithelial component which was absent. Immunostaining was performed using pan cytokeratin (panCK), vimentin, CD34, S100, BCL2, estrogen receptor (ER),

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progesterone receptor (PR), Ki 67, smooth muscle actin (SMA), desmin, vimentin, and leukocyte common antigen (LCA). The tumor was strongly positive for vimentin (Figure 3) and negative for all other markers (Figure 4). CD10 was performed later which was moderately positive (Figure 5). A diagnosis of mammary sarcoma - NOS was made. For the past six months, the patient has been seen for follow up visits without any evidence of recurrence. The patient remains on a follow up schedule.

Discussion

Primary sarcomas of the breast that arise from mesenchymal tissue of mammary glands are very rare. The term stromal sarcoma includes all sarcomas other than cystosarcoma phyllodes.² Usually stromal sarcomas are subclassified according to the histological description of the cell of origin. Some of the subtypes reported as case series and case reports are malignant fibrous histiocytoma (MFH), fibrous sarcoma, angiosarcoma, spindle cell sarcoma, leiomyosarcoma, liposarcoma, rhabdomyosarcoma, osteosarcoma, chondrosarcoma, and synovial sarcoma.¹

Primary sarcoma of the breast with no specific differentiation based on morphology or immunohistochemistry has been described as an NOS type of mammary sarcoma.² This is a very unique entity, not much has been described in the

literature. Liebl and Moinfar described this sarcoma in 2006.³ There were seven cases of mammary sarcoma which did not fit into any specific soft tissue category. Histologically these were composed of spindle cells with pleomorphic nuclei and abundant mitosis with collagenous stroma. A similar case of undifferentiated mammary sarcoma has been reported in a young female which was a recurrence of a benign phyllodes tumor.⁴

The diagnosis of stromal sarcoma NOS type should be made after taking into consideration all other tumors with similar histological features. Sarcomatous overgrowth in a high grade phyllodes tumor, metaplastic carcinoma, fibrosarcoma, leiomyosarcoma, and MFH of the breast are to be ruled out completely. Dermatofibrosarcoma protuberans, fibromatosis, and myoepithelial carcinoma of the breast are other important tumors for differential diagnoses.^{1,2} A thorough, adequate sampling and immunohistochemistry is needed to carefully exclude these differentials.^{2,3} Tumor cells of mammary sarcoma-NOS type are negative for most the immune markers. These include several epithelial markers such as pancytokeratins, CK 5/6, CK 14, CK17, CK 34 β E12, CD34, SMA, desmin, h caldesmon, ER, and PR.^{2,3} Vimentin is strongly positive in all cases of mammary sarcoma-NOS. Striking immune features of these tumors are their positivity for CD10 and epidermal growth factor receptor

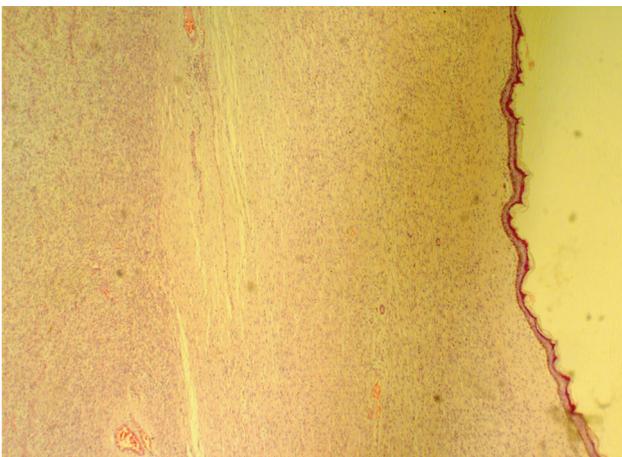


Figure 1. Tumor cells in sheets and fascicles under the skin in the breast. (Hematoxylin and eosin, 10 \times).

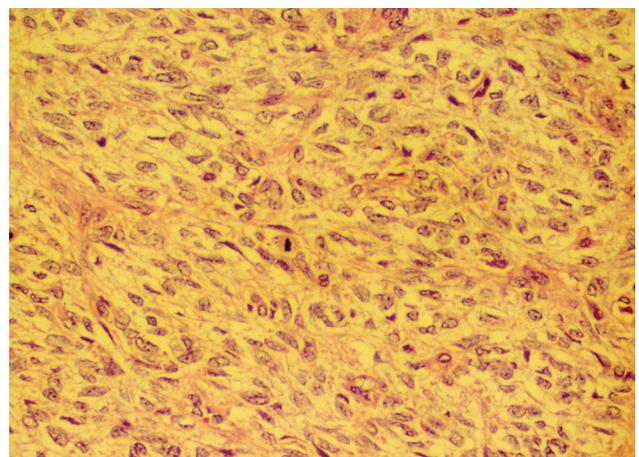


Figure 2. Spindle shaped tumor cells with nuclear pleomorphism, mitotic activity, and interspersed collagen. (Hematoxylin and eosin, 40 \times)



Figure 3. Immunohistochemical study showing positivity for vimentin in malignant cells. (Vimentin immunostain, 20×)

(EGFR-1). CD29, SMA, p63, and calponin may show focal positivity in a few cases.^{2,3}

In the breast, CD10 or the common acute lymphoblastic leukemia antigen (CALLA), a cell surface neutral endopeptidase and member of the metalloprotease family is known as a marker for myoepithelial cells.^{5,6} Moderate to strong expression of CD10 is also seen in cystosarcoma phyllodes.⁷ Malignant phyllodes tumors consist of a predominant mesenchymal component and a benign epithelial component. The mesenchymal component simulates pleomorphic sarcoma which requires an extended sampling to demonstrate the presence of ductal involvement.

Immunochemistry usually points out the positivity of stromal cells for vimentin, actin, CD34 and Bcl-2.^{7,8} Myoepithelial lesions are also positive for SMA, S100, CK 5/6 and high molecular weight keratin.² Metaplastic carcinoma is biphasic and contains both a carcinomatous component and heterogeneous sarcomatous component which show co-expression of S100, vimentin and cytokeratin.^{1,2} Fibrosarcoma is a highly cellular spindle cell tumor that displays an interdigitating fasciculated herringbone pattern. Leiomyosarcomas exhibit similar morphological features but are usually positive for desmin, SMA, and h-caldesmon.¹ MFH of the breast is extremely rare and most cases of primary MFH in the breast may actually represent undifferentiated sarcoma

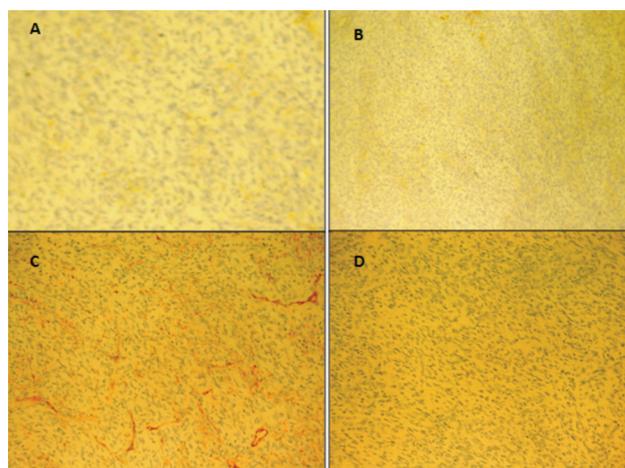


Figure 4. Immunohistochemical study of: A) smooth muscle actin (SMA), B) pan cytokeratin (panCK), C) CD34, and D) estrogen receptor (ER) that indicated negativity in the malignant cells. (10×)

of the breast that cannot be further classified. MFH is composed of pleomorphic spindle cells with abundant mitosis and inflammatory cells along with histiocyte-like cells. Cells in MFH may be actin, desmin, factor VIII, α 1antitrypsin, and lysozyme positive.² Dermatofibrosarcoma protuberans is composed of relatively monomorphic spindle cells that form a storiform pattern around the sweat glands of the deep dermis and subcutaneous fat vacuoles. The tumor cells show strong immunoreactivity for CD34.¹ Fibromatosis consists of uniform, plump proliferating spindle cells with low mitosis with varying amounts of collagenization and focal myxoid areas.^{1,2}

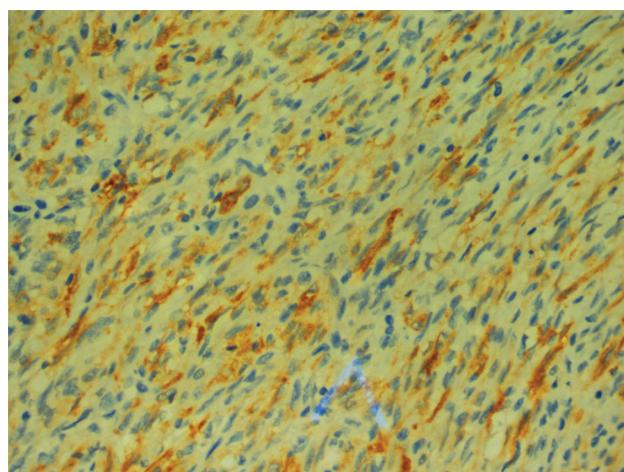


Figure 5. Immunohistochemical study of CD10 showing moderate positivity for malignant cells. (CD10 immunostain, 40×).

Breast sarcomas differ totally from epithelial breast cancers and require different management. The first-line treatment is surgical excision with adequate margins, with small survival advantages shown for post-operative radiotherapy.⁹ High EGFR-1 expression in these undifferentiated sarcomas can be evaluated further for molecular target therapy. The prognosis of mammary sarcoma is difficult to determine because of the small number of reported cases. However a study has associated CD10 expression in stromal cells in breast carcinoma with poor prognosis, estrogen receptor negativity and high grade tumor.¹⁰

Conflict of Interest

No conflict of interest is declared.

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