

# Stromal Sarcoma of the Breast: A Case Report

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We report a case of a 36-year-old lady who presented with a huge fungating tumour that involved the entire right breast. The tumour was diagnosed histologically as undifferentiated primary stromal tumour of the breast with axillary lymph node metastasis. We review the literature of this rare malignant tumour. Stromal sarcomas of the breast lack epithelial participation, and diagnosis of these tumours can be difficult. Genome-wide expression profiling is currently used to determine the cell of origin of most sarcomas. Surgery offers the best therapeutic option. Adjuvant radiotherapy is not very beneficial, while chemotherapy has, to date, no established role in the management of this disease. The prognosis is dismal for patients with lymph node involvement. The size of the tumour has a lesser bearing on outcome. [*Asian J Surg* 2004;27(4):339–41]

## Introduction

Soft-tissue sarcoma usually arises from the deep fascia or skeletal muscles of the extremities. Primary breast sarcoma not associated with other malignancies or previous irradiation is extremely rare.<sup>1,2</sup> Management remains controversial because of the rarity of the disease, and various authors hold different opinions as to how breast sarcomas should be managed. Furthermore, histological grading correlates poorly with clinical behaviour.<sup>3</sup> Advances in diagnostic methods, such as immunohistochemistry and electron microscopy, have enabled more accurate identification of the cellular origin of specific tumours.<sup>4</sup> In this report, we present a case of primary stromal tumour of the breast and review the relevant literature.

## Case report

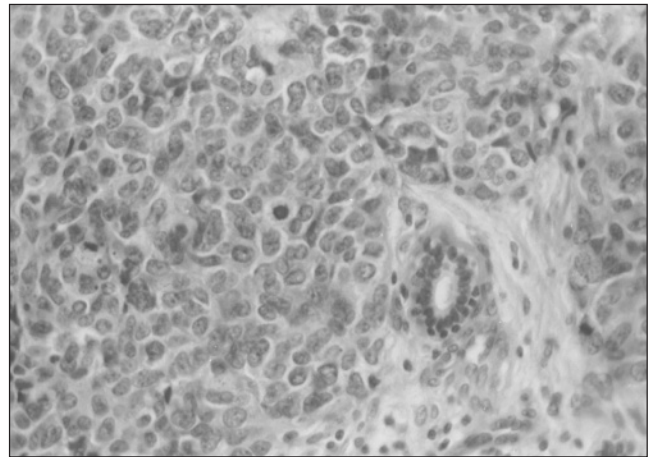
A 36-year-old lady gave a history of a palpable mass in the right breast for 3 years. She was nulliparous and had no family history of breast cancer. There was no history of breast implants or irradiation. On physical examination, the patient was pale. She had a temperature of 38°C, blood pressure of

110/70 mmHg and a pulse of 110 bpm. Breast examination revealed a huge, fungating, irregular mass involving the entire right breast. It was approximately 35 × 25 cm with areas of haemorrhage and necrosis (Figure 1). The right axillary lymph nodes were palpable. The contralateral breast was normal. Haemoglobin was 5.3 g/dL. Chest roentgenography, bone scintigraphy, abdominal ultrasound and computed tomography of the chest, abdomen and brain were all normal. The patient underwent right mastectomy with axillary lymph node sampling. The wound was covered with a split-thickness skin graft. The postoperative course was uneventful, and the graft took completely. Microscopic examination showed a poorly differentiated malignant tumour with large areas of necrosis (Figure 2). Some areas showed entrapped atrophic breast parenchyma. Normal and abnormal mitotic figures were seen in the range of 8–9/high-power field. Immunohistochemistry showed that all epithelial markers were negative except for vimentin (Figure 3), and there was a weak reaction for smooth muscle antigen; all other mesenchymal markers tested were negative. Lymph nodes showed extensive tumour deposits with large areas of necrosis. The tumour was diagnosed as undifferentiated primary stromal sarcoma with extensive lymph node metastasis.

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**Figure 1.** Huge fungating irregular mass involving the entire right breast with areas of haemorrhage and necrosis.



**Figure 2.** Poorly differentiated malignant tumour with frequent normal and abnormal mitotic figures (8–9/high-power field). Some areas show entrapped atrophic breast parenchyma.

## Discussion

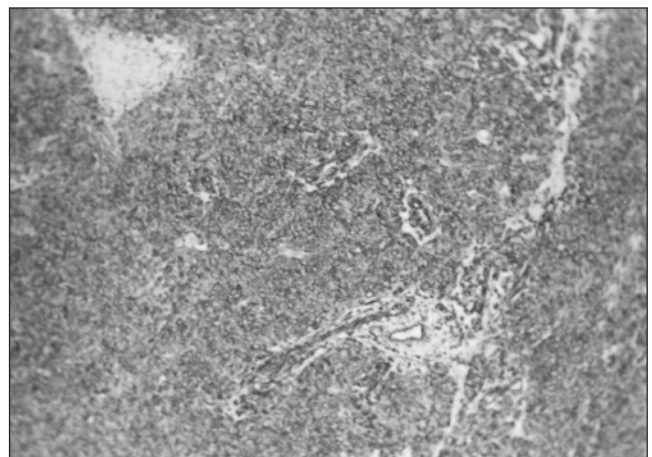
Breast sarcoma is a malignant neoplasm that arises from mesenchymal tissue and is part of a wide spectrum of connective tissue tumours, accounting for less than 1% of malignant breast lesions.<sup>5,6</sup> The incidence, histological type and clinical course of sarcomas of the breast are not well established. This is due to the rarity of these tumours and a lack of definition in various reports. Malignant phyllodes, lymphomas and even carcinomas with pseudo-sarcomatous changes have been considered by some authors to be breast sarcomas.<sup>7</sup> These neoplasms, therefore, pose a significant diagnostic and therapeutic challenge.

Stromal sarcomas of the breast were first defined by Berg et al in 1962.<sup>8</sup> They excluded malignant phyllodes, lymphoma and angiosarcoma from their study. They described 25 cases of homogeneous tumours with fibrous, myxoid and fatty patterns seen on light microscopy. The tumours were named stromal sarcoma of the breast because they were considered normal variants of mammary duct stroma. If the morphological features of a specific type of sarcoma are related to its cell of origin, e.g. “liposarcoma, fibrosarcoma” a more meaningful classification for prognosis and therapy may be proposed. The term “stromal sarcoma of the breast” should be reserved for tumours lacking an epithelial component on thorough histopathological sampling in which the cell of origin cannot be determined.

Recently, genome-wide expression profiling has been used to improve classification of soft-tissue sarcomas. Molecular characterization of these tumours using a cDNA expression microarray identified a group of genes associated with malig-

nant soft-tissue sarcoma such as fibrous histiocytoma and liposarcoma. This technique may help in elucidating the cell of origin of many of these tumours and may lead to recognition of new diagnostic and/or prognostic markers with therapeutic implications.<sup>9</sup> The most commonly described breast sarcomas are fibrosarcoma, liposarcoma and undifferentiated high-grade sarcoma.<sup>2,4,10</sup> Other sarcomatous lesions such as leiomyosarcoma and sarcoma with bone and cartilage are rare.<sup>6</sup> The aetiology of breast sarcoma is unknown. It has been suggested that a possible association exists between implantation of a silicon prosthesis and breast sarcoma, but this has not been proven. Angiosarcoma may be induced by radiation.<sup>11</sup>

Breast sarcoma may spread by direct invasion and/or via blood. Axillary lymph node involvement is very rare and is usually associated with end-stage disease.<sup>12,13</sup> In our patient,



**Figure 3.** Strong vimentin staining.

there was no distant metastasis, despite the advanced nature of the primary tumour and extensive involvement of axillary lymph nodes. Nodal disease is known to carry a poor prognosis, tumour contour being an important prognostic factor. Tumours with pushing margins seem to have a better prognosis than those with infiltrating borders; tumour size is of less prognostic value. Lesions with a high mitotic rate have a poor prognosis.<sup>14</sup> Simple mastectomy with attainment of clear resection margins gives optimal results.<sup>15</sup> Axillary lymphadenectomy is important to ensure local clearance of the primary tumour with the removal of clinically involved lymph nodes.<sup>16</sup> Adjuvant radiotherapy may be indicated when the primary tumour is bulky and/or when local clearance is not considered satisfactory.<sup>17</sup> The role of adjuvant chemotherapy has yet to be defined.<sup>3</sup> Breast sarcomas do not have hormonal receptors and adjuvant hormonal therapy is, therefore, ineffective.<sup>18</sup>

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