

# Primary stromal sarcoma of the breast

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## SUMMARY

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*Primary sarcomas of the breast are rare and there are only a few hundred cases reported in the literature; hence, their significance is primarily educational. We report a case of a 57-year old patient who developed breast sarcoma without previous history on any breast diseases.*

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**Key words:** Breast Neoplasms; Sarcoma; Diagnosis; Immunohistochemistry; Antigens, CD

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## INTRODUCTION

Breast cancer is the most common cancer of female population worldwide. In Cancer Statistics 2010, breast cancer remains the heading type of cancer with an estimate of 28% in American women (1). The vast majority of breast cancers are carcinomas, while breast sarcomas are few (2-5). Sarcomas represent less than 1% of all primary breast malignancies end less than 5% of all sarcomas (6-8) and because of their rarity, they are not commonly encountered by the members of medical profession.

Primary breast sarcomas derive from breast mesenchymal tissue. The annual incidental rate is 44.8 new cases per 10 million women (9). There is still no consensus according to the exact definition of breast sarcoma. Some authors excluded cystosarcoma phyllodes from their studies due to its epithelial components (6, 10, 11), but others do not distinguish between other subtypes of breast sarcoma and cystosarcoma because of the similar survival and clinical course (7, 8, 12, 13).

Almost every previous reference on this entity in the literature is in form of small case report series. In almost all cases, patients had been diagnosed clinically as having a breast carcinoma and the correct tissue diagnosis was established by the histology (14-16).

We report a case of primary stromal sarcoma of the breast.

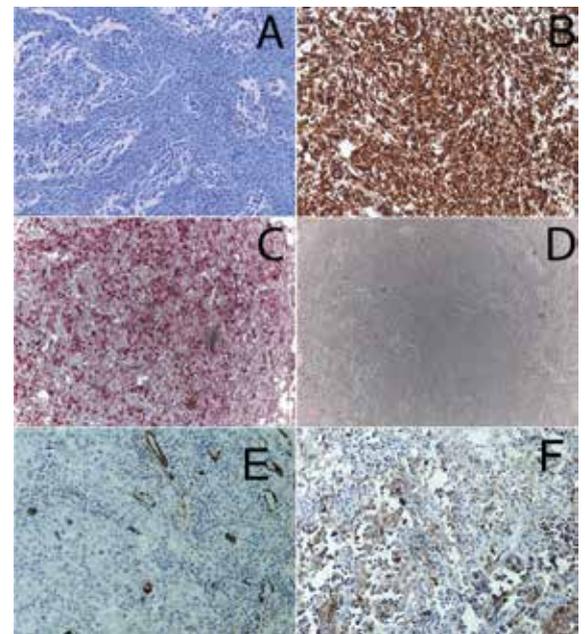
## CASE PRESENTATION

A 57-year old female patient presented to the surgical department with a complain of progressive swelling of the left mammary gland without pain. There was no history of previous breast trauma, bleeding, or family history of breast cancer. On examination there was a single 4x5.5x3.5cm mass, firm and nontender in the upper right quadrant. There was slight retraction of the overlying skin. Excision biopsy was done to confirm the suspicion of carcinoma of the breast.

Grossly, the cut section revealed firm, discreetly lobulated, infiltrative unencapsulated gray-white tumor, measuring 4x5.5x3cm. There were no necrotic areas, areas of haemorrhage or calcification. Surgical margins were tumor negative.

Microscopically, hematoxylin and eosin (HE) stained section revealed whorls and nests of spindle cells with mild differences in cellular size. The nuclei were round to oval with marked hyperchromasia. There were neither prominent nucleoli, nor cytoplasmic granules. Focally, tumor cells were round to oval with abundant cytoplasm, separated by delicate fibrous bands with lymphocytic infiltrates and were infiltrating adipous tissue. The suspicion on dedifferentiated carcinoma was not supported by immunohistochemical findings. The APAP method was used to analyze: vimentin, desmin,

cytokeratin, MAC, pan T, pan B, proliferating cell nuclear antigen (PCNA), epithelial membrane antigen (EMA), CD34, CD68 and S100. The tumor was found to be vimentine and CD68 positive. Very strong expression for PCNA was demonstrated (Figure 1).



**Figure 1.** A – Tumor is composed of whorls and nests of spindle cells. Trichrome x40; B – Vimentin x40 was strongly positive; C – Strong immunoreactivity to CD68 x100; D – Cytokeratin staining was negative x100; E – Actin x 20 was negative; F – S-100 x 20 was negative

## DISCUSSION

Primary sarcomas of the breast are rare and there are only a few hundred cases reported in the literature. Cystosarcoma phyllodes are much more common and their behavior, management and treatment are different from pure sarcoma. The reported case was the first stromal sarcoma diagnosed in our laboratory during the period of 32 years. The defect in experience was prevailed over by using appropriate antibodies for immunohistochemical method as recommended (15). The controversy still exists on the term „stromal sarcoma“. Berg and coworkers have unified all breast sarcomas other than cystosarcoma phyllodes under stromal sarcoma to indicate their origin from breast stroma (17). In 1985, Callery et al. proposed the term „stromal sarcoma“ for tumors arising from specialized hormone sensitive breast stroma. It is now recommended to use histological description by cell of origin of the neoplasm (18). The most common sub-types of breast sar-

comas are: malignant fibrous histiocytoma, fibrous sarcoma, angiosarcoma and spindle cell sarcoma. Several other sub-types (leiomyosarcoma, liposarcoma, rhabdomyosarcoma, osteosarcoma, chondrosarcoma, synovial sarcoma and neurosarcoma) have been described as smaller percentages of case series or as case reports. (5, 8, 14). The presenting case is stromal sarcoma-fibrous sarcoma, according to the cell of origin.

Although rare, one must have in mind the possibility of such neoplasm in the breast any time there are spindle cells in the sections of the tumor. The breast is the place of metastatic sarcomas (19, 20) as well as the sarcomas secondary to radiotherapy (21).

The risks of developing breast sarcoma are largely unknown, but those often proposed are: external beam radiation of the breast or chest wall; chronic lymphedema of the breast and arm (especially for angiosarcomas); pre-existing fibroadenoma or hereditary diseases, like neurofibromatosis or Li-Fraumeni syndrome (22).

It appears that breast sarcomas behave like extremity sarcomas and hence, it is rational for using the same treatment protocol (15, 22). The first therapeutic line is surgical resection with postoperative radiotherapy with or without chemotherapy. In the present case, the conservation of the breast was done with radiotherapy and without chemotherapy. The patient was followed for 22 months and was in good condition.

### Conflict of interest

We declare no conflicts of interest.

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