



## Case Report

## Primary breast stromal sarcoma: A rare case report

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## ABSTRACT

Primary sarcoma of the breast comprises less than 1% of all the breast malignancies. It is a locally aggressive neoplasm with only a few hundred cases published till date. The index case is being reported due to rarity of this entity. Breast sarcoma should always be considered whenever spindle cells are seen in the histological sections. The main diagnostic confusion arises with malignant phyllodes tumor and metaplastic carcinoma from which distinction is important from prognosis as well as treatment point of view. Herein, we report a case of a 47-yr-old female, who presented with a right-sided breast lump which was diagnosed as suspicious of carcinoma breast on fine needle aspiration (FNAC). Thereafter, modified radical mastectomy was performed, and on histopathology, a possibility of fibro-histiocytic tumor of intermediate malignancy was rendered, which was subsequently confirmed by immunohistochemistry (IHC).

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## 1. Introduction

Breast cancer is the most common cancer among women worldwide. Most breast cancers are epithelial in origin while only a small minority, the breast sarcomas, arise from the mesenchymal component of the breast. With an annual incidence of 4.6 cases per million women, breast sarcomas comprise less than 1% of all breast malignancies. They may exist as primary tumours that have heterogeneous histological subtypes and present at a slightly early age (mean 40 years) or may develop later in life (45-50 years of age) following radiotherapy to the chest wall (secondary tumours).<sup>1</sup> Primary breast sarcomas are infrequent as compared to secondary ones, which makes the index case an unusual entity.<sup>2</sup>

## 2. Case Report

A 47-year-old female presented to the Surgery Outpatient Department in our hospital with the complaint of a painful lump in the right breast since 6 months. On examination, a

firm, tender, 10×10 cm lump, with irregular margins, was palpated in the upper outer quadrant of right breast. There was no history of nipple discharge or nipple retraction. The overlying skin was apparently normal. No history of breast trauma or any other significant family history was elicited.

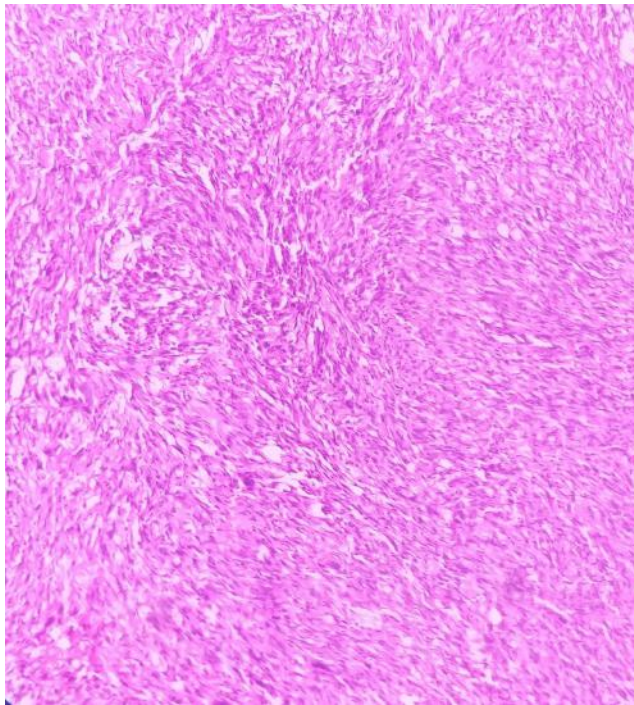
Patient had previously undergone Fine needle aspiration cytology (FNAC), a mammography and a Magnetic Resonance Imaging (MRI) breast elsewhere. FNAC report showed the lump as suspicious of carcinoma breast. MRI breast with contrast revealed a lesion measuring 7 × 6.7 cm, (BIRADS III/IV) in the right breast while mammography right breast suggested the possibilities of a complex cyst/ large fibroadenoma/ phyllodes tumor with the impression of BIRADS III category. Also, a previous biopsy done in a reputed hospital suggested the diagnosis of leiomyosarcoma.

Subsequently, a modified radical mastectomy was planned and the specimen was sent for histology. On gross examination, the cut section revealed a vague 5×6×2.5 cm cavitory lesion, filled with blood clots and necrotic debris, along-with a solid area measuring 1 cm in diameter. Totally, 12 lymph nodes were isolated from the specimen.

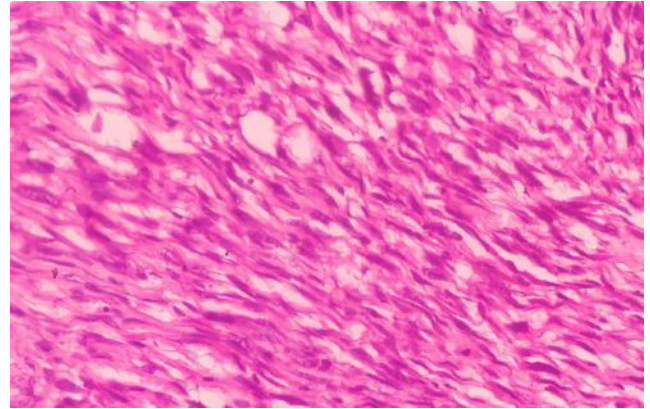
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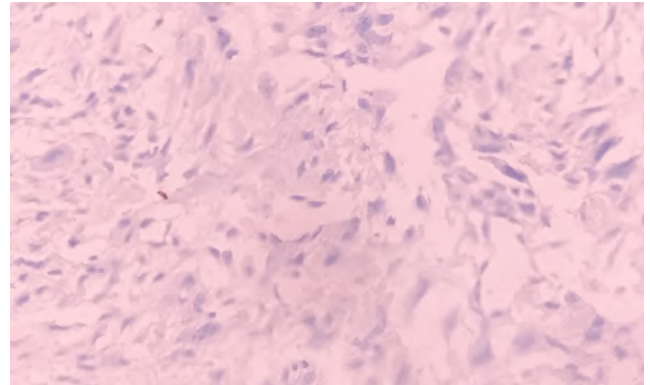
On microscopic examination (Figures 1 and 2), hematoxylin and eosin stained sections revealed a tumor comprised of spindle to plump, ovoid cells and epithelioid to polygonal cells displaying moderate pleomorphism. About 50% of the tumor area showed degenerative/necrotic changes. The individual tumor cells had moderate to abundant cytoplasm with pleomorphic and hyperchromatic nuclei and inconspicuous to prominent nucleoli. Fair number of giant cells and few bizzare cells were noted. Stroma showed delicate collagen fibrils with focal areas of myxoid change. The tumor cells were seen infiltrating the adjacent adipose tissue to some extent. Mitosis was less than 5/10 High power field (HPF). Surgical margins were free of tumor. Lymph nodes showed no evidence of metastasis from tumor. Possibility of fibro-histiocytic tumor of intermediate malignancy was considered. Immunohistochemistry (IHC) was advised to rule out malignant phyllodes tumor and metaplastic carcinoma. Consequently, immunohistochemical panel was applied including cytokeratin (CK), vimentin, p63, CD10, CD34, Smooth Muscle Actin (SMA) and desmin (Figures 3, 4, 5, 6 and 7). Vimentin and SMA were diffusely positive in tumor cells, CD10 and desmin were focally positive while CK, p63 and CD34 were negative. Further imaging studies were done and they did not reveal any mass elsewhere in the body. Based on the above findings a final diagnosis of primary breast sarcoma with focal smooth muscle differentiation was made.



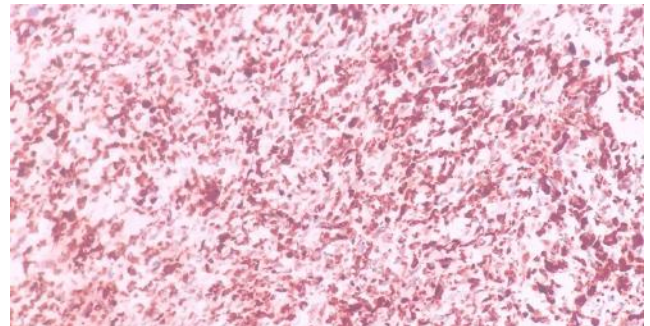
**Fig. 1:** Photomicrograph showing tumour cells arranged in whorls and fascicles (H&E, 100X)



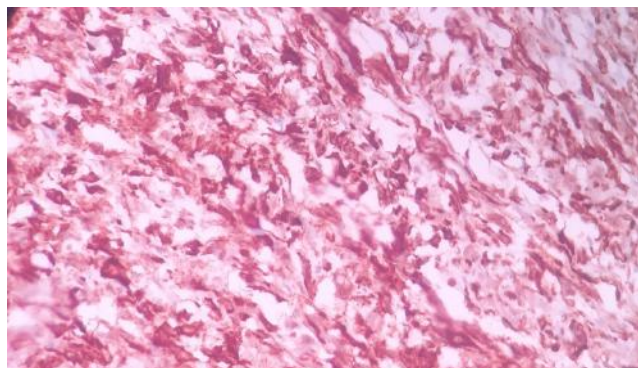
**Fig. 2:** Photomicrograph showing spindle to plump ovoid cells displaying moderate pleomorphism (H&E, 400X)



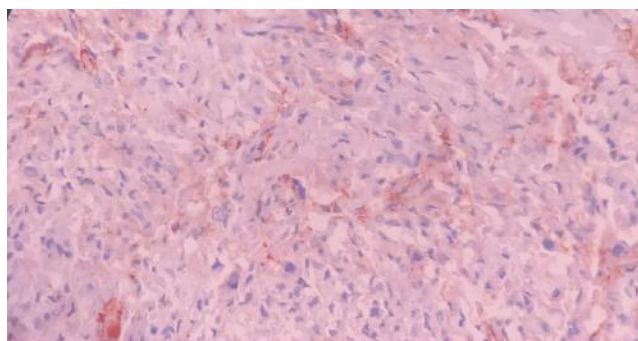
**Fig. 3:** Photomicrograph showing negative cytokeratin staining in spindle cells



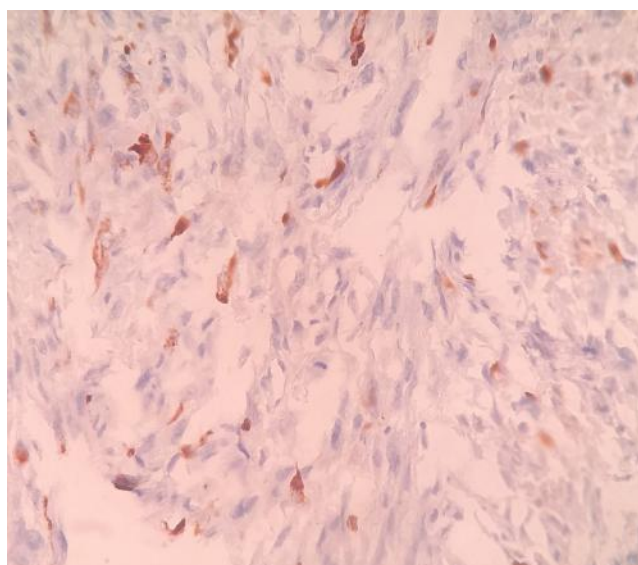
**Fig. 4:** Photomicrograph showing tumour cells diffuse positivity for vimentin



**Fig. 5:** Photomicrograph showing diffuse positivity for SMA



**Fig. 6:** Spindle cells showing focal positivity for CD10



**Fig. 7:** Few spindled tumour cells showing positivity for Desmin

### 3. Discussion

Breast sarcoma is a rare and aggressive, malignant neoplasm that presents in women of age 48-60 years with mean reported age of 45 years. Observed to be mostly unilateral, it does not have a side predilection and can occur equally in both right and left breast. It usually presents as a lump, often painful, with average reported duration of six months. Mean stated size is 5.7 cm and it is noted that angiosarcomas are larger in size. Contralateral and extramammary involvement has been reported in scattered case reports. Barnes et al observed distant metastasis to lungs, liver, spleen and skin. No lymph node metastasis have been recounted.<sup>3,4</sup>

The heterogeneity and rarity of this neoplasm renders its etiology difficult to comprehend. Some factors associated with the increased risk of development of breast sarcoma are previous radiotherapy to the breast and chest wall, previous history of fibroadenomas, chronic lymphedema and some genetic diseases like Li-Fraumeni syndrome and neurofibromatosis.<sup>5</sup>

Primary breast sarcoma is a heterogeneous entity with various subtypes. The most common subtypes are malignant fibrohistiocytoma, angiosarcoma, spindle cell sarcoma and fibrosarcoma. Less common varieties include rhabdomyosarcoma, leiomyosarcoma, liposarcoma, stromal sarcoma, synovial sarcoma, osteosarcoma, neurosarcoma, hemangiopericytoma and chondrosarcoma. Angiosarcoma is the most common radiation-induced sarcoma of the breast.<sup>6</sup> Breast sarcomas should always be differentiated from other spindle cell tumors of breast like metaplastic or sarcomatous carcinomas.<sup>7</sup>

Breast sarcoma tends to be a rapidly expanding mass with pushing border, hence it appears as an oval, hypervascular lesion on imaging studies with circumscribed or ill-defined margins, although they show invasion microscopically. Imaging studies like mammography, ultrasound and MRI are useful adjuncts to diagnosis. On mammography, they appear as non-calcified oval masses and on sonography as oval, hypoechoic lesions with posterior shadow. These findings can sometimes be overlapping with that of phyllodes tumour but they are in stark contrast with that of infiltrating carcinoma of breast which has stellate/ill-defined borders.<sup>8</sup>

Diagnosis by FNAC is troublesome due to nonspecific cytomorphologic features. Smears usually show dispersed population of malignant, pleomorphic, sometimes bizarre, spindle cells with the absence of any epithelial component and yields a differential diagnosis of malignant spindle cell tumor, malignant phyllodes tumor and metaplastic carcinoma (spindle/sarcomatoid variant).<sup>9</sup>

Hence the most reliable diagnostic modality for this entity is histopathology supplemented by immunohistochemistry (IHC). Since breast sarcoma is a heterogenous entity, it is imperative that a pathologist

should be familiar with an array of histomorphological features of various subtypes. Malignant fibrous histiocytoma consists of fibroblast and histiocyte like cells, along with pleomorphic giant and inflammatory cells and is vimentin and CD68 positive on IHC. Leiomyosarcoma shows fascicular growth pattern comprising of atypical spindle cells with cigar-shaped hyperchromatic nuclei and eosinophilic cytoplasm and is immunoreactive for SMA and desmin. Rhabdomyosarcoma shows an alveolar growth pattern along with the presence of rhabdomyoblasts and positivity for desmin, myogenin, and MyoD1 on IHC.<sup>10</sup> Fibrosarcoma consists of sheets and fascicles of atypical spindle cells, forming storiform and “Herring” bone pattern with diffuse immunoreactivity for vimentin.<sup>11</sup> Angiosarcoma is characterized by irregular vascular network with hyperchromatic and irregular nuclei and is confirmed by immunohistologic staining for CD31, the most sensitive and specific indicator of angiogenic proliferation.<sup>12</sup>

In the index case, histopathology revealed spindle to plump, ovoid cells and some epitheloid to polygonal cells displaying moderate pleomorphism. Stroma showed delicate collagen fibrils with focal areas of myxoid change. Absence of a biphasic tumor, with leaf-like architecture and epithelial components on extensive sectioning ruled out the possibility of cystosarcoma phyllodes in our case. The cytokeratin negativity on IHC and absence of carcinomatous component ruled out the possibility of metaplastic carcinoma. This distinction is important in terms of both prognosis and treatment of patient.

Breast sarcomas clinically behave like sarcomas in the extremities and they should be treated with the similar protocol.<sup>5</sup> The first line of treatment is surgical resection. The treatment of choice is mastectomy with postoperative radiotherapy with or without chemotherapy. As breast sarcomas rarely metastasize through lymphatics, axillary dissection is generally not performed.<sup>13</sup>

Age, size, grade, histological type, involvement of more than one quadrant, and tumor spread are considered the most significant prognostic factors in cases of primary breast sarcoma; and are found to be in common with that of soft tissue sarcomas. Tumors with size measuring more than 5 cm are associated with worse prognosis. Yin M et al observed fibrosarcoma and liposarcoma to be associated with better survival, while osteosarcoma was associated with a worse outcome whereas according to Zelek L et al, angiosarcoma was associated with worse prognosis due to its high recurrence and its infiltrative nature.<sup>4,5,8,14,15</sup>

#### 4. Conclusion

Breast sarcoma should always be considered whenever spindle cells are seen in the histological sections. It is a very rare neoplasm and behaves in a very aggressive manner. Hence the importance of an early diagnosis and immediate

treatment.

#### 5. Key Points

1. Primary sarcoma of the breast comprises less than 1% of all the breast malignancies. It is a locally aggressive neoplasm with only a few cases published till date which makes the index case an unusual entity.
2. Herein, we report a case of Primary Breast Stromal Sarcoma which was diagnosed by using a combined approach of histopathology and immunohistochemistry.
3. Differentiation from malignant phyllodes tumor and metaplastic carcinoma is necessary from prognosis as well as treatment point of view.
4. Poor prognosis and behaves in a very aggressive manner. Hence the importance of an early accurate diagnosis and immediate treatment.

#### 6. Consent

Informed consent was obtained from the patient for publication of this case report.

#### 7. Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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