# Cash Report

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# Primary fibrosarcoma of breast: a rare case presentation

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#### **ABSTRACT**

Primary Fibrosarcoma of breast is an extremely rare tumor accounting for <1% of all the primary breast malignancies and <5% of all the sarcomas. Breast fibrosarcoma is a rare tumor characterized by marked rapidity of growth. Diagnostic investigations like FNAC, mammography and USG are relatively insensitive and histopathological diagnosis is mainstay and it is usually a diagnosis of exclusion. Surgical therapy is most effective treatment and chemotherapy should be considered for large tumors with high mitotic rates. In all the diagnosis and management should be multidisciplinary team approach. In this case report we are reviewing a case of primary Fibrosarcoma of breast in a 45 years old female presenting with a rapidly growing painless mass in left breast since 3 months. Repeated FNAC were inconclusive and hence incisional biopsy was done which revealed Fibrosarcoma. Modified radical mastectomy was done with axillary clearance and histopathology reports further confirmed the diagnosis. Being a very rare tumor, the primary aim of this presentation is to provide an overview of literature on the diagnosis and management protocols for breast fibrosarcoma patients.

Keywords: Fibrosarcoma, FNAC, Incisional biopsy, Modified radical mastectomy

#### INTRODUCTION

Sarcomas of breast are extremely rare breast tumors accounting for <1% of all the primary breast malignancies and < 5% of all the sarcomas. They arise from mesenchymal tissues of the breast. The annual incidence is about 44.8 new cases per 10 million women. The exact definition of breast sarcoma is still not uniform. Some authors excluded cystosarcoma phylloids from their studies due to presence of epithelial components but others do not because of their similar clinical and surgical course. Most publicized articles are limited to small retrospective reviews and case reports, making clinicopathological study difficult.

The overall incidence of primary fibrosarcoma of breast is not known nor has it been reported in literature. <sup>10</sup> In a review conducted by Poland et al of 25 cases of primary breast sarcoma, during 80 yrs. period, only 4 cases of

fibrosarcoma reported.<sup>1</sup> Terrier et al, in his study of 33 cases of sarcoma of breast only 2 cases of fibrosarcoma were reported.<sup>2</sup> Blanchard et al, in his study of 55 cases of sarcoma only 2 cases of fibrosarcoma were reported.<sup>11</sup>

The peak incidence of fibrosarcoma occurs in  $5^{th} - 6^{th}$  decade of life, but for fibrosarcoma of breast the peak incidence is unknown. Roberson Et al reviewed the literature of 49 reported patients with fibrosarcoma of breast and half of them were 41-60 years of age. <sup>12</sup>

The most common presentation of fibrosarcoma of breast is rapidly progressing painless lump in breast which attains a large size. Metastasis from breast fibrosarcoma commonly spread haematogenously typically to lungs, bone and liver.<sup>1-3,9</sup> Lymph node metastasis is rare.<sup>3,13</sup>

The rarity of this pathology led to lack of consensus on the optimal treatment and hence a multidisciplinary approach is needed for diagnosis and treatment of patients.<sup>3,14</sup>

#### **CASE REPORT**



Figure 1: Clinical photograph of fibrosarcoma breast.

A 45 years old female patient,  $G_3$   $P_3$   $L_3$   $A_0$  in her third year of menopause presented with rapidly growing painless lump in left breast since 5 months. The lump was extending over whole of breast, measuring  $20 \times 15 \times 10$  cms in dimensions. It was firm to hard in consistency with nodular surface with reddish brown discoloration of overlying skin. There was fixity to skin but no fixity to pectoralis major muscle. The mass was involving the entire nipple-areola complex. There were no axillary lymph nodes palpable.

Repeated 3 FNAC of tumor came to be inconclusive and so incisional biopsy taken which was suggestive of fibrosarcoma of breast. USG of breast reveals solid hyperechoic mass with no cystic component. X-ray chest and USG abdomen did not reveal any secondary deposits in chest and abdomen.

After proper pre-anesthetic workup and fitness modified radical mastectomy was done with primary closure of defect. Specimen sent for histopathology examination confirming the diagnosis.

Grossly the tumor was big, nodular involving the whole breast tissue. Histopathology examination shows tumor mass composed of spindled shaped cells arranged in whorls and band pattern. Nuclei are hyperchromatic, pleomorphic with prominent nucleoli. Mitosis rate is increased. There was no duct like or epithelial gland like structure.

No recurrence at operative site or distant metastasis reported in follow up period of 6 months.



Figure 2: Gross appearance of tumor.

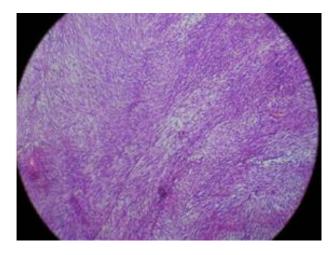


Figure 3: Microscopic appearance of tumor.

The second most proposed risk factor is chronic lymphedema of breast or arm. 19 (Especially for Angiosarcoma)

People with hereditary disease like neurofibromatosis or Li fraumani syndrome have an increased risk of developing soft tissue sarcoma. <sup>20</sup>

## Clinical presentation

The most common presentation of fibrosarcoma of breast is rapidly progressing painless lump n breast which attains a large size. Metastasis from breast fibrosarcoma commonly spread haematogenously typically to lungs, bone and liver. <sup>1-3,9</sup> Lymph node metastasis is rare. <sup>3,13</sup>

#### Diagnosis

Diagnosis on basis of FNAC is often difficult to interpret and has limited accuracy. So histopathological tissue examination is must for clinical diagnosis. On histopathological examination, breast sarcomas are

divided several subtypes- Angiosarcoma, Malignant fibrous histiocytoma, Stromal sarcoma, Spindle cell sarcoma and Fibrosarcoma. Among them Angiosarcoma is most common.

Cellular pleomorphism, mitotic numbers, stromal atypical and infiltrating borders have been found to be main prognostic factors.6,7,23

Mammography nonspecific demonstrating is nonspiculated dense mass without microcalcification. 21,24

On USG they appear as hyperechoic with no shadowing.<sup>25</sup> MRI may display rapid enhancement with washout characteristics of lobules.<sup>26</sup>

#### Staging

Table 1: AJCC system for soft tissue sarcoma<sup>27</sup>

Stage	Histological grade	Size	Location: Superficial or Deep	Systemic metastasis
ΙA	Low	<= 5 cm	Superficial	No
ΙB	Low	>5 cm	Superficial	No
II A	Low	>= 5 cm	Deep	No
II B	High	< 5 cm	Superficial or Deep	No
II C	High	>= 5 cm	Superficial	No
III	High	>= 5 cm	Deep	No
IV	Any	Any	Any	Yes

## **Surgery:**

For all breast sarcomas surgical resection is the first modality of treatment. In comparison to wide local excision, mastectomy was considered gold standard but recent study conflict this comparison and considered both of them equivocal.<sup>3,5,7,9,13</sup> For smaller localized breast sarcomas most of them recommended wide excision with greater than 3 cm normal tissue margin.<sup>28</sup> But to reduce the risk of local recurrence, mastectomy may be considered for those whose tumors are too large.

Breast sarcomas like any other sarcomas commonly metastasize haematogenously.<sup>29</sup> Fong Et al reported lymph node metastasis accounted for 2.7% in 1772 cases of soft tissue sarcomas including Angiosarcoma (13.5%), embryonal rhabdomyosarcoma (13.6%), epithelial sarcomas (16.4%).<sup>30</sup> As fibrosarcomas have significantly small proportion of all, conventional axillary clearance is not necessary.<sup>29</sup>

#### Radiotherapy

In contrast to surgical therapy, there is widespread disagreement for the benefits of radiotherapy in breast sarcomas.<sup>31</sup> Majority of studies did not find any benefit for adjuvant radiotherapy.<sup>1,7,11,32</sup> But McGowan et al and Johnstone et al demonstrated significant benefits for patients.<sup>3,33</sup> They recommended post-operative irradiation of at least 60 grays to whole tumor bed. It may help to reduce the local recurrence of tumor. Fibrosarcomas are relatively insensitive to radiotherapy.

#### Chemotherapy

The role of chemotherapy for breast sarcomas remains unclear, but adjuvant chemotherapy is worth a shot in patients with highly malignant sarcoma, with positive surgical margins or post-operative recurrence.<sup>34</sup> The most effective chemotherapy regimen is Adriamycin (ADM) + Ifosfamide (IFO).

## **Prognosis**

It is generally agreed that the prognosis of disease is related with tumor cell differentiation, tumor size and surgical approaches.<sup>29</sup> Large studies have demonstrated a 5 years disease free survival ranging from 44-66% and a 5 year overall survival ranging from 49-67%. <sup>2,3,5-7,9,13,35,36</sup>

### **CONCLUSION**

Breast fibrosarcoma is a rare tumor characterized by marked rapidity of growth. Diagnostic investigations like FNAC, mammography and USG are relatively insensitive and histopathological diagnosis is mainstay and it is usually a diagnosis of exclusion. Surgical therapy is most effective treatment and chemotherapy should be considered for large tumors with high mitotic rates. In all diagnosis and management multidisciplinary team approach.

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