

## Presentation of Low-grade Fibro Myxoid Sarcoma in Male Breast: Rare and Challenging Entity for Diagnosis - A Case Report with Review of Literature

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### ABSTRACT

Primary breast sarcomas are very rare tumors accounting <1% of primary malignant breast tumors and <5% of soft tissue sarcomas. They are seen in young adults with tendency for multiple recurrences. They can metastasize if incompletely excised, so the prognosis is poor. There is rarity because of lack of consensus and a few case reports have been reported till date. We report a case of 31-year-old male with history of left areolar swelling. He had history of excision biopsy one year back from same site which on histopathology was diagnosed as low-grade fibro myxoid sarcoma. The patient was asymptomatic for one year and now presented with history of swelling at scar site. The lumpectomy was done. There was no systemic dissemination. The tumor was histologically diagnosed as recurrence of low-grade fibro myxoid sarcoma. We review the literature about breast sarcomas and low-grade fibro myxoid sarcoma of the breast.

**Keywords:** Low grade fibro myxoid sarcoma, Primary breast tumors, Soft tissue sarcoma, Breast cancer, Recurrence.

### Introduction

Soft tissue sarcoma of the breast are very rare tumors and account for less than 1% of all primary malignant breast tumors and less than 5% of all soft tissue sarcomas [1]. Breast sarcomas have very high chances of recurrence, leading to poor prognosis. They can be primary or secondary. Primary sarcomas occur de novo and secondary sarcomas are associated with chronic lymphedema, radiotherapy or genetic syndromes like neurofibromatosis, Li-Fraumeni syndrome and familial adenomatous polyposis and as a malignant conversion of phyllodes tumour [1],[2]. Primary breast sarcomas occur in female patients exclusively and constitute less than 1% of all breast malignancies in both sexes.

In males, primary leiomyosarcoma, myxofibrosarcoma, fibrosarcoma, primary angiosarcoma, and undifferentiated pleomorphic sarcoma has been reported [3]-[6]. Clinically, this tumor occurs in young patients and has tendency for multiple recurrences, with a risk of metastasis if incompletely excised.

It appears as a slow growing painless mass. Myxofibrosarcoma is the most common differential diagnosis but is seen in elderly patients and has more prominent atypia with usually uniform myxoid stroma.

### Case Report

A 31 year old male came to our hospital with history of left areolar swelling. The patient had history of excision biopsy 2 year back and on histopathology it was diagnosed as low-grade fibro myxoid sarcoma. Immunohistochemical staining was negative for SMA & S-100. A PET-CT scan at that time showed ill-defined soft tissue density lesion measuring (1.7x1.3) cm at left areolar region, without significant radiotracer uptake. He also gave history of paralysis in the same year and NCCT scan of brain was done which came out to be normal. The patient gradually recovered. The patient was non-smoker but gave history of on & off drinking. The patient complained of

swelling at same left areolar region scar site now, which increased in size. There was no history of pain or any nipple discharge or any axillary lymphadenopathy. Repeat PET-CT was done which showed mildly FDG avid bilateral cervical level-II lymph node and non FDG avid mild thickening of left areolar region-post surgical/residual. Few small lymph nodes seen at right paratracheal, bilateral hilar and subcarinal region, with specks of calcification with no significant radiotracer uptake. Rest no abnormality was detected. We received a wide local excised lumpectomy specimen measuring (8x6x3) cm.

On serial sectioning single grey, white area with focal cystic areas identified measuring 2 cm in diameter. Tumor was 1.1 cm away from distal resection margin. Grossly, tumor was non-encapsulated with intermixed fibrous and cystic areas with gelatinous appearance. Microscopic examination showed cytologically bland spindle cells arranged in a whorled pattern in variably myxoid and fibrous stroma. The tumor showed no nuclear pleomorphism, necrosis or mitosis and it was reported as recurrence of low-grade fibro myxoid sarcoma. All resection margins as well as skin nipple areola complex were free of tumor.

### **Discussion**

Primary breast sarcomas are very rare. There is lack of sufficient consensus and clarity about the management guidelines for this entity. The most commonly used staging system for breast carcinomas is the American Joint Committee on cancer staging system for soft tissue sarcoma, which includes histological grade, tumor size, lymphatic involvement and distant metastatization [7]. The most common mode of dissemination is hematogenous typically to lungs, bone marrow and liver [8].

Lymphatic spread is uncommon and so axillary lymph node involvement is not a frequent finding [7]-[10]. The breast skin and nipple areola complex are rarely involved in the breast sarcomas [7]. Prognosis is based on tumor size, the presence of regional or distant metastasis and histological grade [8]. Detection of FUS translocation is a strong confirmatory finding for low grade fibro myxoid sarcoma [11].

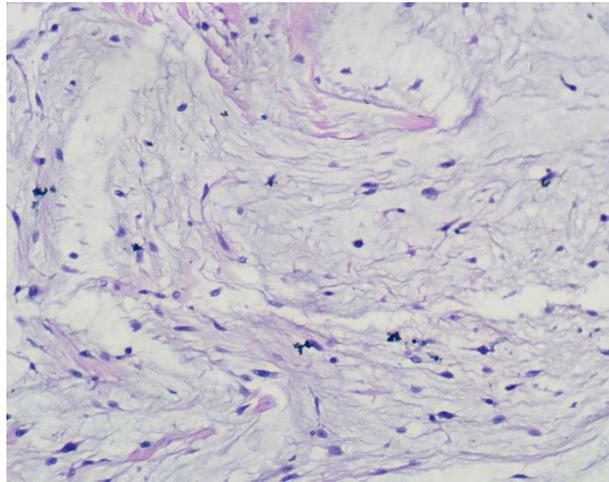
Low grade fibro myxoid sarcoma was first recognized by Evans in 1987 as a rare soft tissue tumor with a high metastatic potential, despite a benign histological appearance. It occurs frequently in young adults, slightly more often in men, presenting as a slowly growing painless mass usually in a subfascial location [12].

Zhang et al [13] in 2018 reported first case of LG-FMS in a male patient followed by Czopnik et al [12] in 2021. Diagnosis should involve a core biopsy. Excisional biopsy should not be performed unless previous attempts at diagnosis are unsuccessful [8],[14],[15]. The published literatures are very rare and there are no definite guidelines regarding diagnosis and management of this entity. Complete surgical resection with negative resection margins is although suggested. So, a cautious multidisciplinary approach should be used for management of such rare entity.

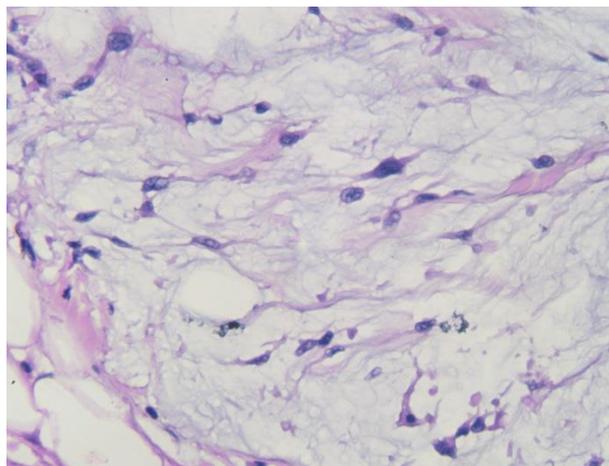
### **Conclusion**

Being a rare and aggressive tumour with rapid growth, surgical excision with resection margins is apparently the curative treatment. Though the role of adjuvant therapy remains controversial, but it can be tried in patients with high risk of recurrence. The prognosis is poor so a complete follow up of the patient for the subsequent years is recommended.

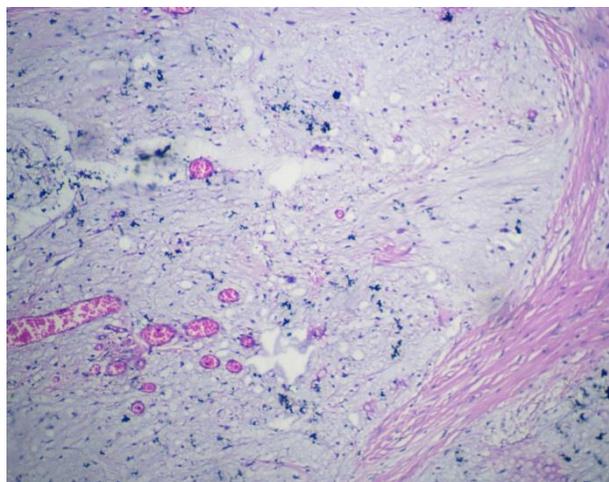
***Legends - low grade fibromyxoid sarcoma***



**Fig.1a.** Histology fibro-myxoid sarcoma 10x20 (H&E) Tumor showing myxoid areas with bland nuclei showing minimal atypia and low to moderate cellularity



**Fig.1b.** Histology fibro myxoid sarcoma 10x40 (H&E) Tumor showing minimal nuclear pleomorphism interspersed with myxoid and fibrous areas



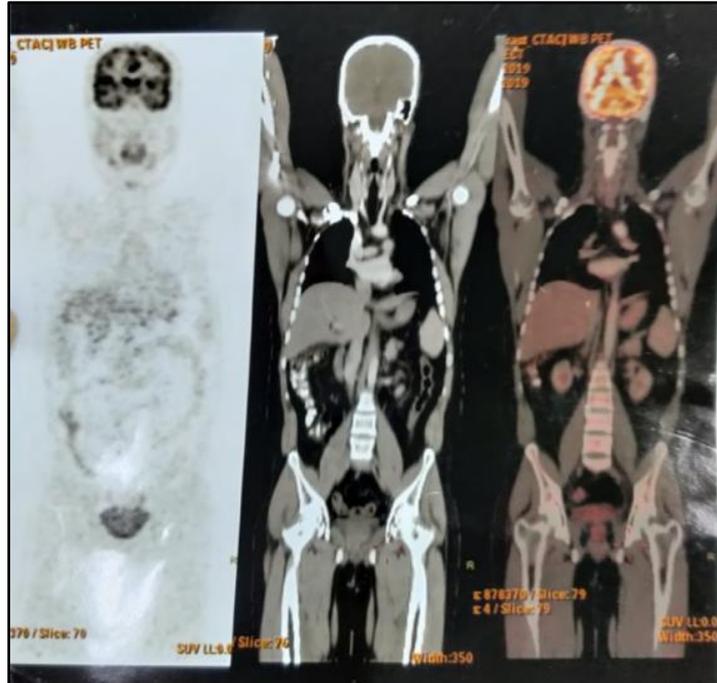
**Fig.1c.** Histology fibro myxoid sarcoma 10x10 (H&E) Tumor showing fibrous and myxoid areas with curvilinear vessels



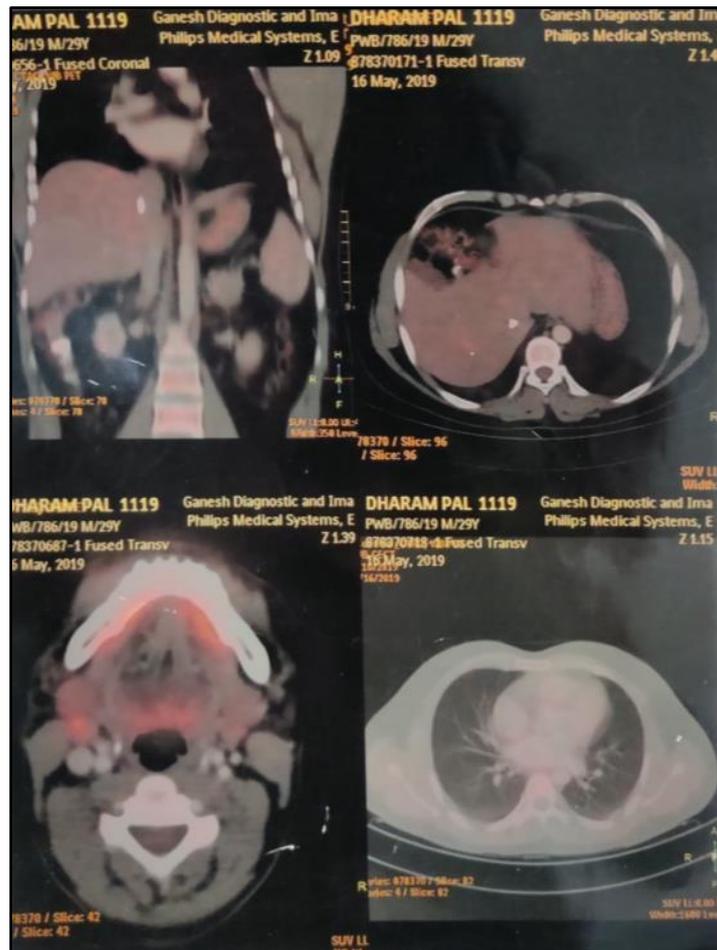
**Fig.2.** Gross image of breast showing glistening mucoid areas



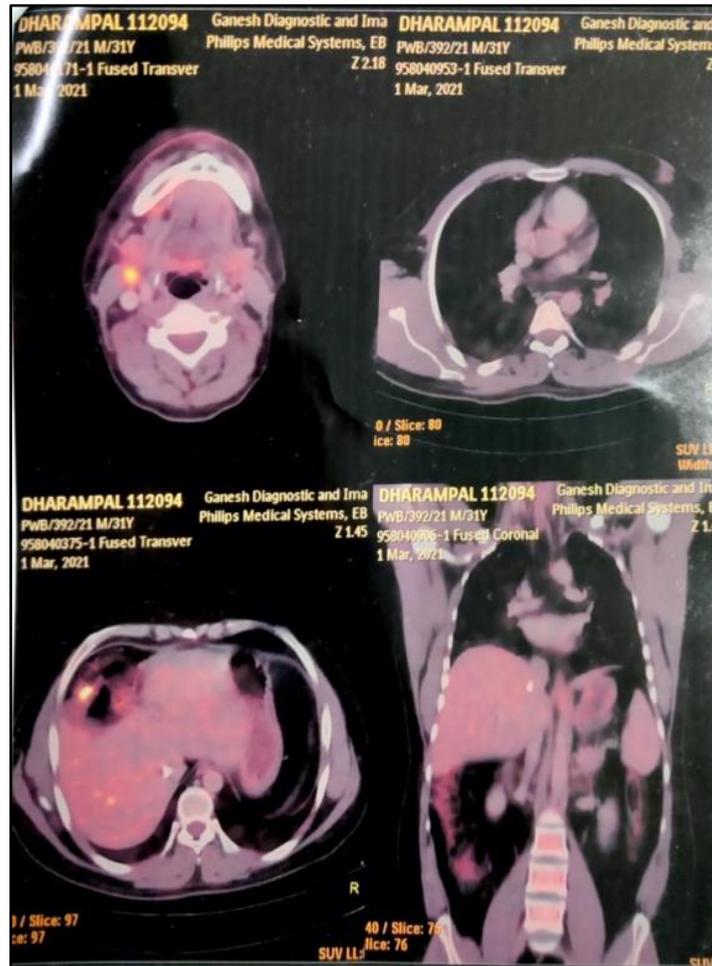
**Fig.3a.** NCCT brain in fused transverse/fused coronal plain showing no obvious intracranial radiological pathology



**Fig.3b.** NCCT head using volume axial sections through head using multi slice CT scanner showing brain with no intracranial pathology



**Fig.4a.** 18-FDG whole body PET-CT study showing ill-defined soft tissue density lesion 1.7x1.3cm at left retro areolar region, without significant radiotracer uptake



**Fig.4b.** Repeat 18-FDG whole body PET-CT study, post excision biopsy showing Non FDG avid mild thickening of the left areolar region showing post surgical/ residual changes with no metabolically active lesion elsewhere in the body. Mild FDG avid bilateral cervical lymph nodes at level II seen

#### **Declarations**

#### **Patient Consent**

*The authors certify that they have obtained all appropriate consent forms. In the form the patient(s) has/have given his/her consent for his/her images and other clinical information to be reported in the journal. The patients understand that their name and initials will not be published, and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.*

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#### **Conflicts of interest**

*There are no potential conflicts of interest.*

#### **Consent for publication**

*Authors declare that they consented for the publication of this research work.*

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