



## CASE REPORT

# Breast Fibromatosis: Case Report, Case Series, and Mini Review

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## Introduction

Fibromatosis of the breast is a rare, benign tumor, lacking metastatic potential, but can be aggressive and create issues with loco-regional control if not adequately treated. In the literature, fibromatosis of the breast is often discussed only in case reports and small case series. The consensus on treatment remains wide local excision, however, other therapies have been described. In discussion of an interesting case presentation of what we have defined as secondary breast fibromatosis, we reviewed relevant treatment modalities and discussed proper treatment for this patient. Our aims of this review of breast fibromatosis were fourfold. 1. We present an interesting case report of a secondary breast fibromatosis with an aggressive surgical resection strategy. 2. We prompt a comprehensive discussion of fibromatosis with a review of the literature. 3. We offer the delineation between primary and secondary breast fibromatosis and determine if the behavior of the two are different. 4. We present a case series obtained by conducting a retrospective review of the patients with breast fibromatosis at the University of Miami/Jackson Memorial Hospital Systems from 2003-2016 and assessed their outcomes based on medical, surgical, and radiation treatments in conjunction with prior literature reviews. Overall, this comprehensive review highlights past and present recommendations in the treatment of this locally aggressive disease.

## Case Report

A 49-year-old female with a previous diagnosis of T1N0M0 ER-/PR+/HER2- invasive ductal carcinoma of the left breast in 2009, presented in November 2016 with a new left breast mass. In 2009, at the time of her

diagnosis of her left breast cancer, she was pregnant. She underwent PET/CT and there was a large hypermetabolic axillary lymph node, however this was never biopsied. She received neoadjuvant chemotherapy. After giving birth, she underwent a left modified radical mastectomy, which showed no residual invasive carcinoma, but showed DCIS and no lymph node involvement. She received radiation therapy and completed five years of tamoxifen. She presented with a left chest wall mass in November 2016 (the mass had been present for several months, but the patient had missed several follow-up appointments). The mass continued to grow in size and was approximately 6 × 7 cm on physical exam and fixed to the chest wall. Biopsy showed fibromatosis. CT chest and MRI revealed the mass was invading the pectoralis muscle and at least two ribs. In addition to the left breast cancer, she also had von Willebrand's disease. On May 9, 2017, she underwent resection of the left chest wall mass including the pectoralis muscle and ribs 3, 4 and 5. Mesh was placed in the defect and a latissimus flap was rotated for soft tissue coverage. Final pathology showed fibromatosis, 8.0 × 7.5 × 4.0 cm. The tumor invaded muscle and abutted the ribs, but lacked invasion. There were negative margins with the closest margin being 0.5 cm from the superior aspect. The patient is currently two months out from surgery with no issues and did not require any further treatment such as chemotherapy or radiation.

## Fibromatosis

Fibromatosis of the breast (or extra-abdominal desmoid tumor) is a rare, benign tumor, however it can create issues with locoregional control if not managed appropriately. Fibromatosis arising within the breast parenchyma accounts for 0.3% of breast tumors. When



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fibromatosis of the breast is discussed in the literature, it is often presented as case reports. Large multi-center trials do not exist in the treatment and management of fibromatosis. It is agreed upon that treatment with wide local excision is sufficient. The recurrence rate is as high as 57% for fibromatosis arising from the pectoralis muscle, and 21% for those arising in the breast primarily [1,2]. Fibromatosis most often recurs in the first three years after resection [2].

### Clinical Characteristics

Patients often present with a painless firm mass and may even have associated skin dimpling. Pain and nipple discharge are uncommon. Mammographic findings mimic breast carcinoma and are often indistinguishable. Additionally, ultrasound findings may show a well-circumscribed mass that resembles a fibroadenoma, which can result in inadequate resections [1,3]. Pain is often a finding in advanced disease when the chest wall is involved.

### Pathologic Features

Grossly, fibromatosis may appear as a white or gray fibrous mass that is often well-circumscribed, but can be ill-defined as well. Microscopically, the features include spindle-cell, fibroblastic proliferation with some mitoses [1]. Although they grossly may appear well-circumscribed, a case series in 1989 by Rosen, et al., microscopically, they found that out of 22 patients, all had some finger-like stellate extensions into the surrounding fat and glandular parenchyma. This finding may contribute to the high rate of recurrence. Moreover, although these tumors developed within the breast parenchyma, the center of the tumors lacked any parenchymal elements and the surrounding breast tissue in the periphery was often a normal cellular configuration with only one case from their series containing mild hyperplasia. In conjunction with this series, our pathology results also demonstrated similar findings and seem to be their own entity within normal breast tissue [1].

### Primary and Secondary Fibromatosis

The distinction between primary and secondary breast fibromatosis has not been described in the literature. Patients may develop fibromatosis primarily within the breast parenchyma or may develop fibromatosis secondarily after receiving radiation to the chest, often due to treatment of breast cancer. When this occurs it has been observed in our own data that the tumors that arise with secondary fibromatosis appear to be well-circumscribed and less infiltrative. The secondary fibromatosis is often mistaken as a recurrence of breast cancer. This may result in less than adequate margins given that breast cancer is often not resected with 1 cm margins. Nonetheless, regarding primary or secondary fibromatosis, treatment remains the same to include wide local excision.

### Treatment and Management of Fibromatosis

As mentioned previously, the mainstay of treatment remains radical resection. Currently, at our institution, our goal is to obtain 1 cm margins circumferentially, however, this may be difficult due to the proximity or the involvement of the tumor with vital structures. Despite obtaining margins less than 1 cm in our series, patients overall have done well. Aggressive radical surgical approach is necessary due to the aggressiveness of these tumors. However, in patients with surgical resection that will result in significant deformity or mutilation or major loss of sensory or motor function, surgery should be avoided [4]. The presence of chest wall or rib involvement should not be a contraindication to surgical resection. As supported by authors Povoski, et al. in 2006, a patient with recurrent fibromatosis of the breast required a radical excision including chest wall and rib resection. This aggressive approach is acceptable to obtain loco-regional control and it also provided the patient symptomatic relief due to chest wall pain [3]. Furthermore, our patient presented previously also required resection of the chest wall and ribs, which resulted in negative margins and without significant morbidity. Of note, involvement of specialized surgical teams, such as thoracic surgery, and/or plastic surgery is imperative to successful resection.

As far as medical treatment of fibromatosis, this should only be reserved for those with the inability to resect or the presence of persistent positive margins.

Patient should be followed closely, especially in the first three years, since this is when recurrence is the highest. CT scans of the chest, mammography and ultrasound of the breasts are important imaging modalities to achieve proper surveillance.

### Case Series Review

A retrospective review was performed at our institution at the University of Miami/Jackson Memorial Hospital Systems. Data was collected from patients diagnosed with primary or secondary breast fibromatosis, and factors, such as margin status, treatment received, interval follow-up and presence of recurrent disease were evaluated. Upon review of our data collected from 2003-2016, a total of 15 women were included who were diagnosed with fibromatosis of the breast confirmed on final pathology. The age ranged from 28 to 64-years-old with a median age of 40 and mean age of 43. The average size of the lesion was 6.37 cm (range of 1.5 cm to 15 cm in greatest dimension). Thirteen of the fifteen patients underwent surgical excision including lumpectomy, mastectomy, and mastectomy with chest wall excision including ribs (two patients). Eight patients had positive margins on initial resection and required re-excision with negative margins on final pathology (Of note, some patients were treated elsewhere initially and re-excision was performed at our institution). This

was often contributed to a benign pre-operative diagnosis, such as a fibroadenoma, thus wide margins were not planned. Additionally, 8 out of 15 of the patients had previous history of radiation to the chest, majority from the presence of an infiltrating ductal carcinoma, which would support a diagnosis of secondary fibromatosis. Two patients in our series were treated non-surgically. One patient received tamoxifen and the other received chemotherapy, both with stable disease. Mean follow-up of our patients was 40.4 months and our recurrence rate was 7.7%.

## Discussion

Fibromatosis remains a rare disease, for which treatment is based largely on case reports and case series reviews. The last large case series review of breast fibromatosis was in 1989 by Rosen, et al. which included data from 22 patients. Our case series included 15 patients, with similar pathologic data and treatment strategies. Our recurrence rate was 7.7% compared to their recurrence rate of 27%. This difference is unlikely due to a differing surgical strategy, but more likely explained by the advancements in imaging techniques that provide better delineation of the extent of disease. This factor allows for better pre-operative planning and success of obtaining negative margins. In our discussion of primary versus secondary fibromatosis, treatment guidelines remain the same, which remains wide local excision. Medical treatment should be reserved for those who are not surgical candidates, whether due to medical co-morbidities, or a surgical resection is unable to be performed due to large size, inability to achieve negative margins, etc.

Radiation therapy and anti-estrogen therapies have also been described as potential treatment strategies, but data is still lacking to substantiate a true benefit over surgical resection [2]. For example, according to Sheer, et al. they presented a case of a patient given medical management of her recurrent fibromatosis. Tri-

als of NSAIDs, tamoxifen, triptorelin, sorafenib, interferon alpha 2b, and sunitinib were ineffective. The patient had a 57% response to treatment with tyrosine kinase inhibitors [3]. The exact mechanism of its effect is unknown, and although the patient had a partial response, initial treatment with surgical excision when feasible remains at the forefront.

Imperative to the treatment of fibromatosis is to perform adequate imaging, including CT scan of the chest to determine underlying chest wall or rib involvement. This will facilitate resection decision-making to obtain negative margins as well as involve multiple surgical teams if the resection will be extensive.

Although fibromatosis treatment is primarily surgical therapy, it is recommended that the management employs a multi-disciplinary approach, given that multiple medical modalities may be need to be explored. The disease itself can become complicated in advance cases, however, involving multiple specialty teams is clearly of benefit to provide the patient with the optimal care. Until further data is collected, potentially in a randomized, prospective trial, for medical treatment for breast fibromatosis, surgical resection remains the standard of therapy.

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