CASE REPORT
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Multidisciplinary management of breast fibromatosis with chest wall resection and reconstruction: a case report

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ABSTRACT

Breast fibromatosis, also known as desmoid tumor of the breast, is a rare and benign condition, yet it is locally aggressive and prone to recurrence. Due to its infiltrative nature, early diagnosis and surgical excision with wide margins are essential to minimize the risk of recurrence and preserve function and aesthetics. We report the case of a 45-year-old female patient who presented with a palpable mass in the inframammary fold of the left breast. Imaging studies and a core needle biopsy suggested fibromatosis. The diagnosis was confirmed through excisional biopsy and immunohistochemical analysis. Surgical resection of the chest wall tumor was performed in a multidisciplinary approach involving mastology, thoracic surgery, and plastic surgery teams. The resection included part of the sternum and ribs, followed by reconstruction using a latissimus dorsi flap and chest wall prosthetic materials. Histopathological and immunohistochemical examination of the surgical specimen confirmed the diagnosis of breast fibromatosis, with tumor-free margins. The patient had an excellent functional and aesthetic outcome and remains disease-free after 1 year of follow-up. This case highlights the importance of a multidisciplinary approach in managing breast fibromatosis, a rare but challenging entity. Long-term follow-up is necessary to detect potential recurrences, which are not uncommon despite complete excision.

KEYWORDS: fibromatosis; breast neoplasms; desmoid tumor; surgical procedures.

INTRODUCTION

Desmoid-type fibromatosis (also referred to as desmoid tumor) is a rare, benign, but locally aggressive soft tissue neoplasm. Histologically, it is characterized by a clonal proliferation of fibroblasts and myofibroblasts arranged in long fascicles within a collagenous stroma[1]. The lesion demonstrates an infiltrative growth pattern, minimal cytologic atypia, and a low mitotic index, which helps distinguish it from malignant spindle cell tumors. Despite being benign and lacking metastatic potential, desmoid-type fibromatosis is clinically significant due to its tendency for local invasion and high recurrence rates after surgical excision[1].

In the breast, desmoid-type fibromatosis accounts for less than 0.2% of all breast tumors, making it an uncommon clinical finding[2,3]. The majority of these tumors occur in the third decade of life, although the affected age range is broad, between 15 and 60 years[3].

Desmoid tumors may be classified as extra-abdominal or intra-abdominal, with the most common extra-abdominal locations being the shoulders, chest wall, and thighs[3]. Although this tumor does not have metastatic potential, it is locally aggressive and can infiltrate adjacent structures[2].

The etiology of breast fibromatosis remains unclear[4]. Clinically, it usually presents as a palpable, painless, firm, and mobile nodule, sometimes fixed to the pectoral muscle fascia. Retraction of the skin or nipple may also be observed[5].

In imaging examinations, including mammography, ultrasound, and magnetic resonance imaging (MRI), it resembles carcinoma, thus requiring histopathological analysis (via core biopsy, vacuum-assisted biopsy, or excisional/incisional biopsy) to establish the diagnosis[6-8].

This study aimed to describe the clinical presentation, diagnostic process, treatment, and follow-up of a patient diagnosed with breast fibromatosis at Hospital Federal da Lagoa. Data were collected retrospectively from medical records between late 2022 and 2023, with ongoing post-treatment monitoring to assess outcomes. This case exemplifies the application of evidence-based management in a public healthcare setting and provides insight

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into the therapeutic decision-making process and multidisciplinary care involved in treating this rare tumor.

CASE REPORT

This report was conducted in accordance with the CARE guidelines for case reports. Data were obtained retrospectively from the patient's medical records, imaging studies, surgical reports, and pathology results. The case was approved by the Ethics Committee (CAAE: 87557825.0.0000.0193), and the patient provided written informed consent for publication of her clinical information and images. In addition, a literature review was performed using PubMed and SciELO databases with the keywords "breast fibromatosis" and "desmoid tumor of the breast," without language restrictions, to contextualize the findings of this report.

A 45-year-old female patient (initials BSM) presented with a palpable nodule in the lower inner quadrant (LIQ) of the left breast, near the inframammary fold. She sought medical evaluation and was assessed by the mastology department at Hospital Federal da Lagoa.

On clinical examination, both breasts were symmetrical with no skin retraction. A nodular bulge was observed in the LIQ of the left breast. Palpation revealed a 4 cm, firm, slightly mobile, fibroelastic mass near the inframammary fold. The right breast and axillary and supraclavicular regions were unremarkable.

Initial imaging included a mammogram (21/10/2022), classified as BIRADS 1, and an ultrasound (17/10/2022), which identified a BIRADS 4 lesion in the left breast characterized by a 24×18 mm hypoechoic mass with irregular and spiculated margins in the LIQ. A core biopsy was performed.

Histopathological examination (14/11/2022) revealed a low-grade spindle cell neoplasm suggestive of the fibrous component of a fibroepithelial tumor, but without identifiable epithelial breast structures. Immunohistochemistry showed negative beta-catenin and positive smooth muscle actin, favoring a diagnosis of breast fibromatosis.

Chest CT (05/01/2023) demonstrated a 43×30 mm irregular soft tissue mass in the left inframammary region, extending between the subcutaneous tissue and sternum, with preserved bony cortical integrity. To confirm the diagnosis, an incisional biopsy was performed (25/01/2023), which revealed atypical spindle cell proliferation infiltrating collagenized stroma and adipose tissue, consistent with fibromatosis, with no evidence of malignancy.

Given the progressive growth and chest wall involvement, surgical resection was scheduled for 18/05/2023. The surgical plan included an extensive chest wall resection by the mastology and thoracic surgery teams, followed by reconstruction with a latissimus dorsi flap by the plastic surgery team.

The surgery was particularly challenging due to the tumor's proximity to vital thoracic structures and the need for wide resection to achieve negative margins. An anterior circular thoracotomy was performed with a 4 cm margin around the tumor, extending through skin, subcutaneous tissue, muscle, and intercostal structures. Resection involved partial removal of the lateral aspects of the 5th, 6th, and 7th ribs and the lower half of the sternum. The resected specimen measured 12.5×11 cm (Figure 1A–1F). The excised mass is shown separately (Figure 1C).

Chest wall reconstruction was performed using a multidisciplinary approach. A Marlex mesh was placed to cover the defect (Figure 1D). Three neoribs were then created using steel wires anchored in the rib and sternal remnants, followed by injection of bone cement through a drain system to provide structural support and prevent thoracic instability (Figure 1D–1F). Finally, the plastic surgery team rotated a latissimus dorsi muscle flap to achieve soft tissue coverage.

The main intraoperative risks included chest wall instability, pleural injury, respiratory compromise, chronic postoperative pain, and the possibility of incomplete margins. Despite these challenges, the surgery proceeded without complications.

The patient remained in the intensive care unit for 5 days, with stable respiratory function and good pain control. She was discharged from the ward on postoperative day 6. Outpatient

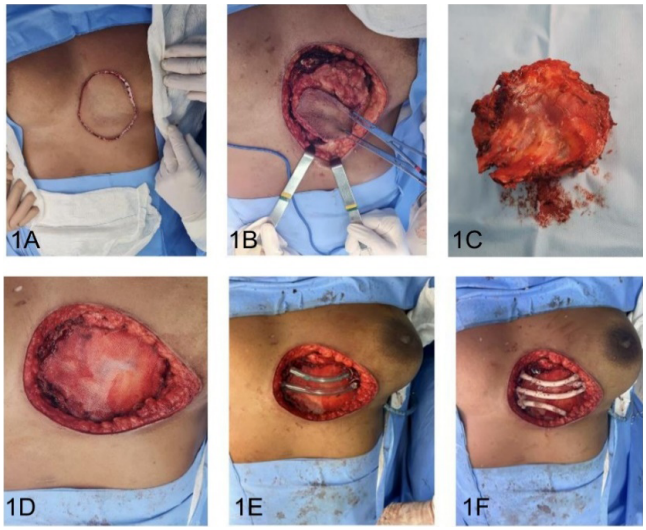


Figure 1. Intraoperative images of surgery resection and reconstruction. (A) Preoperative marking of the tumor margins on the left breast before resection. (B) Intraoperative view showing the thoracic wall tumor after skin and soft tissue incision, with exposure of the lesion. (C) Macroscopic image of the resected tumor, displaying well-defined but irregular margins and a fibrous appearance. (D) Placement of Marlex mesh for initial thoracic wall reconstruction following tumor resection. (E) Surgical placement of neocostal structures using steel wires and thoracic drainage tubing for support. (F) Formation of three neoribs using bone cement injected through fenestrated drains to complete chest wall reconstruction.

follow-up demonstrated satisfactory wound healing and good flap viability (Figure 2).

The histopathological report of the surgical specimen (18/05/2023) described a 9.5×7.0×8.0 cm segment weighing 324 g, composed of skin, subcutaneous tissue, fascia, muscle, and rib segments. Within it, a firm tan mass measuring 6.0×6.0×1.5 cm was identified, adherent to fascia and muscle, but with no infiltration of the deep margin. Microscopy revealed a uniform spindle cell proliferation with pale cytoplasm within collagenized stroma, infiltrating subcutaneous tissue, fascia, and muscle. The margins and osteocartilaginous tissue were free of neoplasia. Minimal atypia and a low mitotic index were noted, confirming desmoid-type fibromatosis. Immunohistochemistry was positive for beta-catenin, smooth muscle actin (SMA), and calretinin, and negative for S-100, CD34, and cyclin D1.

Given the complete resection and absence of malignancy, no adjuvant therapy was indicated.

At her 1-year postoperative follow-up in May 2024, the patient was asymptomatic. Mammography and ultrasound (April 2024) were classified as BIRADS 2, showing benign post-surgical changes. She continues to be monitored annually with clinical examination and imaging (mammography and ultrasound), under mastology follow-up, as part of a structured long-term surveillance plan (Figure 3).



Figure 2. Early postoperative frontal view of the patient showing favorable aesthetic and functional outcomes following oncologic resection and reconstruction with latissimus dorsi flap.



Figure 3. Postoperative image showing late scarring aspect of the surgical site, with adequate healing and no signs of recurrence.

DISCUSSION

Fibromatosis, also known as desmoid tumor, is characterized by the clonal proliferation of fibroblasts and myofibroblasts, typically arising from muscle or aponeurotic fascia[1]. Desmoid tumors can be classified as abdominal (abdominal wall), intra-abdominal, or extra-abdominal, based on the site of origin. The breast is an uncommon site for this tumor, accounting for only about 0.2% of all breast tumors[1].

It is therefore a rare, benign, and locally aggressive tumor with a propensity for recurrence—even after complete surgical excision with negative margins. Recurrence rates can reach approximately 30–35%[2,5]. Although its growth is slow, it is progressive and can become quite large[3]. Importantly, it does not metastasize[3].

The etiology of breast fibromatosis remains unclear. However, associations have been reported with trauma (incidental or surgical), familial multicentric fibromatosis, familial adenomatous polyposis, Gardner's syndrome, silicone or saline implants, and hormonal factors—especially estrogens[3,4].

Patient age ranges from adolescence up to advanced age (13–83 years) without racial or ethnic predilection; it predominantly affects middle-aged women, with only a few cases described in men[9].

Clinically, desmoid tumors typically present as painless, solid, firm nodules that may be fixed to the skin or underlying muscle fascia. Skin and/or nipple retraction may also occur[10]. Tumor size varies considerably, averaging around 2.5 cm, though it can be significantly larger. Multiple tumors are rare and generally associated with genetic conditions such as Gardner's syndrome[9].

Radiologically, on mammography, these tumors usually appear as high-density, non-calcified masses with irregular or spiculated margins[5]. Occasionally, they may present as asymmetric densities, architectural distortion, or well-defined lobulated masses[9]. Ultrasound commonly reveals a hypoechoic, ill-defined mass with thick echogenic margins and posterior attenuation; lymphadenopathy is not typically associated[11,12]. MRI is particularly useful for assessing chest wall involvement and surgical planning, as desmoid-type fibromatosis often appears poorly defined, iso- to hypointense on T1-weighted images, heterogeneously hyperintense on T2, and demonstrates slow enhancement after contrast administration[6,8].

As highlighted above, clinical examination and imaging findings frequently mimic carcinoma. Therefore, biopsy is essential for proper management[13,14]. Fine-needle aspiration cytology (FNAC) does not contribute meaningfully to diagnosis, as the cytologic material typically contains spindle cells and scattered glandular elements that are also present in other breast lesions[4]. Thus, FNAC alone cannot exclude differential diagnoses such as dermatofibroma, schwannoma, solitary fibrous tumor, or spindle cell lipoma[2].

A definitive diagnosis is established through histopathological analysis of the lesion obtained via core needle biopsy or

excisional/incisional biopsy[3]. These tumors are typically unencapsulated and irregular, characterized by spindle cell proliferation forming fibers that infiltrate adjacent glandular tissue. Most exhibit minimal or no nuclear pleomorphism[15]. Differentiation from malignant sarcoma is made by the presence of well-differentiated fibrous tissue and the absence of mitotic figures[15].

Once diagnosed, the treatment of choice is wide-margin surgical excision. In some cases, removal of adjacent structures is necessary to ensure adequate margins[2,11]. Recurrence is common—ranging from 25% to 35%—and is associated with positive margins; most recurrences occur within 3 years postoperatively[2]. A retrospective institutional study reported a 29% recurrence rate with a median follow-up of 15 months, with younger age and larger tumor size linked to higher recurrence risk[3].

Notably, reports of extensive chest wall resection with reconstruction are rare in the literature. Most published cases have been managed with local excision or mastectomy alone[16,17]. Multidisciplinary approaches involving thoracic and reconstructive surgery for en bloc resection and reconstruction remain exceptional but are critical in complex presentations like this[16].

Adjuvant radiotherapy may improve disease-free survival in cases where complete surgical resection is not feasible[12]. Hormonal therapies (e.g., tamoxifen), NSAIDs, and targeted agents such as tyrosine kinase inhibitors show promise in select cases—especially when conservative or non-surgical management is preferred[12].

Therefore, it is essential that mastologists are familiar with this disease and the most current diagnostic and therapeutic strategies, as this knowledge is crucial for achieving the best outcomes for patients.

CONCLUSIONS

Breast fibromatosis is a rare, benign, locally invasive disease with no potential for metastasis and a recurrence rate of approximately 25%. Its etiology is not well-defined and is more common in middle-aged women. Clinical examination and imaging studies resemble those of breast carcinoma, and fine-needle aspiration biopsy does not confirm the diagnosis. Histopathological examination of the biopsy material and, sometimes, immunohistochemistry are necessary.

The gold standard treatment is surgical resection with wide margins, with other options including radiation therapy, endocrine therapy, and systemic cytotoxic and non-cytotoxic therapies. After treatment, close patient follow-up is important to monitor the risk of recurrence, especially in the first 3 years. If recurrence occurs, resection, if possible, remains the first choice.

AUTHORS' CONTRIBUTION

FOGL: Conceptualization, Investigation, Methodology, Visualization, Writing – original draft. RHSM: Supervision, Validation, Writing – review & editing.

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