

## Case Report

# A Scary Onset of a Rare and Aggressive Type of Primary Breast Sarcoma: A Case Report

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## Keywords

Primary breast sarcoma · Malignant phyllodes tumor · Breast cancer

## Abstract

Primary breast sarcoma, arising from connective tissue within the breast, is extremely rare, accounting for less than 1% of all primary breast malignancies and no more than 5% of all sarcomas. The rarity of this pathology limits most studies to case reports and small retrospective studies, which has led to a lack of consensus on the clinical management. We report a clinical case of a 52-year-old woman, perimenopausal, previously healthy, with regular breast surveillance, who presented with a large (>20 cm) and rapidly expanding hypervascularized tumor of the left breast developed over 10 days, with a very thin preulcerative skin over the last 4 days. There was no systemic dissemination. The patient was submitted to total mastectomy and excision of axillary adenopathy. The tumor was diagnosed histologically as malignant phyllodes tumor associated with areas of high-grade sarcoma. Due to rapid growth and aggressive histological characteristics, adjuvant chemotherapy and radiotherapy were performed. There is a lot of evidence that tumors larger than 5 cm are associated with a poor prognosis. Despite the poor prognosis associated with this aggressive entity, the patient had no recurrence during 5 years of follow-up. We review the relevant literature about primary breast sarcomas.

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## Introduction

Breast sarcoma is an extremely rare and heterogeneous disease, which includes a group of several subtypes of cancer [1–7]. They can appear as primary forms or secondary to radiation therapy or chronic lymphedema [3, 8]. Primary breast sarcomas (PBS) are malignant tumors arising from the connective tissue within the breast and account for less than 1% of breast cancer and no more than 5% of all sarcomas [1, 9]. The published literature regarding breast sarcomas is predominantly composed of small retrospective analyses and case reports. These studies include patients with a wide variety of histologic features, treated over many decades with great variability in surgical approaches, and inconsistencies on what concerns the effectiveness of adjuvant therapies [10]. PBS is mostly diagnosed in middle-aged patients in their fifth or sixth decade of life [1, 10]. The typical patient presents with a painless well-circumscribed and mobile unilateral mass, with variable size (ranging from 1 to 40 cm) and a faster growth rate compared to epithelial breast cancer [1, 10, 11]. The gross appearance is influenced by the specific histologic features [1, 3]. Unlike epithelial breast carcinoma, the most common mode of dissemination is hematogenous, typically to the lungs, bone marrow, and liver [3]. Lymphatic spread is uncommon and so axillary lymph node involvement is not a frequent finding [1, 3, 4, 12]. The breast skin and the nipple areola complex are rarely involved in breast sarcomas [1]. Prognosis is based on tumor size, the presence of regional or distant metastases, and histological grade [3].

There is no definitive consensus regarding the treatment of breast sarcomas. Due to the rarity and heterogeneity of breast sarcomas, there are no guidelines or published randomized trials that validate any particular treatment strategy. However, there is general agreement that surgical excision with adequate margins free of cancer cells should always be the first modality of treatment [1–3, 5, 12]. Radiation therapy and chemotherapy, although controversial, may be considered in patients with high risk of recurrence due to positive margin resection, tumors larger than 5 cm, or a high-grade sarcoma, since these lesions have a tendency to recur locally and can metastasize [1–4]. Nonetheless, response rates to systemic therapies remain poor [13].

Breast sarcomas usually have a poor prognosis, with high risk of recurrence, 80% developing during the first 2 years after diagnosis [3].

## Case Presentation

We report a case of a previously healthy 52-year-old woman (perimenopausal), 1 delivery at 25 years of age, with regular breast surveillance and a normal mammography 2 years before. No previous breast disease or familial cancer was reported.

She presented an extremely voluminous tumor in the left breast with rapid growth, which appeared 10 days before admission, associated with a very thin and preulcerative skin over the last 4 days. Physical examination revealed quite a voluminous left breast, subverted by a very large tumor (>20 cm), with bosselated contours, hypervascularized, firm, with soft cystic and hemorrhagic areas (Fig. 1). No axillary adenopathy was detected during clinical examination. A few hours after admission, the breast started to ulcerate and bleed. Ulceration increased dramatically in 3 days (Fig. 2). Hemoglobin dropped to 8.8 g/dl during this occurrence. In order to control bleeding, she was submitted to a hemostatic radiotherapy flash.

Core biopsy was suggestive of malignant phyllodes tumor, with areas of primary sarcoma. Breast MRI was undertaken and revealed no muscular invasion. Staging for systemic metastases was completely negative.

A total mastectomy and homolateral axillary sentinel node biopsy were performed as it was surgically achievable and no systemic dissemination was found. Histopathology displayed a malignant phyllodes tumor, with 24 cm of longer axis, widely ulcerated, with focally tangential margins, and the malignant mesenchymal component matched a high-grade leiomyosarcoma, with marked mitotic activity and a diffusely infiltrative pattern. The sentinel lymph node was free of metastasis. The postoperative course was uneventful.

Due to very fast development and growth as well as the aggressive histological characteristics, 10 cycles of adjuvant chemotherapy with doxorubicin and dacarbazine (ADIC) + ifosfamide (IFX), followed by radiotherapy (a total dose of 50 Gy in 25 fractions over 5 weeks to the chest wall, plus a boost to the primary site of 20 Gy in 10 fractions over 2 weeks) were administered after informed consent, and it was well tolerated. The patient chose not to perform reconstruction surgery.

Clinical and ultrasound examination were performed every 6 months, and mammography every year for the surveillance of the contralateral breast. After 5 years of follow-up, no recurrence was found and the patient is alive and free of disease (Fig. 3).

## Discussion

PBS is a rare and very aggressive entity, comparable to other soft tissue sarcomas and is totally independent of epithelial breast cancers [9, 11]. Due to its rarity, there is no sufficient knowledge to support consensus regarding best management [8]. The optimal treatment for breast sarcoma involves a multidisciplinary working team, and patients should be treated according to the guidelines referring to soft tissue sarcomas [1].

Diagnosis should involve a core biopsy, once fine needle aspiration has limited accuracy. Excisional biopsy should not be performed unless previous attempts at diagnosis were unsuccessful [3, 10, 11].

The most commonly used staging system for breast sarcomas is the American Joint Committee on Cancer staging system for soft tissue sarcoma, which includes histological grade, tumor size, lymphatic involvement, and distant metastization [1].

The treatment regimen should be individualized, and a multidisciplinary approach is mandatory [8, 11]. The first-line treatment is complete surgical excision of the tumor with adequate margins, regardless of histological subtype, based on the evidence that local recurrence is more frequent in patients with surgical margins less than 1–2 cm. Negative surgical margins are more effective markers for local recurrence and overall survival than the extent of surgical resection. Lymph node involvement is rare and may occur in the carcinosarcoma subtype or when the disease is already disseminated. Thus, axillary lymph node dissection is only recommended in case of enlarged lymph nodes, suspicious lymph nodes on breast imaging studies, or in case of the carcinosarcoma subtype [1]. Routinely lymphadenectomy does not seem to improve outcomes. Sentinel lymph node biopsy was performed in this case considering the very large and aggressive tumor that could comprise areas of carcinosarcoma.

Response rates to systemic therapies remain poor and their role remains unclear. Adjuvant radiotherapy and chemotherapy for PBS is not consensual and depends mainly on the risk of tumor recurrence [8].

There is evidence of benefit with adjuvant radiotherapy for large (>5 cm), high-grade tumors or in patients with positive surgical margins in reducing rates of locoregional recurrence [11]; the impact on overall survival remains uncertain [8].

Soft tissue sarcomas are often minimally chemosensitive, with response rates ranging from 20 to 40% [1, 14]. Adjuvant chemotherapy is a reasonable option for patients who have a good functional status and a high-risk primary sarcoma or recurrent sarcoma.

Breast sarcomas have a high recurrence rate and poor prognosis. Size seems to be the most frequently reliable prognostic factor in many series, with 5 cm serving as a valuable cut point. Studies have demonstrated a 5-year disease-free survival ranging from 44 to 66% and a 5-year overall survival rate ranging from 49 to 67% [1]. Most detrimental events, such as local or distant recurrence and death, occur most frequently during the first years after diagnosis [1, 3, 11].

### Conclusion

Although rare, PBS is a very aggressive disease, with rapid growth, distinct from breast carcinoma. Dissemination is primarily hematogenous, typically to the lungs, bone marrow, and liver. Lymphatic spread is uncommon. There is no definitive consensus regarding best treatment for breast sarcoma. Surgical excision with adequate margins represents the only consensual potentially curative treatment. Adjuvant therapy, although controversial, may be considered in tumors with high risk of recurrence. The prognosis is poor and detrimental events occur most frequently during the first years of follow-up.

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### Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and the accompanying images. A copy of the written consent is available for review.

### Disclosure Statement

The authors certify that they have no conflicts of interest.

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**Fig. 1.** On admission, the patient presented an extremely voluminous tumor in the left breast (>20 cm), with bosselated contours, firm, and hypervascularized with soft cystic and hemorrhagic areas.



**Fig. 2.** A few hours after admission, the breast started to ulcerate and bleed. Ulceration increased dramatically in 3 days.



**Fig. 3.** Breast scar 5 years after diagnosis.