Case Report

# Primary myxofibrosarcoma of the male breast: A case report

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## ABSTRACT

Myxofibrosarcoma is an unusual type of malignant fibrous histiocytoma that arises in subcutaneous tissue in the limbs of elderly patients and uncommonly involves the breast. We report a rare case of myxofibrosarcoma that occurred in a male breast. A 57-year-old man was referred to our department for a right breast mass. Physical examination revealed an irregular lump without skin involvement. Ultrasonography and mammography showed an oval and hypoechoic opacity with increased internal vascularity. The patient had a core needle biopsy that showed a sarcoma of the breast. The patient underwent a simple mastectomy. The pathology report confirmed the diagnosis of high-grade myxofibrosarcoma of the breast and the patient had local radiotherapy. After a follow-up of 18 months, the patient has no evidence of local recurrence or distant metastases. Myxofibrosrcoma of the breast is extremely rare but should be diagnosed as soon as possible because of the rapid growth of the tumor and the better prognosis in the early stages.

**Key words:** Breast cancer, Myxofibrosarcoma, Treatment

Mammary sarcomas are a rare heterogeneous group of malignant neoplasms that affect more frequently the female breast [1]. The more frequent types of mammary sarcomas are angiosarcoma, fibrosarcoma, leiomyosarcoma, liposarcoma, and malignant fibrous histiocytoma (MFH) [2]. Myxofibrosarcoma is an unusual type of MFH, first described in 1977 by Angervall *et al*. [3]. It is a slow-growing, partially nodular, and painless tumor that arise s in subcutaneous tissue in the limbs of elderly patients [4] and occurs for 9% of soft-tissue tumors [5].

Herein, we report an unusual case of myxofibrosarcoma which arose in the breast of a middle-aged man.

## CASE REPORT

A 57-year-old man was referred to our department for a right breast mass evolving for 2 weeks. He was a non-smoker and had no history of chest irradiation, but a family history of breast cancer.

The patient was in good general condition. Physical examination revealed a firm, mobile, and irregular lump measuring 2 cm, in the upper inner quadrant of the right breast, without skin or nipple retraction. There were no axillary nor supraclavicular nodes. The contralateral breast was unremarkable. The rest of the physical examination was normal.

Ultrasonography of the breast showed an oval, regular, and hypoechoic opacity with increased internal vascularity, measuring 2 cm. There were no radiological axillary lymph nodes (Fig. 1a). On mammography, the tumor was ill-defined and measured 2 × 2 cm, without microcalcifications. There was no skin thickening or nipple retraction (Fig. 1b).

The patient had a core needle biopsy. Microscopic examination suggested initially a sarcoma of the breast. Thus, the patient underwent a simple mastectomy. Macroscopic examination showed, in the upper inner quadrant, a gray mass measuring 20 mm with poorly defined edges.

Microscopic examination revealed a malignant mesenchymal tumor with variable cellularity in a fibrous or myxoid matrix (Fig. 2a). Tumor cells were spindle-shaped, with eosinophilic cytoplasm and indistinct cell borders; nuclei showed a variable degree of atypia with occasional pleomorphism (Fig. 2b). In low cellular areas, some tumor cells became vacuolized, resembling lipoblasts. Mitotic index was high (23 mitosis/10 HPF (high power field)). On immunohistochemistry, tumor cells expressed CD34. Stains for pan-cytokeratin, EMA (Epithelial Membrane antigen), and p63 were negative.

The pathology report confirmed the diagnosis of high-grade myxofibrosarcoma of the breast that was completely excised.

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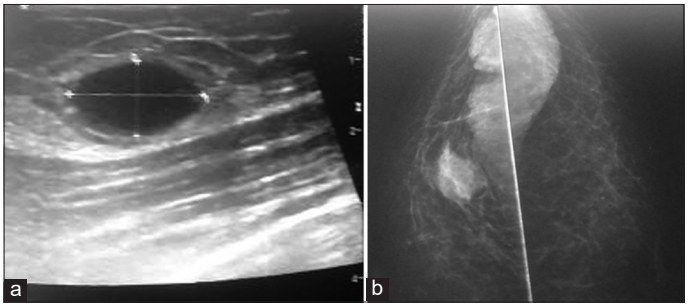


**Figure 1: (a) Ultrasound of the breast showing regular oval opacity and (b) Mediolateral oblique mammography showing oval mass with a smooth and sharp margins**

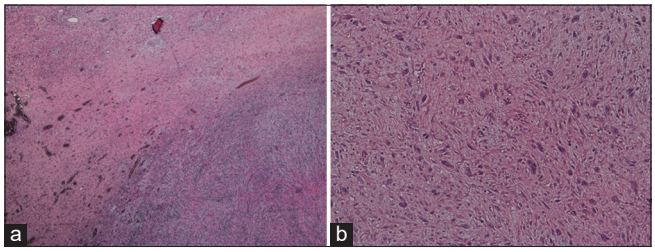


**Figure 2: (a) High-grade myxofibrosarcoma showing a nodular growth with variable cellularity in a myxoid background (HE ×40); (b) At higher power, obvious pleomorphic cells with abundant eosinophilic cytoplasm and irregular nuclei, arranged haphazardly (HE ×250)**

A thoraco-abdominopelvic CT scan was performed that did not reveal any secondary localization.

The patient had local radiotherapy of the chest wall (45 Gy) with conformal radiation technique and no significant toxicity was noticed. After a follow-up of 18 months, the patient has no evidence of local recurrence or distant metastases.

## DISCUSSION

We reported an unusual case of myxofibrosarcoma that arose in the breast of a middle-aged man. Soft-tissue sarcomas are rare and account for 1% of adult neoplasms and <1% of all breast malignancies [6]. Breast sarcomas include angiosarcoma, osteosarcoma, leiomyosarcoma, liposarcoma, fibrosarcoma, rhabdomyosarcoma, and MFH [7].

Myxofibrosarcoma is an unusual type of MFH. The incidence of breast localization was not clearly reported in the literature because of its infrequency [6]. Although myxofibrosarcomas are among the most frequent malignant mesenchymal tumors arising in the extremities of elderly patients, there are rare reported cases of breast involvement in young adults [1].

Moreover, male breast cancer is extremely rare representing <1% of all breast cancers. The most common type of male breast cancer is invasive ductal carcinoma (93.7%), followed by papillary (2.6%), mucinous (1.8%), lobular (1.5%), medullary (0.5%), and breast sarcoma (<1%) [8]. Thus, our patient had a rarely diagnosed cancer in men with a rare histologic subtype.

MFH arises more commonly after irradiation or surgery, while primary MFH of the breast arising *de novo* is extremely rare [9]. Our patient had no history of surgery or irradiation.

With ultrasonography and mammography of the breast, we could highlight the lesion and indicate a biopsy, since it was a lump in a male breast. The core needle biopsy could differentiate it from common breast neoplasms. Besides, the immunohistochemistry study contributes to the accurate diagnosis and exclusion of other differential diagnoses [1]. Smith *et al*. reported that, in 50% of superficial myxofibrosarcomas, positive staining for CD34 is noticed, as in our case [10].

However, according to several authors, magnetic resonance imaging (MRI) is considered mandatory when the diagnosis of soft-tissue tumor is suspected or confirmed, including myxofibrosarcoma. It has a better level of resolution to provide information on the tumor and its relationship with vascular nerves and bone joints [5]. As our patient had a small and mobile lump located in the breast, we performed the mastectomy without MRI.

Because of its extreme rarity, the optimal treatment is not well defined for the primary sarcoma of the breast. Therefore, recommendations are extrapolated from the soft-tissue sarcoma data. Based on the literature review, the treatment of choice for myxofibrosarcoma of the breast is local complete surgical resection with negative margins. Axillary dissection has been considered unnecessary for MFH of the breast since these tumors rarely spread through the lymphatic system [11]. For our patient, although the optimal surgical approach could be achieved with breast conservative surgery, radical treatment was performed.

The role of adjuvant chemotherapy in non-metastatic cases is unclear and hormonotherapy has no place in MFH [12]. Radiotherapy should be considered when surgical margins are inadequate or microscopically involved and when the tumor size is larger than 5 cm [8]. The use of dose escalation of adjuvant radiotherapy of at least 60GY to tumor bed was discussed in some retrospective studies, improving the disease-specific survival [13]. In our case, mastectomy without axillary dissection was performed followed by local radiotherapy.

The 5-year survival rate of myxofibrosarcoma varies from 36.4% to 90.0% and the mortality rate is 4.4% to 18.4% [5]. In their series of 19 patients with breast sarcomas, Pandey *et al*. reported that the disease-free survival at 3 years was 39% [14]. Kijima *et al*. reviewed all the reported cases of MFH in the female breast and noted that in the absence of skin involvement, lymph node metastases were rare. Skin involvement and large tumor size were associated with local recurrence as well as distant lung metastasis [15]. The prognosis is linked to delayed diagnosis, higher tumor stage, and higher tumor grade [16].

## CONCLUSION

Although breast changes in middle-aged and older men are often due to gynecomastia, male breast cancer may rarely occur. Among these breast cancers, myxofibrosarcoma is extremely rare but should be diagnosed as soon as possible because of the rapid growth of the tumor and the better prognosis in the early stages.

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