# Myxofibrosarcoma that Developed Rapidly in the Breast of an Elderly Man and Recurred Early after Surgery: A Case Report

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**Background: Myxofibrosarcoma is a rare disease occurring subcutaneously in the limbs. We report a case of a rapidly growing myxofibrosarcoma in the breast of an elderly man that recurred early after surgery.**
**Case presentation: A 73-year-old man presented with a breast mass. Physical findings showed a large tumor in the right breast, and malignancy was suspected on ultrasonography. Computed tomography (CT) revealed tumor invasion into the pectoralis major and pectoralis minor muscles. Positron emission tomography/CT showed no abnormality in other organs. Needle biopsy results excluded breast cancer but did not provide a definitive diagnosis. However, the tumor grew rapidly before further results were available, so emergency mastectomy was performed. The final pathological diagnosis was high-grade myxofibrosarcoma. Postoperative radiotherapy was started because of remnant tumor. The wound became worsened and swollen, and needle biopsy 10 days after the start of therapy indicated recurrence. Radical resection and thoracoplasty were performed. Postoperative pathological specimens showed no residual tumor. Radical radiation therapy was resumed. The patient has shown no recurrence after an year.**
**Conclusions: It is important to consult a soft tissue oncologist for tumors in the breast and perform appropriate examination and treatment if soft tissue tumors cannot be ruled out.**

**Key words: myxofibrosarcoma, gynecomastia, male breast cancer, malignant lymphoma**

## INTRODUCTION

Myxofibrosarcoma, first reported in 1977 by Angervall *et al*. [1, 2], is a slow-growing, partially nodular, painless tumor that occurs frequently in the limbs of elderly people, and accounts for 9% of soft tissue tumors [2]. There is no sex predilection, and the tumor is rarely found in the chest area or the breast.

The 5-year survival rate of myxofibrosarcoma varies from 36.4% to 90.0%, and the mortality rate is 4.4% to 18.4%. Low-grade myxofibrosarcoma is unlikely to cause distant metastasis, but the local recurrence rate is as high as 50–60%; the rates are similar for high-grade myxofibrosarcoma [2–5]. In some cases, the risk of local recurrence has been noted to increase if the bone or cartilage is involved [6–8]. Distant metastasis most commonly occurs in the lung [9–11]. Therefore, although uncommon, if myxofibrosarcoma is suspected, early referral to a specialist in soft tissue tumor is recommended.

Pathological findings of myxofibrosarcoma include short spindle-shaped or star-like polymorphic cells with an abundant mucous matrix background and characteristic elongated curvilinear vessels. Pseudolipoblasts with numerous vacuoles in the cytoplasm are found among the tumor cells, which contain acidic mucopolysaccharides not fat.

There are no specific markers for myxofibrosarcoma, but CD34 and smooth muscle actin (SM-actin) have reportedly been found on the surface of tumor cells, which may play a role in the biology of myxofibrosarcoma [3, 12]. Vimentin, which is expressed in most non-epithelial tumors, and MIB-1 (index) can be used as a reference for estimating the growth potential of the tumor. However, these markers may or may not be expressed in myxofibrosarcoma. In low-grade tumors, the tumor cells in the mucous matrix are atypical, and malignancy is classified into three or four levels based on cell density, cell type, and proliferation [2, 3, 12]. Many low-grade tumors are generated from the subcutaneous tissue, and many have good prognosis, but high-grade tumors are often found in deep soft tissues and 20?30% may develop distant metastases. If invasion to the muscle is noted, the possibility of recurrence and metastasis should be considered.

The optimal treatment for myxofibrosarcoma is surgical resection, and it is important not to leave tumor cells at the margin [13]. Radiation therapy is effective

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to some extent, when used as an auxiliary to surgery, but the effect of chemotherapy is unclear. Recently, pembrolizumab has been shown to be effective in treating myxofibrosarcoma [1].

Here, we present a case of myxofibrosarcoma in the breast of an elderly man that recurred early after simple mastectomy. The patient has achieved long-term survival after radical surgery.

## CASE PRESENTATION

A 73-year-old man visited our clinic complaining of swelling in the right breast and was admitted as a breast and endocrine surgery outpatient. He had no remarkable family history. He had undergone mitral annuloplasty and coronary artery bypass surgery when he was 67 years old, and visited the hospital cardiovascular surgery clinic as follow-up for these surgeries.

Physical examination revealed overall hypertrophy in the right breast, especially in the right CD area (outside). However, no clear tumor formation was observed on breast ultrasonography; therefore, we made a tentative diagnosis of gynecomastia. However, on the 3-month follow-up visit, the swelling in the right breast was more prominent and showed redness (Fig. 1). Breast ultrasonography revealed a low echoic mass of 83 × 25 × 49 mm centered in the upper-outer quadrant, and a mass of 68 × 12 × 41 mm in the AB (inside) area was observed in the right C area. Both masses were lobular in shape and irregular, with projections into the surrounding tissue (Fig. 2a), and a pulsatile blood flow signal (Fig. 2b). In the C area, the border between the tumor and skin was unclear, so malignancy was suspected. The first differential was malignant lymphoma because of low echogenicity and enhanced posterior echo. Breast cancer was also included in the list of differential diagnoses. Axillary and cervical lymph nodes showed reactive swelling. Blood and biochemical tests, including tumor markers, were normal.

Plain and enhanced computed tomography (CT) examinations from the next to the pelvis area were performed. Uneven increase in the concentration and distribution of fat tissue was observed in the right breast, along with unclear muscle border. A tumor structure invading the chest wall of the right breast was observed, and a malignant breast tumor or soft tissue tumor was suspected (Fig. 3). Other abnormal findings, such as suspected metastasis, were not observed.

In the positron emission tomography/CT examination, a slight accumulation of FDG was observed in the tumor in the right chest wall. Invasion into the pectoralis major and minor muscles was suspected. No abnormal accumulation suggesting metastasis was observed.

Needle biopsy of the right breast tumor was performed. Histopathological findings after hematoxylin and eosin staining excluded breast cancer completely, but immunohistological analysis was required to reach a final diagnosis. However, while waiting for the results, the tumor showed rapid growth, so emergency surgery was deemed necessary and simple mastectomy was performed.

The entire reddish area of the skin was excised during the operation. Since the tumor was suspected to have invaded the pectoralis minor and major muscles, partial muscle resection was also performed. Grossly, the tumor was completely excised without any remnants.

The resected specimen had partially unclear border (Fig. 4) and consisted of spindle-shaped tumor cells and curvilinear thin-walled blood vessels on microscopy (Fig. 5A). Myxoid stroma was partly seen (Fig. 5B). Vacuolated pseudolipoblasts were focally noted (Fig. 5B inset). The cellularity was relatively high, and cells with moderate to severe cellular atypia were also present. Multinucleate giant cells were observed (Fig. 5C, D). Tumor invasion into the muscle was observed at the deep side of the tumor, and it was suspected that tumor cells remained at the margin in this section. Tumor cells had also invaded the fat tissue, septum of the fat tissue, and dermis.

Immunohistological examination yielded negative results for smooth muscle actin (SM-actin), desmin, S-100 protein, CDK4, p16, CK(AE1/3), and MUC-4 and partially positive results for CD34 and MDM2 (Fig. 5) and (Fig. 6). MIB-1 labeling index was 30%. Based on these results, the final diagnosis was high-grade myxofibrosarcoma.

Although the tumor was grossly removed, postoperative radiotherapy was started 2 months after the operation because of unclear surgical margin on microscopy, with a dose of 50 Gy/25 fr for the chest wall. The skin became red on the 10th day after the start of radiation therapy. This was originally thought to be a complication of radiotherapy. Skin redness rapidly expanded and ultrasonography was performed, revealing a solid component under the wound. A needle biopsy confirmed the recurrence of myxofibrosarcoma. Radiotherapy was discontinued, and enhanced CT and MRI scans were performed, which also showed recurrence of myxofibrosarcoma and tumor invasion into the chest wall (Fig. 7).

We immediately consulted a soft tissue oncologist. Radical thoracoplasty was performed by a soft tissue oncologist, respiratory surgeon, and plastic surgeon (Fig. 8). Radiotherapy was performed again after the operation (total 50 Gy). The patient is alive without recurrence a year after the operation.

## DISCUSSION

We reported a case of myxofibrosarcoma in the breast of an elderly man. Although breast enlargements in older men are often due to gynecomastia, male breast cancer or malignant lymphoma may rarely occur. For example, male breast cancer accounts for 0.7% of all breast cancers [14] and 0.17% of all cancers in male [15]. Myxofibrosarcoma occurs frequently in middle-aged and elderly people. It is often found in the extremities, and its occurrence on the trunk is rare. In 2/3 of the cases, the tumor is superficial and is found in the dermis and subcutaneous skin. The tumor is rarely found in deep soft tissues such as in skeletal muscle [16]. Mammary sarcoma accounts for less than 0.1% of primary breast malignancies in both sexes [17] and is reportedly associated with acute traumatic injury [18]. Our patient had undergone heart surgery, which might have triggered the development of the myxofibrosarcoma, although the site of the wound was not close to the site of the tumor.

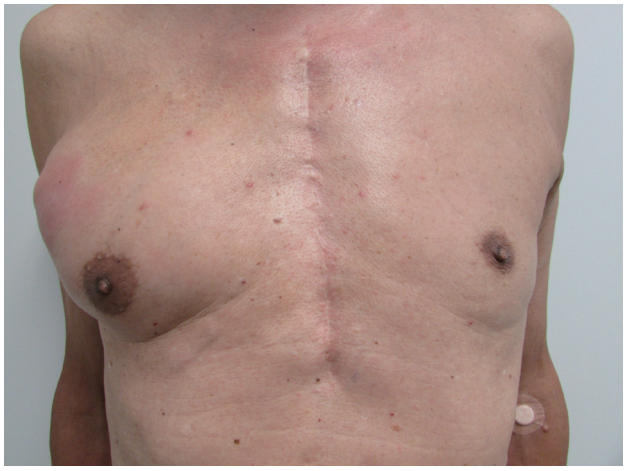

**Fig. 1** Photograph of the chest at the first visit. The right breast had a fist-sized swelling, and the skin was slightly reddened.

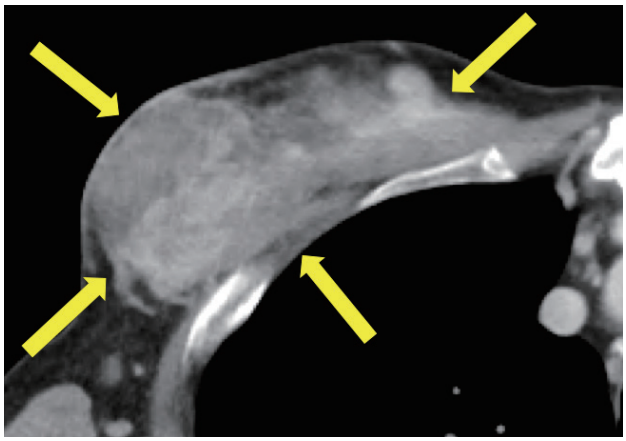

**Fig. 3** Enhanced computed tomography. A heterogeneous increase in density was seen in the right breast (yellow arrow). A tumor structure in the right breast infiltrating the chest wall was suspected. The border with the muscle was unclear.

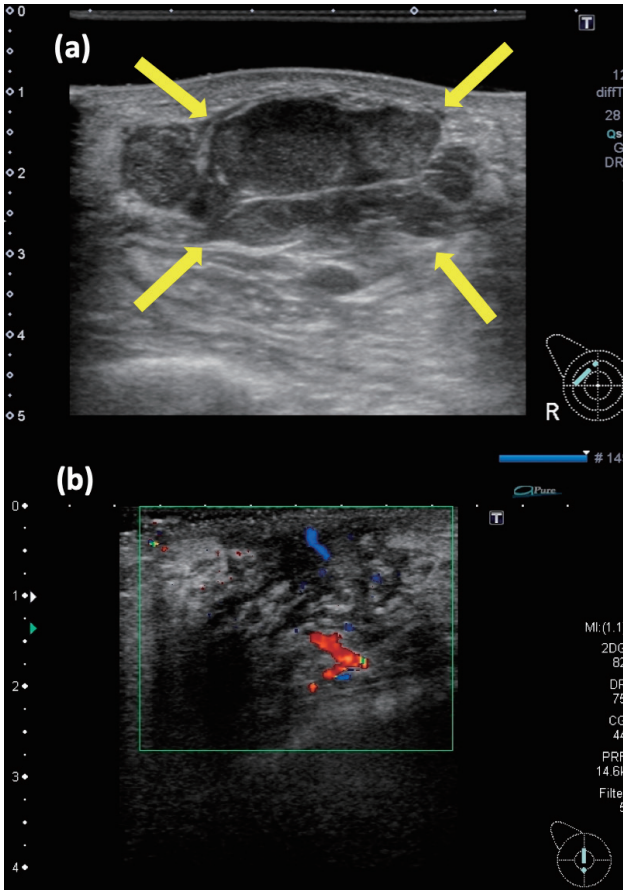


**Fig. 2** Ultrasound of the breast at the first visit. (a) A hypoechoic mass of 83 × 25 × 49 mm in the right C region (yellow arrow) and a mass 68 × 12 × 41 mm in the AB region were observed. (b) A pulsatile blood flow signals were observed inside the tumor.

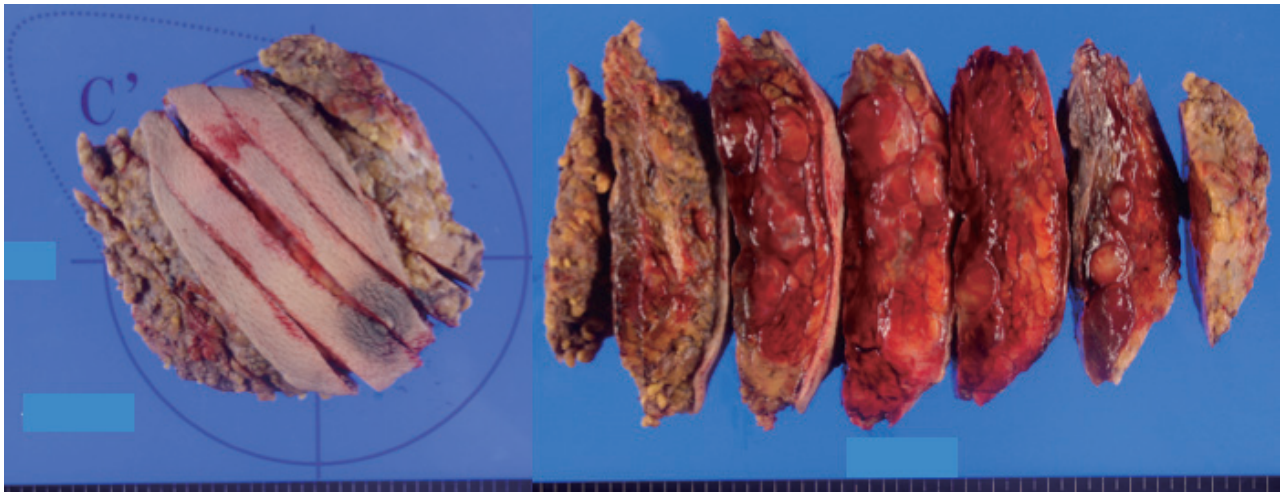


**Fig. 4** Macroscopic image of the right breast.

In this case, we considered the possibility of breast cancer and performed a needle biopsy. Emergency surgery was required because of the rapid growth of the tumor before the results of the fine needle biopsy were available. In cases of male breast cancer, MRI is generally performed after the presence of the rumor is confirmed by needle biopsy. MRI is considered an indispensable test for the diagnosis of soft tissue tumors, including myxofibrosarcoma, as it has excellent resolution and can accurately provide information on location, size, and properties of soft tissue tumors and their positional relationship with vascular nerves and

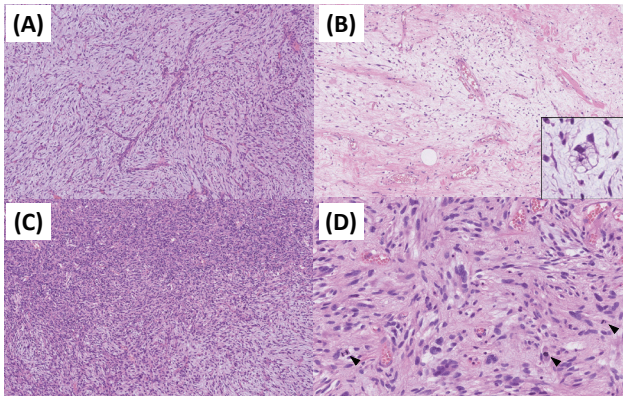


**Fig. 5** Microscopic findings of the resected tumor. (A) Spindle-shaped tumor cells and curvilinear blood vesselss, (B) Myxoid stroma of the tumor with vacuolated pseudolipoblast (inset), (C) Moderate to severe cellularity was observed in the specimen with occasional (D) multinucleate giant cells and mitoses (black arrowhead).

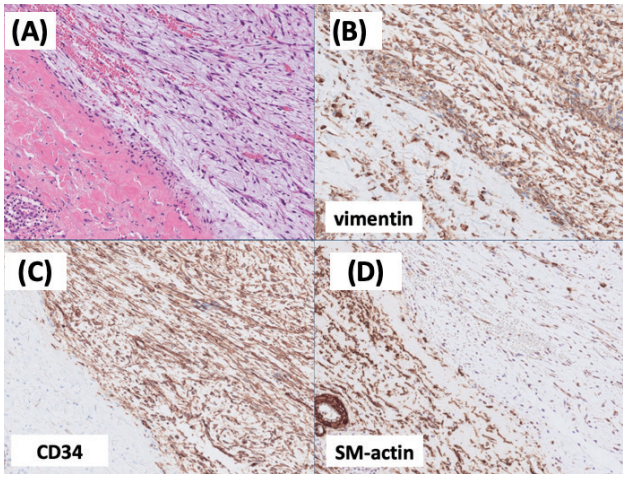


**Fig. 6** Immunohistochemical analysis of the resected tumor. (A) HE stain, (B) Vimentin, (C) CD34, and (D) SM-actin.

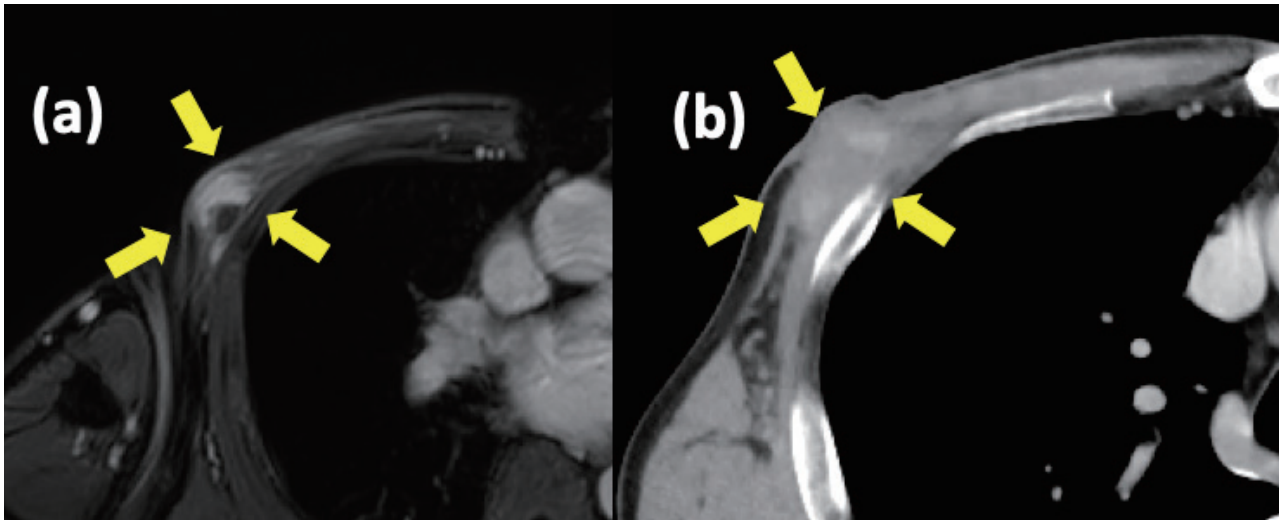


**Fig. 7** Imaging findings at the time of recurrence. Results of both magnetic resonance imaging (a) and computed tomography (b) suggested tumor recurrence and muscle invasion into the chest wall (yellow arrow).

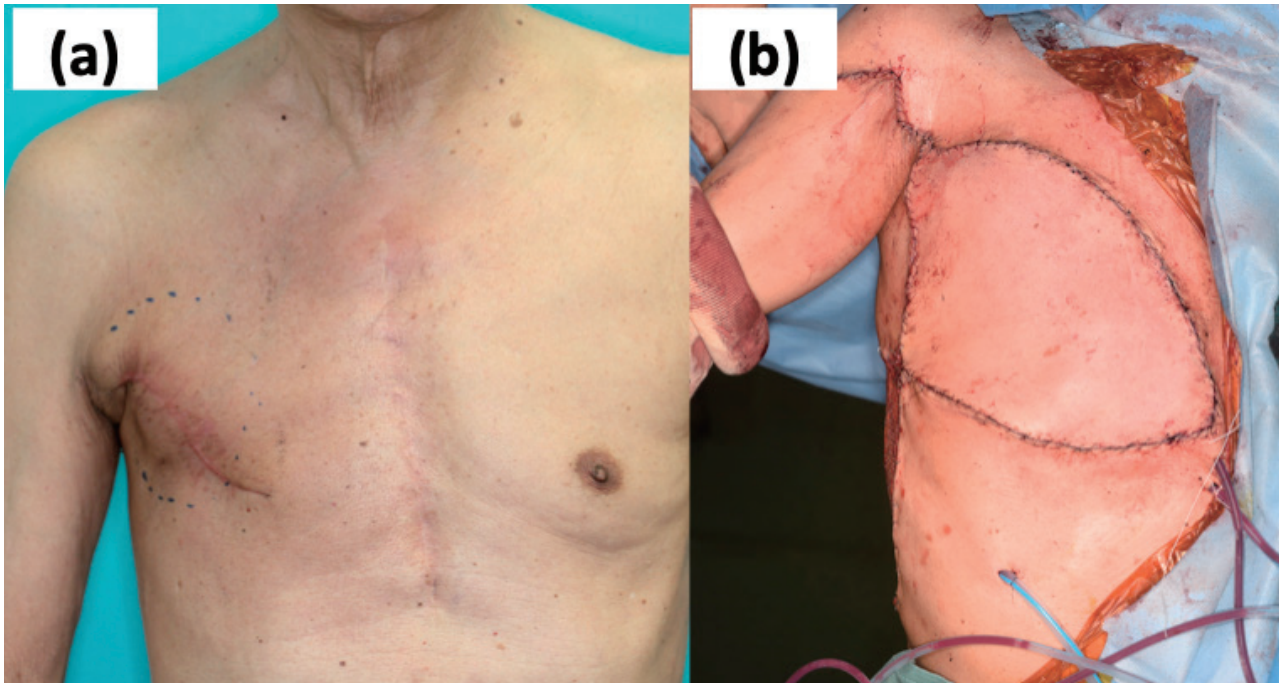


**Fig. 8** Photograph of the chest at the time of recurrence (a) and after the second operation (b).

bone joints, while ultrasound and CT do not have such level of resolution [3]. The findings for myxomas are relatively characteristic on MRI. In T1WI, myxoma-like substrate with low cell density presents with lower signal than muscle, but an area with high cell density is similar to muscle. High-grade tumors and bleeding in the tumor are defined as having a high signal. The myxoma matrix shows a high signal on T2WI/STIR, while the high cell-density region is seen as an area with a low signal. The myxoma-like substrate region has poor contrast enhancement effect, and the solid component has a positive contrast enhancement effect with a tail sign around the mass. These characteristic findings on MRI [19–21] were observed in our patient too. However, in this case, after the first operation, the tumor grew rapidly, and emergency surgery was performed. If MRI had been performed prior to diagnosis, we might have diagnosed the soft tissue tumor earlier and consulted with a soft tissue specialist immediately.

To conclude, oncologists should always have awareness regarding rare tumors such as myxofibrosarcoma and consult with the relevant specialist as soon as possible.

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