

Desmoid Fibromatosis Mimicking Breast Cancer After Nipple-Sparing Mastectomy

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ABSTRACT Desmoid fibromatosis is an uncommon fibroproliferative condition and classified as an intermediate tumor because of low metastatic potential and risk of mortality. In this article, we present a rare condition with desmoid fibromatosis mimicking breast cancer in a 52 years old patient after nipple-sparing mastectomy. A 52-year old multiparous patient referred to the outpatient clinic due to gradually growing mass on the outer quadrant of her right breast. A core needle biopsy was performed and invasive ductal carcinoma was revealed. We did therapeutic mastectomy to the right and contralateral prophylactic mastectomy to the left breast. After one-year follow-up period, the patient applied to our clinic because of a palpable mass on the inferior quadrant of her right breast and chest pain while breathing. Gross pathology results showed a 5.5x1x0.4 cm tumor. Microscopic identification of the lesion showed a nonencapsulated mass with fibrous, gray and white nodular parenchyma. The diagnosis was desmoid type fibromatosis. When desmoid tumor occurs in the chest wall, especially after conservative breast surgery operations, these tumors may resemble the recurrent breast cancer.

Keywords: Aggressive fibromatosis; breast neoplasms; subcutaneous mastectomy

Breast cancer still has a high mortality rate among women aged 40-55.^{1,2} The overall risk of breast cancer in the occidental female population is 9.6-13.1% and the risk of death is 3.4%.³ Surgery is the main approach for the treatment of breast cancer. There are several types of breast cancer surgeries and today conservative breast surgery (CBS) is taking the place of radical procedures.⁴ Mainly, CBS could be classified as wide local excision, quadrantectomy, and nipple and skin sparing mastectomy (NSSM). Although there is still a lack of long term data up to date studies for the NSSM, locoregional recurrence is evaluated less than 1% per year which is acceptable when compared to radical and modified radical mastectomy.⁵

There is still controversy about performing therapeutic NSSM and risk reducing prophylactic mastectomy to the contralateral side at the same time.

Fibrocystic disease with multiple biopsies that result in extensive scarring, lobular carcinoma in situ, atypical ductal or lobular hyperplasia, severe cellular atypia, florid papillomatosis, family history of bilateral premenopausal breast cancer, chronic mastitis with recurrent infections, proven malignancy in breast and suspicious mammogram findings at other side are indications for bilateral NSSM.⁶

Desmoid fibromatosis is an uncommon fibroproliferative condition and classified as an intermediate tumor because of low metastatic potential and risk of mortality. It could occur in any anatomic location but especially arise in the abdominal wall.⁷ They might show locally aggressive nature and invade adjacent bone and soft tissues. It accounts for only 0.2% of breast tumors and fibromatoses of the chest wall represent 10% to 20% of all fibromatoses.^{8,9}

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In this article, we present a rare condition with desmoid fibromatosis mimicking breast cancer in 52 years old patient after nipple-sparing mastectomy.

CASE REPORT

A 52-year old multiparous patient referred to the out-patient clinic due to gradually growing mass on the outer quadrant of her right breast. She had no family history or BRCA mutations. She had no previous history of breast surgery or chest wall trauma. She was only using an oral antidepressant agent as a medication. Breast USG and mammography findings showed that there were 11x7.5 mm, 7x6 mm and 3.5x3 mm multicentric solid masses at 9 o'clock which were resulted in BIRADS-6. Then, the patient admitted to the general surgery department of our hospital. A core needle biopsy was performed and invasive ductal carcinoma was revealed. Concurrent sentinel lymph node biopsy was negative. Breast surgeons consulted the patient to our clinic for performing nipple-sparing mastectomy and immediate reconstruction. After we had taken informed consent approval for surgery from the patient, we did therapeutic mastectomy to the right and contralateral prophylactic mastectomy to the left breast. We created a subpectoral pocket to insert the silicone prosthesis and subsequently placed 375 cc round, textured high profile Mentor® silicone implants. The implant surface was covered with inferiorly based lipodermal flap and pectoral muscle. The patient only received hormonotherapy (tamoxifen) as an adjuvant treatment after the operation.

The drain was left for one week and the patient was discharged without any complication. After one year follow-up period, the patient applied to our clinic because of a palpable mass on the inferior quadrant of her right breast and chest pain while breathing. Preliminary diagnosis according to imaging modalities was desmoid type fibromatosis (Figure 1). Fine needle aspiration was done under ultrasound guidance and the pathology result was spindle cell mesenchymal tumor. Immunostaining was positive for actin, CD34, beta-catenin, keratin and negative for desmin caldesmon, calponin, S100, and STAT-6. Under these circumstances, surgical excision of the mass was performed, including the partial intercostal muscles and

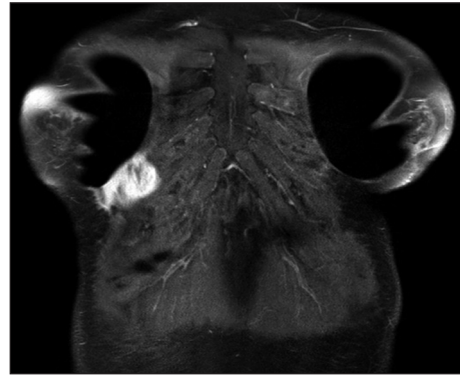


FIGURE 1: Preoperative view of MR findings of right breast desmoid tumor under silicon prosthesis.

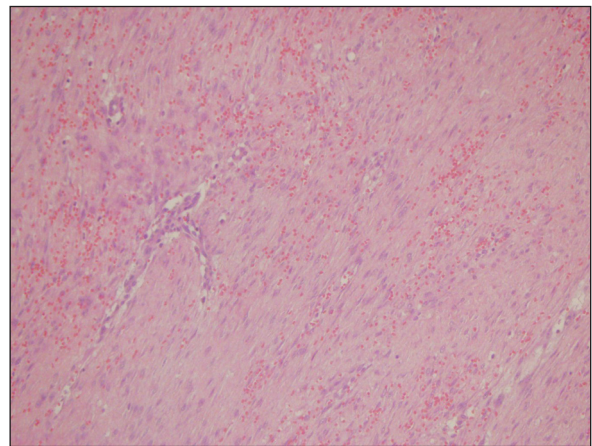


FIGURE 2: Pathological view of the desmoid tumor.

chest wall without resection of the ribs or the parietal pleura. The mass had solid, irregular borders and was adherent to the adjacent tissues. Then, silicon prosthesis was reinserted under the sub-pectoral plane.

Gross pathology results showed a 5.5x1x0.4 cm tumor and the identification of the lesion showed a nonencapsulated mass with fibrous, gray and white nodular parenchyma. The diagnosis was desmoid type fibromatosis with positive medial and inferior margin. Immunostaining was positive for nuclear beta-catenin, actin, CD34 and negative for STAT6, similar to previous findings (Figure 2).

DISCUSSION

Desmoid tumors (DTs) are rare, accounting for 0.03% of all neoplasms and <3% of all soft tissue tumors.^{10,11} It occurs in any anatomic location but es-

pecially arises in the abdominal region. Intra-abdominal desmoid tumors could be related to familial adenomatous polyposis (FAP), Gardner Syndrome and mutations in CTNNB1.⁷ DT has a wide range of age distribution and female preponderance. However, some studies report that it also occurs in men.^{12,13} In our case report, the patient was female and 52 years old which was compatible with the literature. DT was identified in the chest wall without any skin involvement or retraction and abdominal imaging showed no sign of colorectal polyps.

Fibromatoses of the chest wall represent 10% to 20% of all fibromatoses.⁹ There are breast tissue related chest wall desmoid tumors in the literature but after nipple-sparing mastectomy operation, it could resemble the recurrent breast cancer in many aspects.¹¹ As the appearance of the mass is similar to the soft tissue density in radiological modalities, it could be unnoticed easily. Also, progressive enlargement and locally aggressive behavior may lead the clinician to misdiagnosis. Expression of STAT6 is highly sensitive and specific for solitary fibrous tumor therefore, beta-catenin staining assisted us to distinguish these two entities.¹⁴

Magnetic resonance imaging (MRI) is the first modality for evaluating the soft tissue tumors and superior contrast compared to computed tomography (CT). On CT scan, tissue density could be similar and difficult to distinguish from each other. DTs are composed of fibroblasts and collagen fibrils. When fibroblasts are the dominant cells in the tumor, their water content are higher than the collagen and tumors are bright T2 weighted images.¹⁵ As in our case, even if the radiological modalities show strong evidences for the different type of soft tissue tumor, pathological verification is crucial for ruling out the recurrent breast cancer.

According to the literature, there are fewer than 250 reports and most of them are case reports and series.¹⁶ In the largest published study 44% of the patients had a recent breast surgery.¹¹ Additionally, implant-related breast desmoid tumors are even rarer and most of them are associated with silicone implants.¹⁷ As in our case report, it is obvious that desmoid tumors have been associated with an-

tecedent trauma, particularly surgical intervention but it is not clear that whether one traumatic event is enough.¹⁸ Because, either for an aesthetic breast augmentation or reconstruction with silicone implant there may be repetitive micro-traumas due to the prosthesis. As a result of, normal chromosomal analysis of the patient, the reason lying behind this sporadic DT could be the mastectomy operation. On the other hand, the patient was using tamoxifen for one year as adjuvant therapy for breast cancer which is also an initial treatment option for DT.¹⁹

According to the World Health Organization (WHO) classification of tumors of soft tissues, DT is a locally aggressive fibroblastic tumor. Management remains controversial because of the limited data due to its low incidence. Younger age, chest wall location and >10 cm diameter are the strongest predictors of the recurrence thus, surgical resection with negative margins is feasible.²⁰ However, an aggressive approach could result in an increased risk of morbidity and consequent complications.²¹ We couldn't excise the mass with a negative margin therefore, instead of coping with lung-related complications; we opted to treat the residual tumor with radiotherapy. Also, systemic chemotherapy based treatments might be considered according to toxicity and response.

CONCLUSION

Desmoid tumors are locally aggressive intermediate type tumors. When it occurs in the chest wall, especially after conservative breast surgery operations, these tumors may resemble the recurrent breast cancer. Therefore, the clinician must be aware of this kind of tumors and inform the patient in detail.

Informed Consent

The patient signed informed consent form for every step of the procedure and the study was conducted in accordance with the 1964 Helsinki declaration and Institutional Ethical Committee.

Source of Finance

During this study, no financial or spiritual support was received neither from any pharmaceutical company that has a direct connection with the research subject, nor from a company that provides or produces medical instruments and materials which may negatively affect the evaluation process of this study.

Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Haluk Vayvada, Cenk Demirdöver, Alper Geyik;

Design: Haluk Vayvada, Cenk Demirdöver, Alper Geyik; **Con-**

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