Case Report
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Recurrent Primary Fibromatosis in the Breast: A Case Report[1]
유방에 재발한 원발성 섬유종증: 증례 보고[1]

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Fibromatosis is a rare benign tumor that presents predominantly as a unilateral, painless, palpable, and firm to hard mass which may be accompanied by skin dimpling and nipple retraction. It is characterized as an infiltrating fibroblastic and myofibroblastic proliferation that can behave in a locally aggressive fashion if incompletely excised. However, it is not known to metastasize. We report a case of a 39-year-old female with recurrent fibromatosis in the breast. Initially, the mass was detected by a screening mammography. It appeared as an oval mass and recurred twice after surgical excision over a 36-month period with a more suspicious appearance.

Index terms
Breast
Fibromatosis
Ultrasound
Breast Tumor

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INTRODUCTION

Fibromatosis tumor of the breast is an extremely rare entity. It is a benign stromal tumor of the breast that constitutes less than 0.2% of all breast tumors (1). Although histologically benign, this tumor is locally aggressive and may recur in up to 29% of cases (2). Fibromatosis can occur at any age, but most commonly appears in the fourth decade of life.

Fibromatosis is subdivided into two major groups; superficial (fasical) and deep (musculoaponeurotic). Deep fibromatoses are genetically distinct from superficial fibromatosis. Features of deep fibromatosis are rapidly growing, usually large in size and involve to deeper structures. The etiology of fibromatosis is unknown but an association with Gardner's syndrome or surgical trauma has been reported. Failure to accurately diagnose fibromatosis leads to local recurrence after incomplete excision.

Breast fibromatosis arises in underlying fibroaponeurotic fascia of the pectoralis rather than in the breast parenchyma. We report a case of a recurrent primary tumor confined to breast. The purpose of this report is to describe a case of breast fibromatosis which recurred after surgical excision and appeared as malignancy.

CASE REPORT

A 39-year-old female who had a 1.5-cm mass in the left breast, which was detected by a screening mammography was referred to our clinic for an ultrasound. Ultrasound shows an ill-defined, oval inhomogeneously mixed echoic mass, measuring 1.5 cm at the 12 o'clock-position (Fig. 1A). The mass was categorized as BI-RADS 3 (probably benign). She underwent a surgical excision as per her wishes. A hostologic analysis revealed an ill-defined lesion, which presented as a slightly myxoid stromal change between preexisting mammary lobules and proliferation of spindle-shaped immature fibroblasts forming vague fascicles in slightly myxoid stroma (Fig. 1B). She was diagnosed with a fibromatosis.

In her routine follow-up after 1 year, the patient reported

feeling a small-sized (0.6 cm), palpable lump at the excision site of her left breast and that lump was a non-tender mass. Ultrasound showed a well-circumscribed, round hypoechoic mass (Fig. 2A). It was considered as a BI-RADS 3 (probably benign) and a short term (six-month) follow-up was recommended. On serial follow-up ultrasounds, the mass morphed into a lobular shape and had a taller than wider mass (Fig. 2B, C). Finally, the mass was categorized as BI-RADS 4a (suspicious for malignancy) and was recommended for a biopsy. A 14-gauge core needle biopsy was performed with ultrasound guidance. The result was adenosis and stromal fibrosis. Despite this result, imaging findings led to excision for a definite diagnosis. The histology result was fibromatosis, which is a solid mass with a stellate appearance and peripheral entrapped breast lobules, and is composed of the fascicular growth of more mature fibroblasts in collagenous stroma (Fig. 2D). No evidence of carcinoma was seen. Immunohistochemical staining for smooth muscle actin and β-catenin were positive and S-100 protein was negative and the CD 34 was focally positive. Histologic examination of the lesion revealed a recurrent fibromatosis which formed mass.

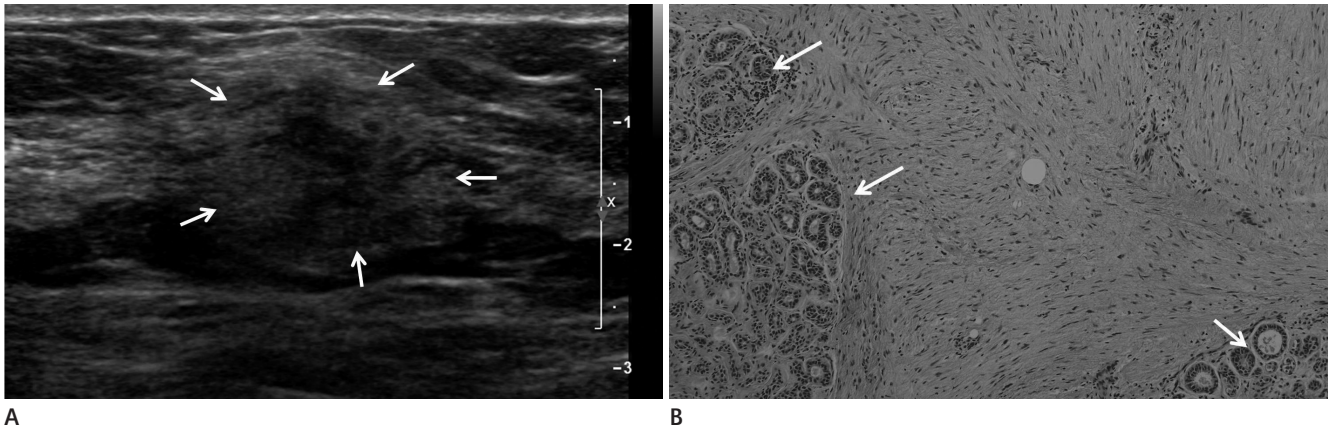
After 1 year, she complained of the second recurrent lump in the same site. Ultrasound showed a 1.5 cm, ill-defined, irregular mass with peripheral halo which was typical findings of malignant mimicking fibromatosis (Fig. 3). Recurrences were treated by wide reexcision and 1-cm negative margins were acquired. The histology was fibromatosis which extended into surrounding fat and glandular parenchyma. She is currently two years out from her last wide excision and she remains disease free.

DISCUSSION

Mammary fibromatosis is a rare, benign, nonmetastasizing stromal tumor. Fibromatosis presents as a palpable mass that is clinically suspicious for malignancy. Dimpling or retraction of the skin may be present, and the mass may adhere to the chest wall. The etiology of fibromatosis is poorly defined. Fibromatosis may occur sporadically but also may occur after trauma or a previous surgical procedure such as breast reduction (3) or breast augmentation (4). Fibromatosis was reported in a patient with familial adenomatous polyposis syndrome, or Gardner syndrome (2). Nonetheless, few cases have been reported (1). Unlike abdominal desmoid tumors, mammary fibromatosis does not appear to be associated with pregnancy (2). Most cases are reported in women, though there have been rare cases in men.

On sonography, fibromatosis typically appears as a solid, spiculated or microlobulated, irregular hypoechoic mass with straightening and tethering of Cooper ligaments, which is very difficult to differentiate from a malignant lesion. Calcifications are rarely associated. Involvement of the pectoralis muscle or intercostal muscles may be identified, indicating the locally aggressive nature of fibromatosis.

Primary fibromatosis which has been reported in the English language literature are available in 11 cases (Table 1) (5-15). Primary fibromatosis has more often occurred to male (4 of 11) than thought. When those are divided to less than 1.5 cm, two of four cases were defined as a lobular hypoechoic mass on ultrasound and two cases were taller than wide, and the irregular



A B

Fig. 1. A 39-year-old woman with a non-palpable mass in the left breast.
A. Ultrasound shows an ill-defined, oval inhomogeneously mixed echoic mass (arrows), measuring 1.5 cm at the 12 o'clock-position.
B. Pathology reveals an ill-defined, slightly myxoid stromal change which is present between preexisting mammary lobules (arrows) without mass formation (Hematoxylin-Eosin stain, × 12.5).

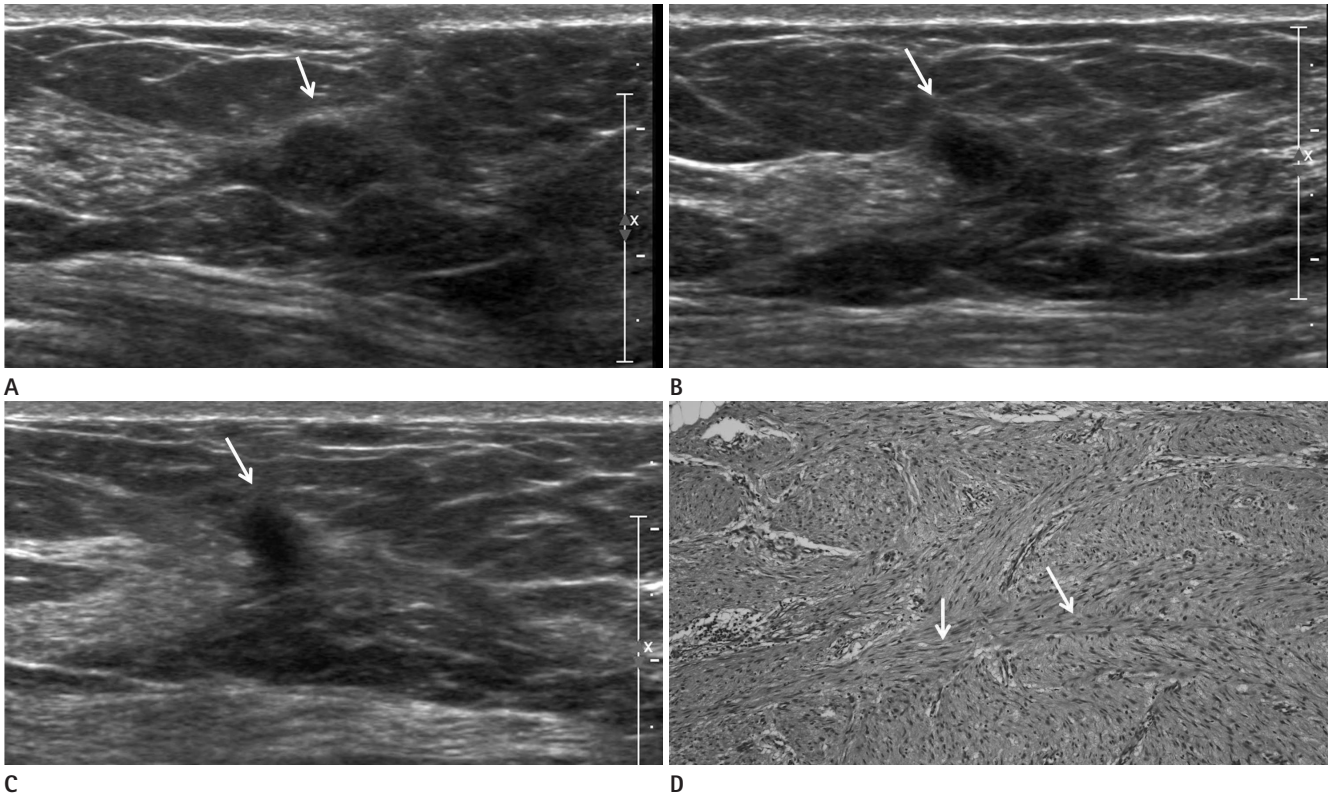


Fig. 2. A 39-year-old patient who developed a recurrent mass.
A. At 12 months after operation, ultrasound shows a 0.6 cm revealed a well-circumscribed, round homogeneously hypoechoic mass (arrow) at the scar.
B. At 16 months after surgery, the mass is changed into a lobular mass (arrow) on 6 months follow up.
C. At 24 months after the operation, the mass (arrow) had a taller than wide appearance and an angular margin.
D. Pathology reveals an ill-defined solid mass with a stellate appearance and peripheral entrapped breast lobules and is composed of fascicular growth (arrows) of more mature fibroblasts in the collagenous stroma (Hematoxylin-Eosin stain, × 200).

masses with spiculated margin. All fibromatosis was 1.5 cm had typical features of infiltrating fibromatosis. Smaller masses tended to be a less spiculated irregular shape and weren't adherent to the pectoralis muscle.

Our cases are small (less than 1.5 cm) lesions. The first lesion is primary fibromatosis, which presented as ill-defined, irregular lesion. The second and third lesions are recurrent fibromatosis, which progressed to a mass with malignant features on imaging. In addition, imaging findings are correlated to pathologic findings. Their differences entail whether the lesions form the mass or not. We don't know how toclassify changes in morphology, but we can assume supposed to be by the effect of operation and progress in growth as time goes by. If previously reported cases were detected earlier, those might be shown as a benign mass like our case.

MRI is helpful for determining tumor extent, and in particular, chest wall invasion. The masses are isointense to muscle on

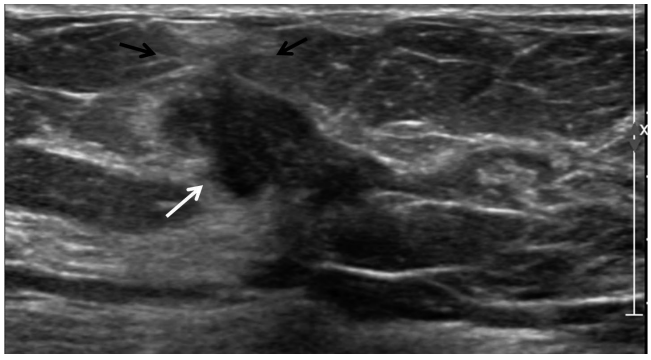


Fig. 3. A recurrent mass at 36 months after the first operation. The mass shows a 1.5-cm, ill-defined, irregular mass (white arrow) with a peripheral halo (black arrows) of fibromatosis as a benign mass mimicking malignancy.

T1-weighted images (WI) and of variable high signal intensity on T2-WI. After chemotherapy or radiation therapy, or both, areas of decreased T1 and T2 signal intensity developped, suggesting decreased cellularity and increased dense fibrosis. The enhancement pattern is generally one of a benign progressive

Table 1. Eleven Cases of Primary Breast Fibromatosis

Case No	Age/Sex	Imaging	Adherent to Pectoralis	Site	Size (cm)
1	53/F	Ill defined, lobular	Not	Left	Small (?)
2	26/F	Taller than wide, irregular mass	Not	Left	1 cm
3	40/M	Ill defined, lobular	Not	Left	1 cm
4	45/M	Spiculated, irregular, taller than wide	?	Left	1.3 cm
5	25/F	Spiculated, irregular	Yes	Left	2 cm
6	46/F	Ill defined, round	Not	Right	2.3 cm
7	72/M	Irregular mass	Yes	Right	2.5 cm
8	47/M	Irregular, posterior shadowing	Yes	Left	3.5 cm
9	72/F	Ill defined, irregular	Yes	Left	4 cm
10	20/F	Spiculated, irregular	Yes	Right	10 cm
				Left	4 cm
11	28/F	Irregular	Yes	Left	?

enhancement, as opposed to the typical washout kinetics of breast carcinoma (4). However, an enhancement pattern can be variable.

Microscopically, an infiltrative stromal process occurs, and is composed of fibroblasts or myofibroblasts, or both, with little to no nuclear pleomorphism and variable amounts of collagen. Mitoses occur rarely to none (2). Lymphoid aggregates often may be seen at the periphery of the lesion. Mutations in the adenomatous polyposis coli and β-catenin pathway are important in the pathogenesis of mammary fibromatosis and are similar to desmoid tumors in the abdomen that are called deep fibromatosis (16). Most cases have shown no estrogen receptor or progesterone receptor positivity, although low estrogen receptor or progesterone receptor reactivity has been shown in patients with familial adenomatous polyposis (3).

The differential diagnosis in pathology includes metaplastic carcinoma of spindle cell type, low-grade fibrosarcoma, and nodular fasciitis (2). Metaplastic carcinoma has an invasive or in situ malignant ductal component. Fibrosarcomas show marked cytologic pleomorphism and increased mitotic activity compared with mammary fibromatosis. Nodular fasciitis contains abundant inflammatory cells dispersed in the loose, myxoid stroma, and multinucleated cells can be present.

Local recurrence of fibromatosis is common. Although recurrence may develop in patients with negative margins, recurrence rates are high in patients with positive margins. Over 60% of margin positive cases recurred within 1 year from the initial surgery (2). Since fibromatosis of the breast is such a rarely reported event in the literature, no clear-cut guidelines have been established to define what exact standards should be applied to the definition of a complete wide local excision. As with the surgical management of breast cancer or phyllodes tumor, a wide excision with a 3-cm surgical margin from the tumor is therefore recommended.

Margins may be clinically difficult to assess during the surgical procedure because of the infiltrative nature of fibromatosis. Frozen sections may be helpful to determine clear margins. However, if the patient had a previous biopsy, differentiating between mammary fibromatosis and the prior biopsy site may be difficult.

Because of chest wall infiltration, a resection of the chest wall, which includes the ribcage, and because of large or extensive recurrences, mastectomies may be necessary. Thus, failure to recognize fibromatosis or misdiagnosis as scar tissue by a previous breast excision may potentially lead to more radical surgery.

In conclusion, we report a recurrent case of breast fibromatosis showing the evolution from an ill-defined irregular lesion like a benign mass, to a taller than wide mass which mimics a malignant mass on an imaging study.

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유방에 재발한 원발성 섬유종증: 증례 보고[1]

임수진[1] · 강영혜[1] · 김루시아[2] · 조영업[3] · 이주원[1] · 김윤정[1]

섬유종증은 섬유모세포와 근섬유모세포의 침윤적 성장을 하며 제거가 불완전할 경우 국소적으로 재발을 자주 하지만 전이는 드물다. 주로 일측에 통증을 동반하지 않는, 단단한 종괴로 만져지면 경우에 따라 피부와 유두의 함몰이 동반될 수 있다. 저자들은 39세 여성에서 발생한 원발성 섬유종증의 재발성 병변을 보고하고자 한다. 이 종괴는 처음에 선별유방촬영에서 발견되었으며 모양은 난원형이었다. 그러나 수술 후 36개월 동안 두 차례 재발하였고 악성소견을 갖는 종괴로 변하였다.

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