

Research article

BREAST FIBROMATOSIS: An uncommon benign breast disease mimicking carcinoma

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Abstract

Breast fibromatosis is exceedingly rare and often misdiagnosed, comprising only 0.2% of breast tumors. Extra-abdominal fibromatosis is an uncommon benign breast lesion resembling an infiltrative carcinoma in its clinical and radiological presentation. Positive diagnosis is provided by histology. The treatment is based on surgical excision with wide margins to avoid recurrence of this locally aggressive tumor. The aim of this study was to report difficulties encountered in the diagnosis and therapeutic particularities. We describe the clinical, radiological and pathological features of a series of five cases of primary fibromatosis of the breast. All patients were women of 38-59 years of age. None of the patients was affected by any genetic disorder characterized by fibromatoses involving multiple sites, including breast. One patient had eight years ago a contralateral breast carcinoma treated by mastectomy axillary lymphadenectomy, chemotherapy and radiotherapy. Other patient was operated four years ago in the same breast and at the same quadrant for adenofibroma. The fibromatosis of the breast, in all cases, presented as a monolateral solid nodule, clinically suspicious for malignancy in 4 patients with no cutaneous and/or nipple retraction. Both echographic and mammographic examinations, revealed in 4 patients, solid masses with irregular margins but without calcifications, mimicking scirrhous carcinoma. All cases were surgically treated by lumpectomy. Histological examination revealed finger-like infiltrating margins entrapping adjacent breast parenchyma and adipose tissue in all cases. The hallmark of the lesion was the

presence of bland-looking spindle cells, with a low mitotic index, organized in long sweeping and intersecting fascicles. Estrogen and progesterone receptor testing were not performed on the specimen in all case.

After a follow-up period ranging from 12 months to 16 years all patients are well and disease-free. In our series breast fibromatoses presented as a palpable masses suspicious for carcinoma clinically and radiologically. Therapy remains primarily surgical, and core biopsy aided in operative planning.

KEY WORDS: breast fibromatosis, desmoid tumors, extraabdominal desmoid tumors, Breast imaging, Microbiopsy, Surgery, spindle cell tumors

INTRODUCTION

Fibromatosis, which is also known as desmoid tumors (DTs) are extremely rare, with an incidence of less than 5 individuals per million of population per year (1, 2) comprising 3% of soft tissue tumors. These tumors usually affect the abdominal wall of parous women within 25 and 35 years of age. The mammary gland is one of the rarest sites in which fibromatosis has been described, representing a 4% of extra-abdominal DTs and only 0.2% of primary breast lesions (2, 3, 4). Age for diagnosis of breast fibromatosis is between 38 to 59 years.

METHODS

We report 5 cases of primary fibromatoses of the breast treated between 1997 to 2013 in the Tunisian institute of carcinology, Salah Azaiez institute. For all cases, the original slides were reviewed by pathologist and diagnoses of desmoids tumors were confirmed; margin status was also assessed.

In patient with a prior history of breast cancer, available slides from the breast carcinoma were reviewed and confirmed. Patients clinical records were reviewed, clinical data, age, history of prior operation on the breast, margin, and tumor size were abstracted. Radiological findings were taken as the written impression documented in the clinical record and images were not reinterpreted. Surgical treatment was a wide-local excision.

RESULTS

All patients were female. One was non menopausal, two was perimenopausal and two was postmenopausal. The mean age was 49 years. No patient had a diagnosis of familial adenomatous polyposis and a prior history of desmoid tumors. A 44-year-old woman had undergone an excisional biopsy for fibroadenoma at the same site 4 years previously. Another 58-year-old woman was treated 8 years previsouly for a controlateral breast carcinoma and she had mastectomy with axillary lymphadenectomy followed by chemotherapy and radiotherapy and hormonotherapy by Tamoxifen. This patient presented with two masses in the right breast. All patients presented with a palpable mass. Four patients were presented with physical findings suspicious for carcinoma. The lesion involved the right breast in 4 cases. The lesions ranged from 10 to 40 mm (tab 1).

Mammography was performed in all patients and visualized tumors as irregular uncalcified hyperdense mass suspicious for carcinoma in four patients (fig1). Breast ultrasound (fig2) was found to have a solid, spiculated irregular hypoechoic masses with posterior shadowing in four patients. Although, one patient had unusual benignlike sonographic characterized by circumscribed borders and posterior acoustic enhancement (fig3-4). No patient underwent preoperative magnetic resonance imaging (MRI) of the breast. One patient had preoperative a core biopsy that was interpreted as benign breast tissue with fibromatosis. All five patients had wide local excision with wide margins (fig5). The histopathological exam showed that all the lesions were composed of a spindle cell (fig6) proliferation forming sweeping or interlacing fascicles. The cellularity varied from mild to moderate. A variable amount of collagen was present and was predominantly located in the center of the lesions. Estrogen and progesterone receptor testing were not performed on the specimen. Neither hormone therapy nor radiotherapy was done for our patients. No recurrence was observed. After a follow-up period ranging from 12 months to 16 years all patients are well and disease-free.

DISCUSSION

Fibromatosis, also termed a desmoid tumor, is a group of soft tissue tumors that may arise from musculoaponeurotic structures, in many anatomic locations of the body (5, 6). Although it is relatively benign, it is locally aggressive and has a tendency to recur without metastatic potential (7). Breast fibromatosis is exceedingly rare and often misdiagnosed, comprising only 0.2% of breast tumors. As an extra-abdominal desmoid tumor, Fibromatosis of the breast is extremely rare, comprising 3% of soft tissue tumors and accounting for less than 0.03% of all neoplasms (1, 2, 6). The first case was reported in 1923 (8). Breast fibromatosis originates from the pectoralis major muscle or rarely from the breast parenchyma, its etiology remains unclear; however, there have been a few reported cases associated with genetic disorders such as Gardner syndrome, familial adenomatous polyposis, silicone breast implants, and trauma (9, 10).

Most cases are reported in women, with six cases reported in men (3,8). These lesions affect patients of many ages, from 18 to 70 years, in our reported cases, age ranges from 38 to 59 years. Clinically, breast fibromatosis presents as a firm mobile palpable mass suspicious of malignancy that may adhere to the chest wall, sometimes with dimpling or retraction of the skin (2). In our patients, the only clinical finding was the presence of the nodule with no other sign or symptom, such as pain, skin retraction, or nipple secretion. There is no axillary lymph node at palpation. The lesions are most commonly unilateral (6, 7). Similar to that which was observed in our series.

Breast fibromatosis frequently shows typical mammographic and sonographic features that may be indistinguishable from those of breast cancer (6,11). Mammographically, breast fibromatosis appears as irregular uncalcified hyperdense masses with spiculated margins that mimic breast cancer (12). In one series, fibromatosis was visible in mammography in only one third of cases. The explanation is that lesions arise from musculoaponeurotic structures and this anatomic region of the body is not well explored by mammography. (7,13). By sonography, desmoid tumors are frequently poorly marginated, hypoechoic masses with a thick echogenic rim and posterior attenuation (12, 13). Benign appearances have also been reported and fibromatosis may be mistaken for a fibroadenoma (13). This case of benignlike sonographic and Mammographic findings was revealed in our previous series. There are few reports of the use of MRI in imaging of breast desmoid tumors. In

addition to being used as a diagnostic tool, MRI imaging also has a role in preoperative planning if a pathological diagnosis has been made prior to surgery or in cases of recurrent disease. By MRI, desmoid tumors appear as illdefined, hypo- to isointense masses on T1- weighted images and as heterogeneously hyperintense masses on T2-weighted images. They show suspicious, slow enhancement after contrast administration (14, 15). At histopathology it is important to differentiate fibromatosis from other breast spindle cell tumors such as fibrosarcoma or malignant myoepithelioma as treatment varies depending on the pathology. The lesions have irregular margins, with spindle cells infiltrating the parenchyma and surrounding normal parenchymal elements. The entrapped ductal and lobular units may show mild cellular atypia; however, the spindle cells are usually uniform with a low mitotic index. The pathognomic appearance of fibromatosis has been described as bland-looking spindle cells forming interlacing bundles with positivity of immunohistochemical markers for vimentin and smooth muscle actin supporting the diagnosis (15, 16, 17). Estrogen and progesterone receptor testing were not performed on the specimen in all case. A spindle cell lesion may be suspected on review of core needle biopsy material. When a preoperative diagnosis is suspected as a result of a core needle biopsy, appropriate preoperative planning can be conducted to direct a definitive, single-step surgery (7). One patient in our series underwent core needle biopsy when we suspected a neoplastic lesion and lumpectomy was indicated. The first line of treatment for breast fibromatosis is surgical excision with wide margins. There is agreement of how to surgically manage fibromatosis involving the breast alone, however in cases where the chest wall is involved there remains controversy as to how surgically aggressive one should be in order to balance between adequate excision and patient morbidity (7). Prior series have reported recurrence rates after surgery ranging from 18–27%, with most recurrences occurring within the first 2 years of treatment (7,18). Margin status has been associated with recurrence in a case series of breast desmoids reported by Wargotz et al (18). Other factors found in series of extra-abdominal desmoid tumors to be associated with recurrence include larger tumor size and younger age (19). In our series, the lesions ranged from 10 to 40 mm and there is no recurrence after a follow-up period ranging from 12 months to 16 years. The absence of recurrence is due to the wide margins ranging from 1 cm to 3 cm.

CONCLUSIONS

In breast fibromatosis, most patients present with a palpable breast mass, which is suspicious for malignancy both clinically and radiographically.

Needle core biopsy usually suggests a spindle cell lesion, this aids in preoperative planning to minimize the number of procedures patients require. Therapy for desmoid tumors of the breast remains surgical and no strong predictors of recurrence exist. It seems prudent to strive for negative margins at the time of surgical resection, if feasible.

However, when destructive procedures such as extensive chest wall resections are necessary to achieve a complete resection, observation or a trial of radiation or adjuvant therapy is reasonable.

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Table 1: Detailed summary of patient data

| PATIENTS | GENDER | AGE | SIDE | QUADRANT | SIZE (mm) | RADIOLOGIC SUSPICIOUS LESION | FOLLOW UP (year) |
|----------|--------|-----|-------|----------|--------------|---------------------------------|---------------------|
| 1 | F | 46 | RIGHT | UIQ | 40 | YES | 2 |
| 2 | F | 58 | RIGHT | UIQ | 12 10 | YES | 16 |
| 3 | F | 38 | RIGHT | LIQ | 10 | YES | 13 |
| 4 | F | 59 | LEFT | UIQ | 20 | YES | 10 |
| 5 | F | 44 | RIGHT | UB | 40 | NO | 1 |

UIQ: Upper inner quadrant, LIQ: Lower inner quadrant , UB :Upper breast.

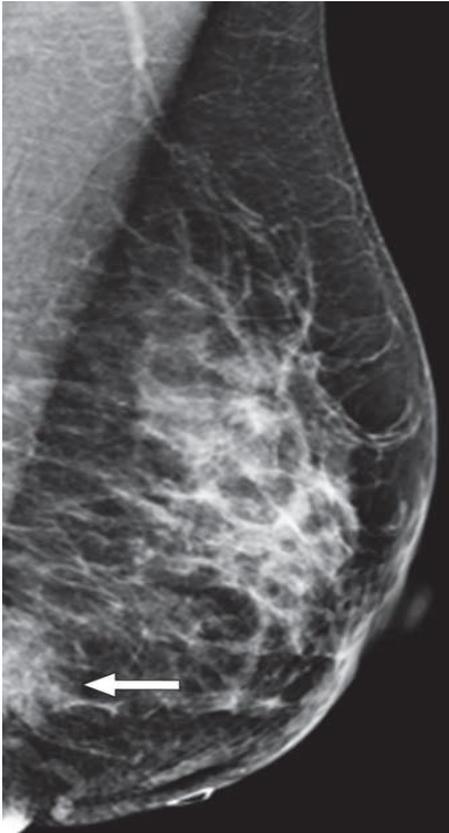


Figure 1: digital mammograms show irregular mass that is incompletely included on films

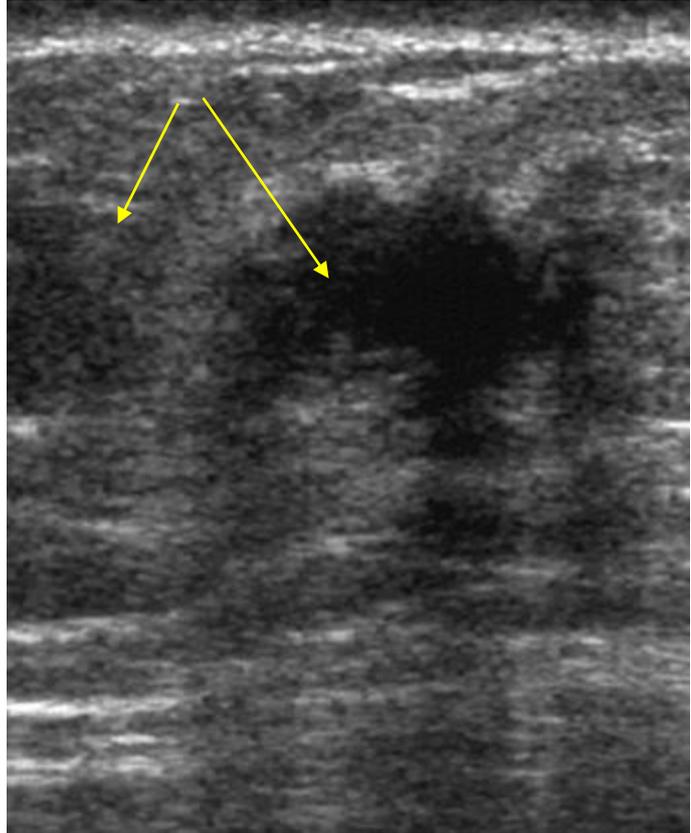


Figure 2 : Sonography image shows irregular, hypoechoic tumor

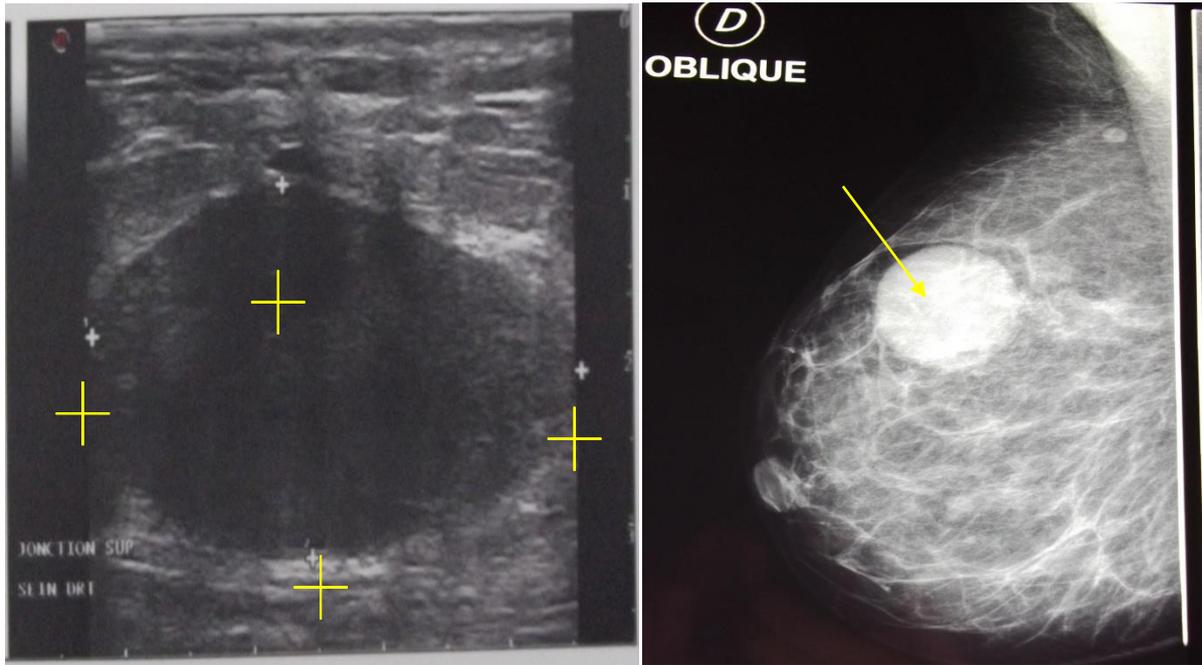


Figure 3, Figure 4: benignlike sonographic and Mammographic findings
fibromatosis may be mistaken for a fibroadenoma



Figure 5: Surgical specimen showing excision with wide margins

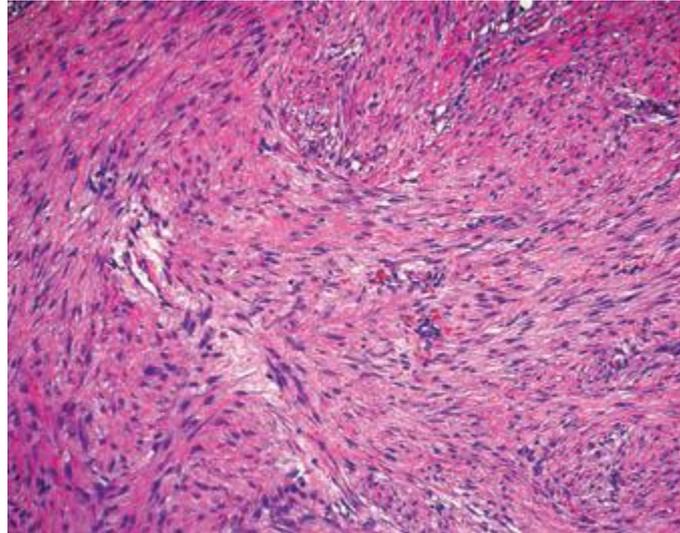


Figure 6: Uniform spindle cells without nuclear atypia are arranged in storiform configuration. (×400)