Case report

FIBROMATOSIS (DESMOID TUMOR) OF THE BREAST

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RESUMEN

El tumor desmoide (fibromatosis) es una entidad patológica extremadamente rara que se desarrolla de la fascia muscular y la aponeusorsis. Aunque sin potencial metastático, estos tumores son localmente muy agresivos y tienden a infiltrarse en los tejidos circundantes. Nosotros presentamos un caso de tumour desmoide de mama, que tuvo apariencias clínicas sugestivas a carcinoma. La paciente, de 56 años presentó una masa palpable de mama derecho. La citología por aspiracion con aguja fina (AGF) no detectó ninguna célula maligna, por lo que se hizo una escisión local conservadora. La paciente no recibió ningun tratamiento postoperatorio adicional, y continúa viva y sana en los siguientes 18 meses.

***Palabras claves**: localmente agresivo, tumor de mama, tumores mesenquimales.*

ABSTRACT

Desmoid tumor (fibromatosis) is extremely rare benign pathological entity that develops from muscular fasciae and aponeuroses. Although without metastatic potential, these tumors are locally very aggressive and tend to infiltrate the surrounding tissues. We present a case of a desmoid tumor of the breast that had clinical appearance suggestive of carcinoma. The patient was 56 years old female with a previous history of surgical trauma who presented with a palpable mass in the right breast. A fine needle aspiration (FNA) cytology did not reveal any malignant cells, thus conservative local excision was performed. The patient did not receive any additional postoperative treatment and was alive and free of disease after 18 months of follow-up.

***Key words**: locally aggressive, breast mass, mesenchymal tumor.*

INTRODUCTION

Desmoid tumor of the breast, also known as breast fibromatosis or aggressive fibromatosis, is extremely rare benign pathological entity that accounts for only 0.2% of primary breast tumours. This is a benign mesenchymal tumour that develops from fibroblasts and myofibroblasts within the breast parenchyma (Al-Yusuf et al, 2005). Despite its lack of metastatic potential, fibromatosis can grow aggressively in a locally infiltrating pattern (Povoski et al, 2006b; Rosen, 2009).

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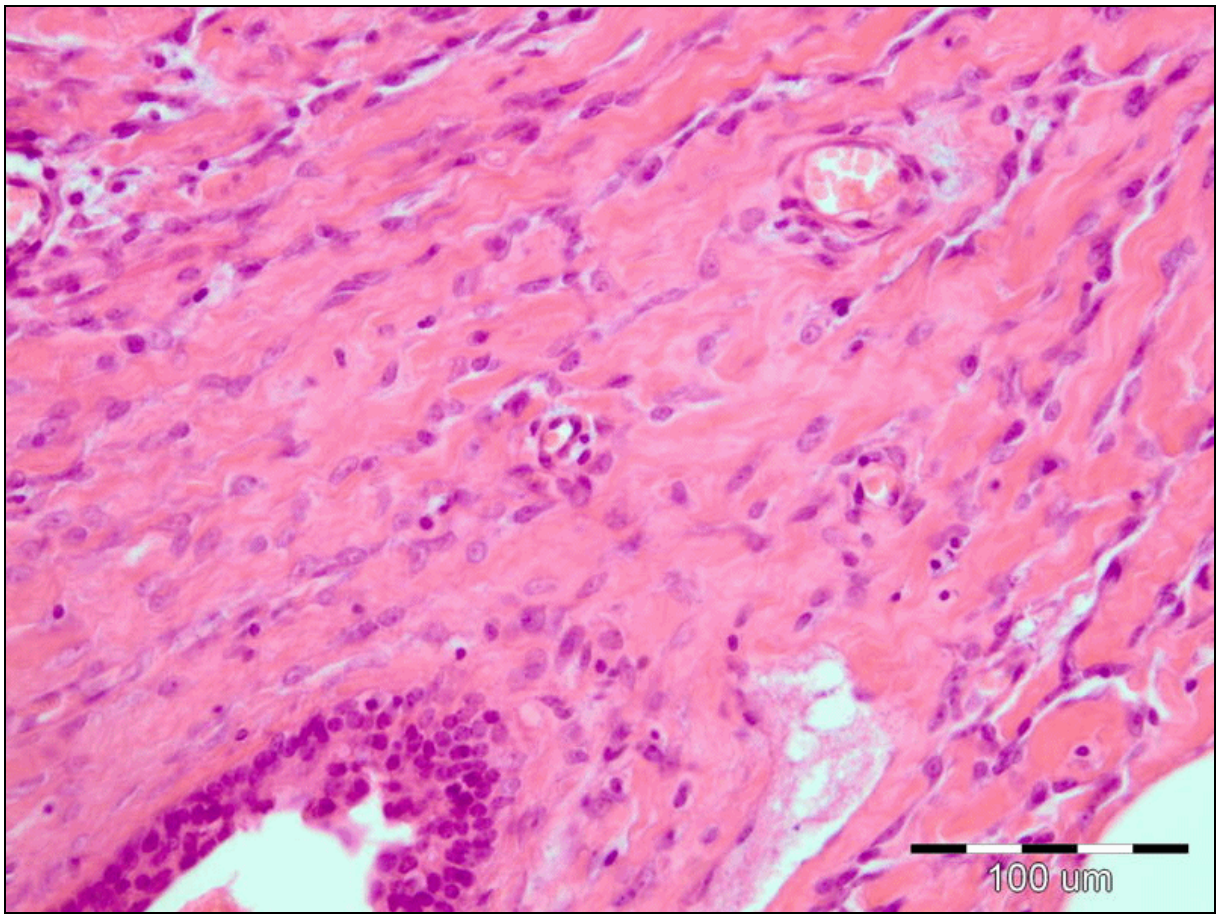

Figure 1. Monomorphic spindle cells arranged in loose fascicles (hematoxylin-eosin, x40)

CASE REPORT

We present a case of a 56 years old Caucasian female patient with a previous history of surgery to the right breast. The patient denied family history of familial adenomatous polyposis (FAP) or Gardner's syndrome. She presented with an uncomfortable, pricking sensation in her right breast and right chest wall region. There was a visible and palpable mass in the inferior aspect of her right breast. In the patient's history, there were two fine needle aspiration biopsies (FNAs), both followed by surgical excisions (lumpectomies) 20 years ago and 5 years ago respectively. Both surgical procedures were performed in the same location as the current tumor.

The two FNAs were reported as benign changes, whereas the materials submitted for histo-pathological analysis from both excisions were reported as fibroadenomas.

Upon physical examination, the two scars from the previous surgical excisions were seen on the inferior-lateral aspect of the patient's right breast. The palpable mass underneath the scars had firm hard consistency and measured, approxim-ately, 4 cm. The tumor clinically appeared to be adherent to the underlying chest wall structures, but was not attached to the skin. The nipple, areola, breast skin and right axilla were normal. The left breast and axilla were normal.

The mammography revealed an area of increased parenchymal density measuring 4.5 x 4 cm in the inferior-lateral aspect of the right breast in the oblique view. There were no microcalcifications or skin thickening. Ultrasound examination showed well-defined hypoechogenic mass lesion close to the chest wall.

Fine needle aspiration biopsy of the mass revealed no malignant cells and was reported as suggestive of fibroadenoma. Nevertheless, a possibility of malignancy was still considered

based on the clinical presentation. Wide local excision of the tumor was performed.

On gross pathologic examination, the specimen measured 5.5 x 5.0 x 2.5 cm in size. The cut surface revealed ill defined, grayish white tumor with fatty streaks. The tumor measured 4.3 x 4.0 x 2.0 cm, surrounded by fatty tissue. Macroscopically, surgical margins were not involved by the tumor.

The specimen was fixed in 10% neutral buffered formalin and processed in routine fashion. Three microns thin sections were cut from the paraffin embedded tissue and stained with hematoxylin and eosin. For the immunohistochemistry, ready-to-use peroxidase-based EnVision™+ kit (K5007) and primary antibodies from Dako (Glostrup, Denmark) was used. For the alpha smooth muscle actin stain, clone 1A4 in dilution 1:400 was used. Polyclonal S100 antibody was used in dilution 1:100. The MIB-1 clone of Ki-67 antibody was diluted 1:400. The reaction product was detected with 3.3'-Diaminobenzidine chromogen (DAB).

Microscopically, the lesion had infiltrative margins into adjacent adipose tissue. The tumor was composed of monomorphic spindle cells arranged in loose fascicles in a sweeping fashion (Fig. 1). Occasional perivascular lymphocytes were noted. The fascicles of spindle cells were separated by keloid-like collagen fibers (Fig. 2). Mitotic activity was inconspicuous. Surrounding breast tissue showed features of fibrocystic change and foci of proliferative breast disease. Excision margins were uninvolved (Fig. 3). Immunohistochemically, the spindle cells were positive for smooth muscle actin (Fig. 4) and S100 protein, whereas and Ki-67 proliferative index was 2% (Fig. 5). The histological features were consistent with the diagnosis of fibromatosis (desmoid tumor) of the breast.

After 18 months of follow up, the patient was clinically free of tumor.

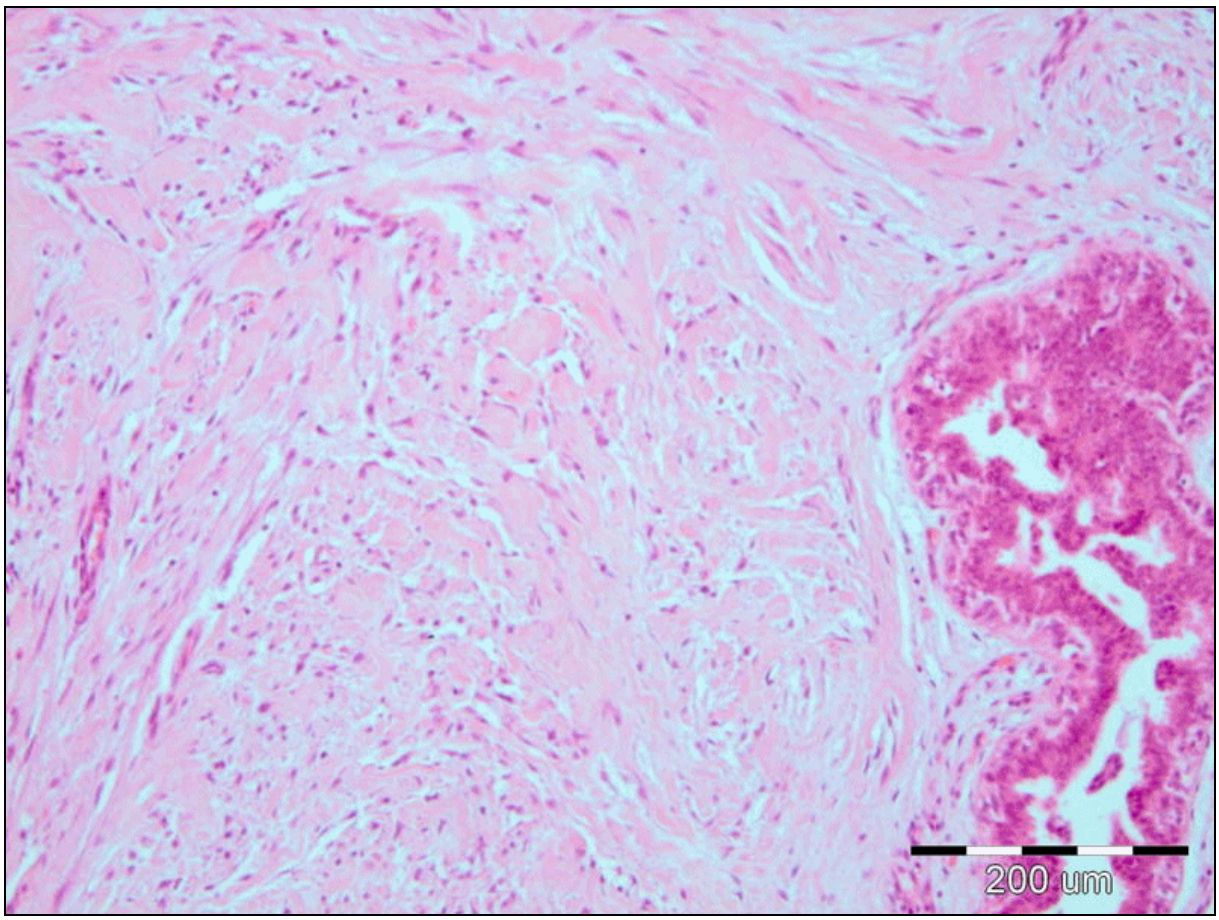

Figure 2. Spindle cells were separated with bundles of keloid-like collagen (haematoxylin-eosin, x200)

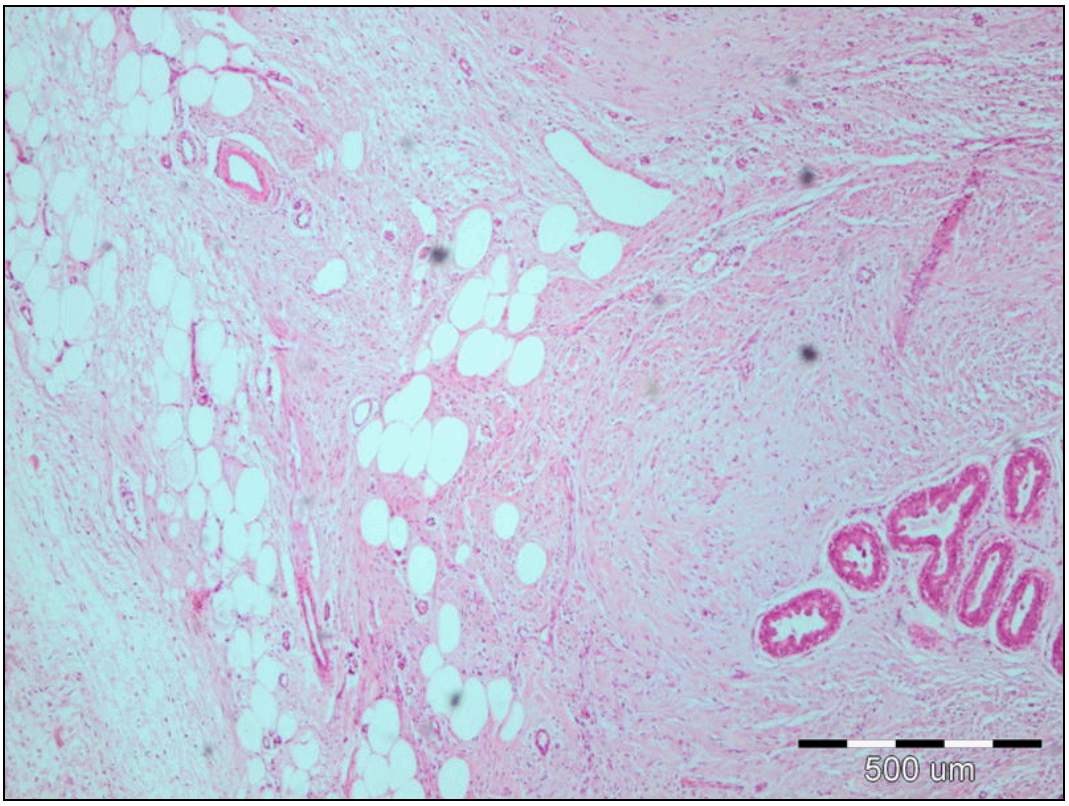

Figure 3. Surgical margin were not involved by the tumour (haematoxylin-eosin, x40)

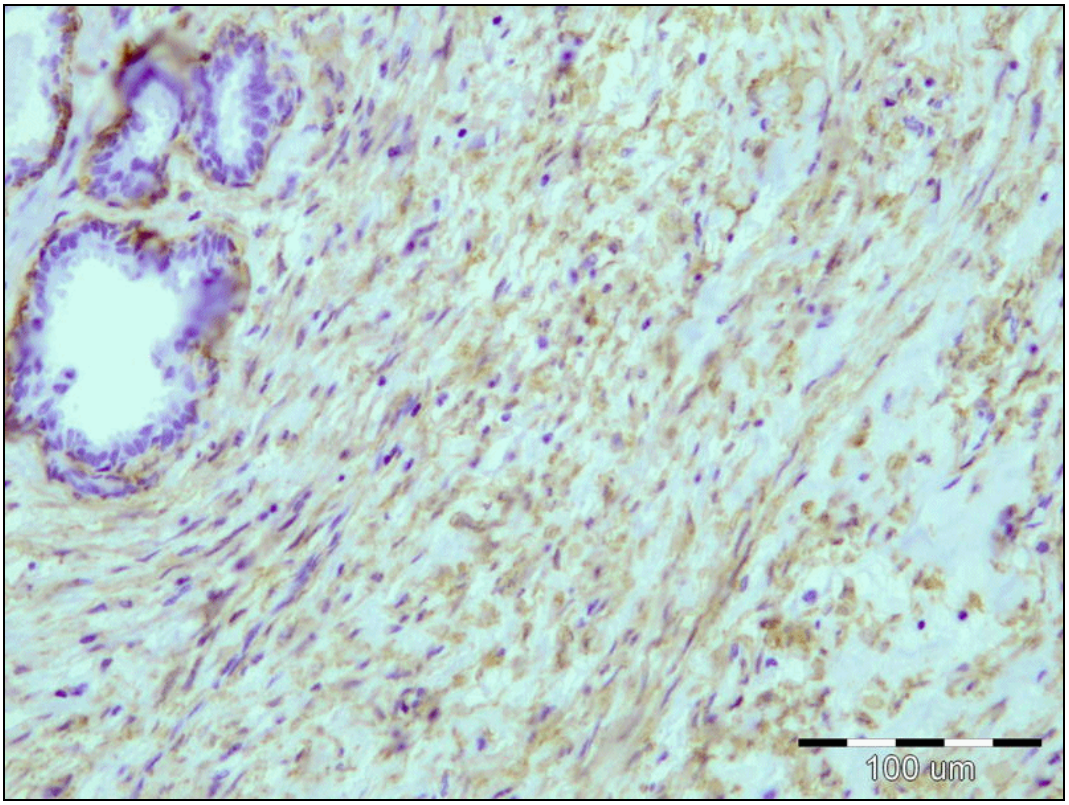

Figure 4. Spindle cells were positive for smooth muscle actin (smooth muscle actin, x200)

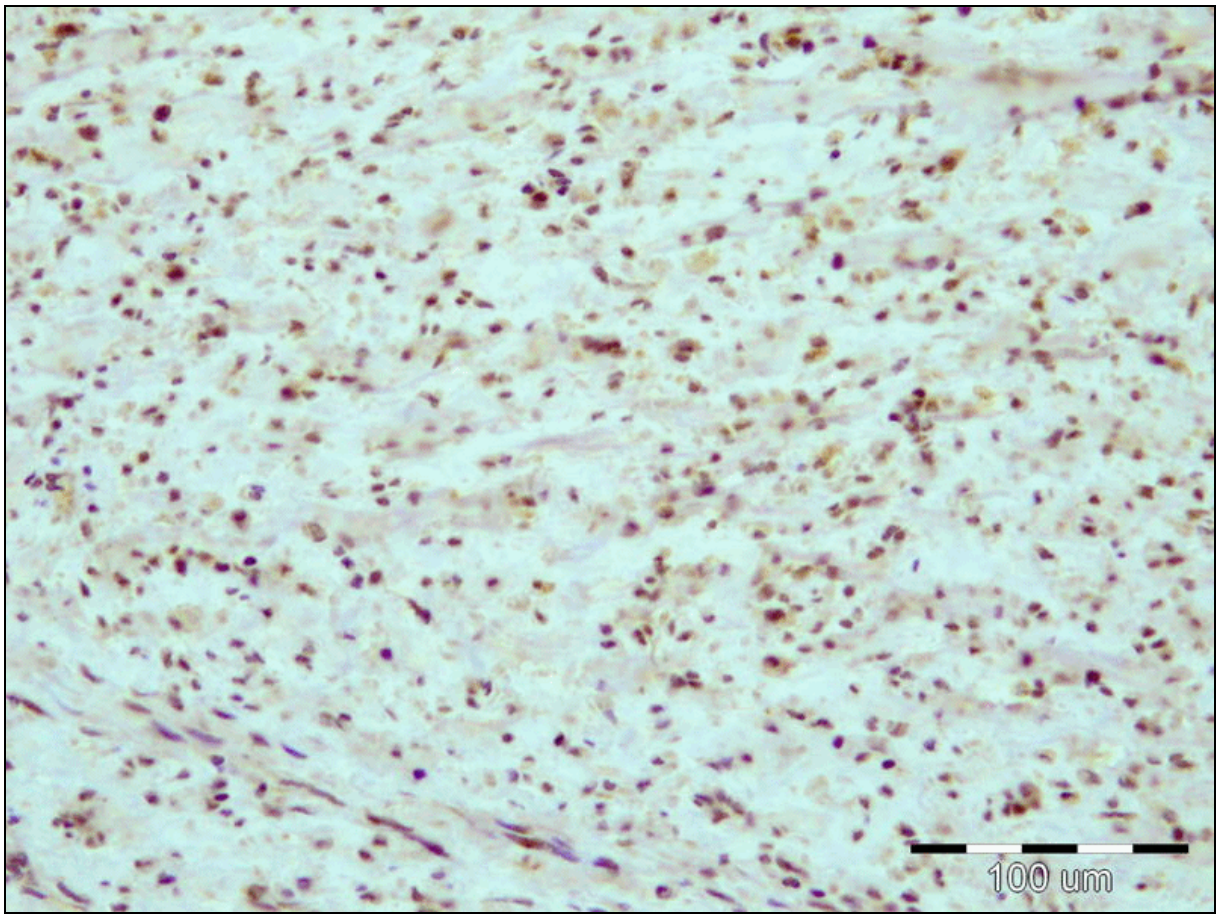

Figure 5. Spindle cells were positive for S100 protein (x200)

DISCUSSION

The incidence of desmoid tumor in general population is very low (2-4 cases per 1.000.000 inhabitants). It accounts for 0.03% of all tumors (Suarez-Artacho et al, 2009) and for less than 0.2% of all primary breast neoplasms (Al-Yusuf et al, 2005; Chummun et al, 2010). Its etiology is unknown, but it has been associated with surgical trauma, minor trauma of the breast, certain genetic disorders, adenomatous polyposis coli (APC) gene or beta-catenin mutation in patient with FAP (Abraham et al, 2000). Since the tumor cells express estrogen and progesterone receptors, hormonal alterations were also linked to the occurrence of these tumors (Erguvan-Dogan et al, 2005; Povoski et al, 2006a, b; Suarez-Artacho et al, 2009; Villarreal-Colin et al, 2008). Several authors reported possible association between augmentation mammoplasty and the development of fibromatosis in the capsule around the breast implant (Chummun et al, 2010; Henderson et al, 2010; Matrai et al, 2011).

This case had previous history of surgical trauma.

On histological examination, the tumor is composed of spindle cells (fibroblasts and myofibroblasts) and collagen fibers (Suarez-Artacho et al, 2009). The edges are characteristically infiltrative, with extension into the surrounding structures, which differentiates this lesion from fibroadenoma. Glandular elements are not present in the tumor and can only be found engulfed at the periphery of the lesion. Mitotic activity is inconspicuous, a finding that helps to differentiate fibromatosis from fibrosarcoma (Serpell et al, 1999).

In our case, the tumor had the characteristic appearance of spindle cells intermingled with collagen bundles. The tumor also had infiltrative borders and the mitotic count was very low.

Fibromatosis arising in the breast parenchyma apparently represents a separate entity from

extra mammary fibromatosis. The latter entity most probably arises from the muscular fascia or aponeurosis, whereas the former appears to originate from fibroblasts and myofibroblasts within the breast parenchyma (Al-Yusuf et al, 2005; Muller et al, 2011).

Fibromatosis can clinically, mammographically, and ultrasonographically mimic breast cancer (Al-Yusuf et al, 2005; Povoski et al, 2006a, b). Mammography usually reveals a stellate tumor indistinguishable from carcinoma. In our case, mammography and ultrasound were not very helpful in differentiating this tumor from carcinoma.

According to our experience, pre-operative diagnosis of fibromatosis by FNA cytology is impossible, but it allows exclusion of breast cancer and other more common diseases. Definite diagnosis can only be made on paraffin sections.

The main differential diagnostic dilemma is between fibromatosis and other benign and malignant entities, such as nodular fasciitis, fibrosarcoma, spindle cell carcinoma low grade fibromatosis-like variant and infiltrating myo-epithelioma (Al-Yusuf et al, 2005; Moinfar, 2007; Rosen, 2009).

Early recognition of fibromatosis and appropriate complete wide local excision with free margins is widely advocated in the literature because of a high propensity towards local recurrence if incompletely excised (Al-Yusuf et al, 2005; Povoski et al, 2006b; Suarez-Artacho et al, 2009). Local desmoid tumor recurrence rates are reported to be as high as 70%. A positive surgical margin is a significant risk factor for recurrence. After surgery, magnetic resonance imaging (MRI) may be useful for monitoring desmoid tumor recurrence (Choi et al, 2005; Dashiell et al, 1978).

Adjuvant antihormonal therapy does not reduce the local recurrence rate, but postoperative radiation therapy can improve the disease-free survival rate (Erguvan-Dogan et al, 2005; Ray and Lawrence, 2006).

In conclusion, mammary fibromatosis or desmoid tumor of the breast is a rare benign spindle cell lesion that mimics malignancy both clinically and radiologically, due to its locally infiltrative growth pattern. Awareness of such lesion and its distinction from breast cancer both clinically and histologically is of great importance, since breast conserving complete local excision is a treatment of choice for these tumors.

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