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(CASE REPORT)

Desmoid fibromatosis of the breast: A case report

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Abstract

Desmoid fibromatosis of the breast is a rare benign condition of the breast (0.2% of breast tumors) that clinically and sonographically mimics breast cancer.

The diagnosis is confirmed by histology, allowing the elimination of the main differential diagnosis of this entity which is metaplastic spindle cell carcinoma.

Breast fibromatosis is characterized by a local evolution and a tendency to recurrence, hence the interest of surgical excision with healthy margins. Radiation therapy should be discussed in such cases.

Through our case and the review of the literature, we will try to focus on the diagnosis of this rare entity and its management because it influences the prognosis.

Keywords: Fibromatosis; Breast; Diagnosis; Risk; Surveillance

1. Introduction

Desmoid tumor is a benign mesenchymal tumor developed at the expense of fascia and muscle fascia. Called desmoid tumor because of its resemblance to tendons, fibromatosis is a histological form close to low grade fibrosarcomas. It is characterized by a strictly local evolution and its tendency is to recur without ever giving metastasis. This pathology clinically and radiologically mimics breast cancer. Only histology will provide the diagnosis. Surgical removal of the tumor with healthy margins is the only treatment. In case of incomplete removal, the risk of local tumor recurrence is increased, but without ever giving rise to metastasis [2].

Desmoid tumor of the breast is an even rarer entity, representing less than 0.2% of breast cancers and 4% of extra-abdominal fibromatoses.

2. Clinical case

2.1. Informations

Mrs FH, a 53-year-old girl, without any notable pathological history, was followed in our consultation for a right breast nodule since February 2021.

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2.2. Clinical examination

The physical examination showed a mass of 3 cm, in the superior-internal quadrant of the right breast, painless, firm, mobile in relation to the two superficial and deep planes and without retraction or nipple discharge, lymph node areas free.

2.3. Paraclinical examination

A mammography + breast ultrasound was done and objectified breasts of fat density type 3 with focal asymmetry with a lesion of the superior-internal quadrant of the right breast measuring 33 mm Right breast Birads 4 Left breast Birads1. (Figure 1 +2).

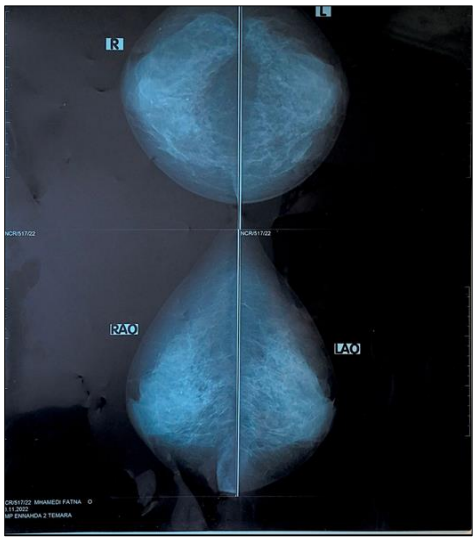

Figure 1 Mammogram showing the lesion in the superior-internal quadrant

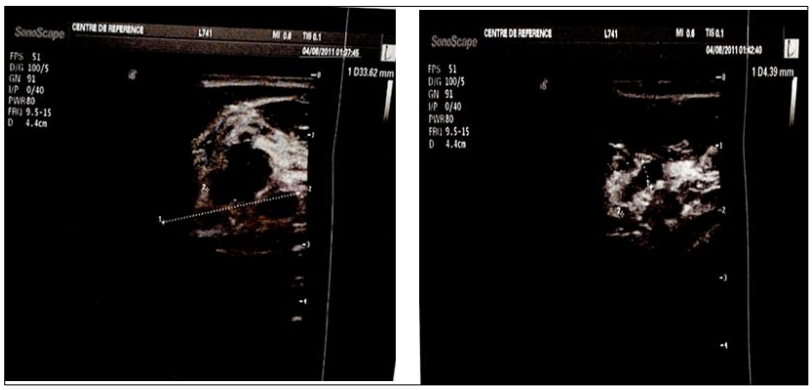

Figure 2 Ultrasound showing the lesion in the superior-internal quadrant

A microbiopsy was performed but came back inconclusive

2.4. Procedure

A large lumpectomy of the right breast was performed.

2.5. Follow-up and outcome

The pathological report concluded to a desmoid tumor measuring 2.5*1cm long axis with fuso-cellular proliferation made of elements of variable density arranged in more or less undulating bundles and punctuated by vessels with slightly thickened walls with or without peripheral retraction. The excisional borders were healthy.

Close monitoring was performed, unfortunately the patient was lost to follow-up.

3. Discussion

Desmoid fibromatosis of the breast represents 0.2% of primary breast tumors and 4% of extra abdominal fibromatoses [1]. It affects women aged between 13 and 80 years with a peak frequency between 30 and 50 years [3]. Its origin is not well known. Three etiologies are suspected: traumatic, hormonal or hereditary origin. In some cases, a genetic component is present; two percent of these tumors are of genetic origin, by mutation of the adenomatous polyposis coli (APC) gene, and fall within the framework of Gardner syndrome associated with familial adenomatous polyposis (FAP) [3]. Cases have been described after trauma, or after breast surgery such as breast reduction surgery [4,5], or after breast prosthesis [6,7]. Unlike abdominal desmoid tumors, breast fibromatosis does not seem to be associated with pregnancy [8]. Mammary fibromatosis is a benign tumor of the breast, but its clinical, mammographic and ultrasonographic presentation is often very suspicious [9].

Clinically, breast fibromatosis presents as a firm, poorly defined, painless, unilateral nodule of variable size (3 cm on average), more often peripheral than peri-areolar, sometimes associated with skin or nipple retraction. This lesion may be painful, especially if it infiltrates the deep layers. Nipple discharge is rare and patients do not have palpable adenopathy. The involvement may rarely be bilateral. Imaging often takes on the appearance of a malignant tumor. The images observed on mammography are suggestive of a malignant lesion (asymmetry of density, poorly defined opacity with blurred and irregular contours, spicules). The most common mammographic appearance of fibromatosis is that of a non-calcified spiculated mass, which is observed in the second patient of our series. Micro or macrocalcifications are rare and correspond to associated breast lesions. On ultrasound, fibromatosis appears as a solid, spiculated or microlobulated, irregular, hypoechoic mass with posterior attenuation, mimicking a malignant lesion [13]. Magnetic resonance imaging is of interest to assess possible parietal invasion [7]. Although few studies have been performed in MRI for fibromatosis, generally, it is a poorly contoured mass, often spiculated, isointense to the muscles, in iso/hypersignal T2 of variable intensity, and heterogeneous. The kinetics of enhancement is variable, sometimes with a progressive benign appearance, sometimes more suspicious in plateau, as in our first case, or with a washout [5].

The preoperative diagnosis can be evoked at microbiopsy but most often, it is affirmed after diagnostic surgery, which is the case in our first patient. Pathologically, desmoid fibromatosis presents macroscopically as a fairly well circumscribed lesion, with an indurated, fasciculated, pearly consistency and varying in size from 5 to 10 cm in diameter. Histological examination shows a pauci cellular proliferation with spindle cells. These cells are fibroblastic and myofibroblastic in appearance, arranged in long parallel bundles within a collagen-rich, often wavy background. The cells usually do not show nuclear abnormalities and mitoses are rare. The poor peripheral limitation with infiltration of the adjacent breast tissue in a finger-like pattern is an important diagnostic feature. There are a few vessels, small and often surrounded by a clear space. Peripheral hemorrhagic raptus and lymphoid clusters are also seen [15]. The immunohistochemical study shows an intense and diffuse expression of smooth muscle actin and beta-catenin by the tumor cells. Estrogen and progesterone receptors are not detectable by immunohistochemistry in mammary fibromatosis, like its extra mammary counterpart. The differential diagnosis is mainly with fibromatosis-like metaplastic carcinoma, as they share many similarities: (1) They are both infiltrative. (2) Both are rich in collagen. (3) Tumor cellularity is low. (4) The cells are not as pleomorphic as the other tumors.

It probably has its indication in case of recurrence and contraindication to surgery or radiotherapy. In conclusion, the clinical, radiological and histological manifestations, and the therapeutic conduct of our two observations were consistent with the literature. It is a very rare pathology that deserves to be studied and known because of its clinical and radiological similarity with breast cancer. The main treatment is surgery with healthy margins since this tumor has a high potential of recurrence without ever giving metastasis. Adjuvant treatments are being evaluated due to the small number of patients affected. Both patients are satisfied with the results and are continuing their follow-ups in our outpatient clinic. They have no tumor recurrence to date.

4. Conclusion

Breast fibromatosis is a benign infiltrating tumor. Its radiological appearance is suggestive of a malignant tumor. Its diagnosis is anatomopathological and its surgical removal must be large and performed in healthy areas in order to reduce the risk of recurrence.

Compliance with ethical standards

Disclosure of conflict of interest

The authors report no declarations of interest.

Statement of Ethical approval

Exempt from ethical approval in my institution.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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