



Case report

Desmoidfibromatosis of the breast after breast reduction: A case report and a review of the literature

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ABSTRACT

Introduction: Desmoids fibromatosis is a rare pathology whose definitive diagnosis is histological. Although it presents clinical and radiological similarities with breast carcinoma.

Case report: In this observation, we report a case of desmoids fibromatosis diagnosed in a 51-year-old female patient. The positive diagnosis was difficult to retain.

Discussion: Desmoids fibromatosis remains a benign pathology with a high recurrence rate and a local aggressiveness requiring an enlarged lumpectomy with sufficient margins up to 3 cm.

Conclusion: Radiotherapy remains a therapeutic option in addition to surgery in incomplete sections and in case of multiple recurrences. The place of medical treatments, in particular anti-estrogens, is not clearly defined.

1. Introduction

Desmoid fibromatosis was first described in 1832 [1] as a benign mesenchymal tumor, developed from musculoaponeurotic structures, characterized by its infiltrative power and its tendency to local recurrence without metastatic power [2]. The breast localization is very rare and represents only 0.2 % of all breast tumors [3], some authors consider it as a subgroup of soft tissue sarcomas and do not differentiate between aggressive fibromatoses and low grade fibrosarcomas [1].

The aim of this work is to highlight the particularities of this entity which remains rare through a case report and a review of the literature. All our work was reported according to the SCARE criteria and guidelines [14].

2. Case report

A 51-year-old female patient, followed for a right breast cancer diagnosed at the age of 49 years for which she received a right mastectomy and axillary lymph node dissection and contralateral breast reduction. It was a 4 cm infiltrating ductal carcinoma, SBR III Luminal B, 0 N+/20 N with presence of fibrous mastopathy without evidence of malignancy at the left breast reduction specimen. The patient received adjuvant chemotherapy, external radiation therapy and hormonal

therapy. One year after surgery,

Two years later, she consulted for a nodule in the left breast measuring 4 × 2 cm in the upper medial quadrant. (Fig. 1), echomammography classified the lesion as ACR 5 (Fig. 2), and biopsy confirmed the diagnosis of breast desmoid fibromatosis. A lumpectomy was performed, but the depth limit was less than 1 mm. Therefore, the decision was made to take the patient back for a mastectomy (Fig. 3).

3. Discussion

Breast tumor is very rare with a frequency of no more than 3.5 % of fibrinoid tumors, and 0.2 % of all breast tumors [4–5]. It affects patients between 20 and 40 years of age. The pathophysiology is still poorly understood, with probably the triggering of cell proliferation following trauma or after breast surgery such as breastplasty [4–7], or after breast prosthesis [8–9], with sex hormones playing a promoting role. Familial fibromatosis has been described in the context of a GARDNER syndrome associating colonic polyposis, bone malformations, squamous cysts and soft tissue tumors, including desmoid tumors [1].

Clinically, this tumor presents as a mass, firm, variable in size, painless [1]. Radiologically, a spiculated opacity having the criteria of malignancy, one can even objectify an involvement of the pectoral or intercostal muscles, it thus mimics at the clinical and radiological level a

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Fig. 1. Clinical aspect.

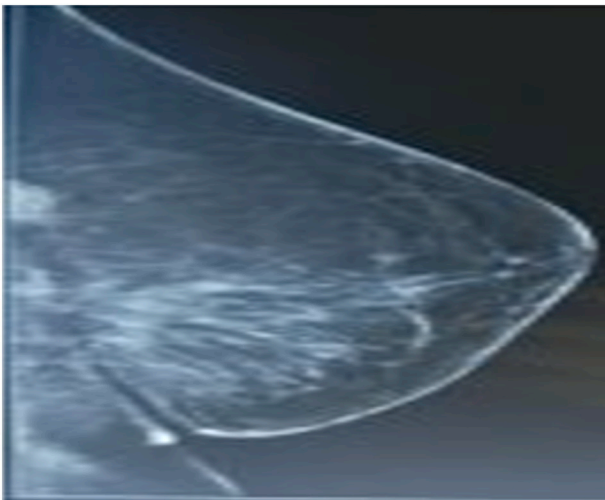


Fig. 2. Radiological aspect.

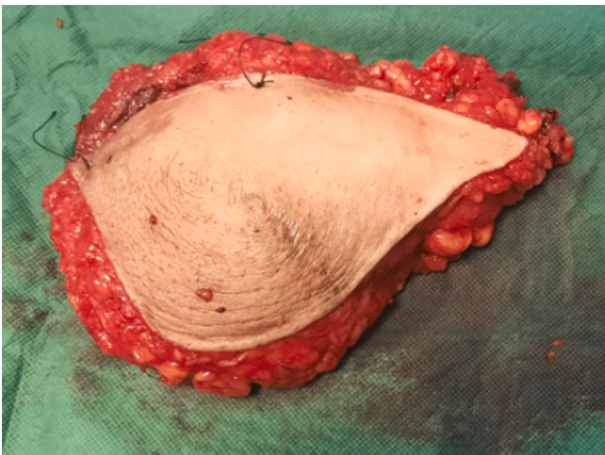


Fig. 3. Mastectomy.

malignant lesion [10]. Histological, it is a firm, whitish lesion, not very limited, with, on microscopic examination, a proliferation of spindle-shaped cells (fibro and myofibroblastic without lymphocytes), arranged in bundles, mixed with bands of collagen, without any associated

epithelial component [1].

On immunohistochemistry, the cells express muscle actin and beta-catenin. Estrogen and progesterone receptors are not detectable by immunohistochemistry in breast fibromatosis [1].

The differential diagnosis is mainly with metaplastic spindle cell carcinoma. It can also be made in the presence of phyllodes tumor of intermediate or malignant aggressiveness, fibrosarcoma or myoepithelial carcinoma, or fibrous carcinoma, since the same spindle cells are involved, but it is the inflammatory contingent present in the case of fibrous carcinoma that makes the difference [1]. The treatment is essentially based on complete surgical excision with healthy margins (up to 3 cm) [11–12]. Recurrence is frequent (18–29 %, 3–6 years) and thoracic muscle and rib involvement is possible [1]. In case of chest wall involvement, a more mutilating excision involving muscles and even ribs is most often performed. A wide excision with healthy safety margins is associated with a significantly higher 5-year recurrence-free survival rate than the opposite case. According to the 2015 CNGOF recommendations [13]. Mastectomy is recommended in case of multiple recurrences, in case of large tumor volume or in case of difficulty in histological diagnosis [1]. The place of radiotherapy is still controversial in the literature, its efficacy is dose-dependent, and tumor control is 60 to 80 % for a total dose of 50 to 60 GY administered. Other adjuvant treatments have also been tried: anti-inflammatory drugs, anti-estrogens and low dose chemotherapy [1]. Positive.

Resection margins, younger age and larger tumor size are also associated with an increased risk of recurrence [13].

4. Conclusion

Breast fibromatosis is a rare entity which, clinically and radiologically, suggests cancer. Only histology will allow the diagnosis to be made. It is a benign fibroblastic proliferation, of strictly local evolution and with a high recurrence capacity. Its treatment consists of a complete removal with safety margins. The role of radiotherapy and medical treatments, in particular anti-estrogens, is not clearly defined and seems limited in this location.

Provenance and peer review

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Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

I declare on my honor that the ethical approval has been exempted by my establishment.

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Nabila youssouf: Corresponding author writing the paper.
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 BOUFETTAL Houssin: correction of the paper.
 MAHDAOUI Sakher: correction of the paper.
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Declaration of competing interest

The authors declare having no conflicts of interest for this article.

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