

Bilateral breast fibromatosis in a 41 years old woman

Abstract

Breast fibromatosis is a rare benign tumor of soft tissues with local spread, frequently recurrent but with no metastatic evolutions. We present a case diagnosed in a 41 yearsold patient referred to a Douala private medical center in February 2012, presenting bilateralbreast tumor. Histological analysis of the excised samples showed fibroblastic proliferation without atypia and metastasis which reappeared 15 months after the initial intervention in the left breast. A mastectomy and lymph nodescurettage has shown resolution of the symptoms. *.Key- words:* fibromatosis - histopathology –breast -Cameroon

Introduction

Fibromatosis is a rare tumor of the breast in females between puberty and menopause. That tumor can arise from the fascia musculo-aponeurotic of the chest wall or from fibroblasts and myofibroblasts of the mammary parenchyma [1, 2].The neoplasm is characterized by being locally aggressive and frequent recurrence without metastasis [3, 4]. The mammary fibromatosis represents only a particular aspect of these tumor, its frequency is less than 0.2% of the primitive tumors of the breast and 4% of the extra- abdominal fibromatosis [3]. Bilateral desmoid tumors have been reported in about 4% of patients [3]. The clinical presentation is a palpable mass that is sometimes associated with skin modification [5]. The diagnosis is based on histological examination of the biopsy or surgical specimen [1, 2, 6]. We are reporting a case of bilateral desmoid tumor of the breast. The informed consent from the patient for this study was obtained.

Case presentation

A 41 years old female patient was referred to a private medical center in Douala town because of bilateral breast masses, a mass in the inferior external quadrant on the right and mass in the upper external left breast. In the two cases, the masses were mobile, painless and they appeared polylobular with no apparent changes in skin pigmentation and without any suspecting lesion.

Mammography showed large opaque masses with regular contours in both cases. The right breast measurement was 10x6x4 cm and the left was 11x7x5 cm. The cytological examination of the tissue aspirate taken under ultra-sound guidance showed abnormal cells.

The excised samples measured a 9x7x5 cm and weighted 58 g in the right breast. In the left, we obtained the sample measurement of 7x4x3 cm and 46 g in weight (Figure 1). There masses were of firm consistency, well circumscribed with smooth and whitish surface homogenous on different sections. The specimens were treated according to the technical routine with fixation in 10% formalin, staining in eosin-haematein and interpretation under optical microscope.

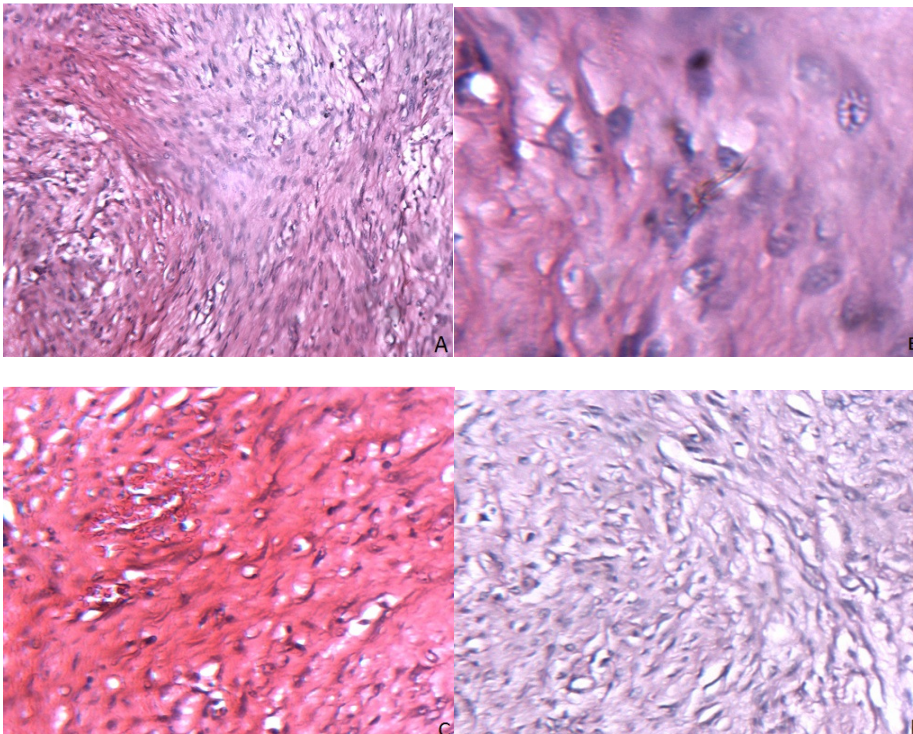


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39 **Figure 1.** Left Breast fibromatosis resected specimen

40 In histological examination, the lesion was characterized by well and differentiated
 41 fibroblastic proliferation, which was rich in collagen without atypia, no images of mitosis. In
 42 some areas, simple mesenchymatous tissue and few blood vessels were observed (figure 2)

43 .



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45

46 **Figure 2.** Diffuse and highly cellular fibroblast proliferation, without atypia or mitosis
 47 (A). Proliferation of low cellularity in this microscopic field, without atypia or mitosis (B).

48 Medium density cell proliferation without atypia or mitosis (C). Medium density cell
49 proliferation (D)

50 The patient was carefully followed up. 15 months after the initial operation, the patient was
51 noticed with a recurrence on the left breast. A left radical mastectomy was done. The
52 histological aspect was similar to the previously described. The right breast and the left
53 axillary nodules were normal. A careful follow-up over 26 months did not show any
54 recurrence.

55 **Discussion**

56 Desmoid tumors (DT) also known as aggressive fibromatosis (AF) constitute a rare
57 fibroblastic proliferative disease. They may occur in any musculoaponeurotic or fascial tissue
58 [7]. These tumors generally are divided by anatomic designation as extra-abdominal,
59 abdominal, or intra-abdominal and the most common locations are around the limb girdles or
60 the proximal extremities, the abdominal wall and intra-abdominal or mesenteric [8, 9]. The
61 mammary fibromatosis is a rare lesion; bilateral type have been reported in up to 4% of
62 patients and mostly it occurs in the breasts of women between 13 to 80 years with an average
63 of 43 years, regardless of their ethnicity [2,3,5]. In our presented case, the breast location, the
64 bilateral position of the tumor and the age of the patient, confirm in the data of the literature
65 makes it a rare and interesting case.

66 Clinically, our patient presented bilateral breast masses, a mass in the inferior external
67 quadrant on the right and another mass in the upper external left breast, associated with
68 painless and no apparent changes in skin pigmentation with no suspecting lesion .
69 Wongmaneeerung et al described two cases of bilateral breast fibromatosis with pain (of the
70 patients) and nipple retraction, the presence of skin involvement and invasion by the tumor
71 (in another patient) [5]. Mehdi et al reported a case with ulceration of all the nipple-areolar
72 plate [1]. That means the clinical manifestations of breast fibromatosis are variable, making
73 each case exceptional.

74 Although etiopathology is not fully understood, some theories had been proposed. Most
75 desmoids arise sporadically, slightly more in women than in men, with some DTs related to
76 pregnancy and trauma, and others associated with hereditary cancer syndromes [10]. Desmoid
77 tumors are results of deregulation of connective tissue growth. Increased nuclear expression
78 of β -catenin, a protein responsible for regulation of gene expression, proliferation and
79 survival, is the characteristic feature in sporadic DT [10]. Antecedent trauma, often surgical,
80 has been noted at the site of the DT in approximately 25% of cases [11]. Estrogen receptors
81 (ER) were observed in 33% of all DT examined, with an equal incidence in males and
82 females and with antiestrogen binding sites found in 79% of samples, including some which
83 were ER negative [11]. Implant-associated breast desmoid tumors may also occur [12].

84 In this case, mammography showed large opaque masses with regular contours in both breast.
85 In the literature, Mammography show typically high density, speculated and stellate tumor
86 without microcalcifications, often indistinguishable from carcinoma [4, 13]. The lesion
87 sometimes appears as lobulated mass or mass with no outlines, but can still be well de-fined.

88 In some rare cases, the lesion has no mammographic presence [14, 15]. Although Ultrasound
89 appearances are various, they are not specific. Desmoidtumours could be poorly marginated; it
90 can be irregular shaped hypoechoic mass or small-sized lesions that are homogeneous with
91 regular outlines [15, 16]. By some authors, the magnetic resonance imaging (MRI) and the
92 scanner are important for the estimation of a possible parietal invasion; MRI can also play an
93 invaluable role in preoperative diagnosis and planning. Desmoid tumors are typically
94 isointense on T1-weighted imaging and demonstrate low to high signal intensity on T2-
95 weighted imaging. After the injection of chelated forms of gadolinium, in T1-weighting, and
96 fat signal saturation, a heterogeneous raise in the ill-defined outlines should be observed [12,
97 15].

98 Based on available reports in the literature, fine needle aspiration cytology of the entire
99 specific may reveal some important information in patients with breast fibromatosis [17].
100 Acytoponction could confirm the diagnosis by isolating fibroblasts with minimal atypia;
101 however, it is mostly a little contribution [2]. In our case, the cytological examination only
102 showed abnormal cells.

103 The histological examination remains the basis for the diagnosis of this pathology. For our
104 patient, histologically they found well and differentiated fibroblastic proliferation, which was
105 rich in collagen without atypia, with no images of mitosis. In some areas, simple
106 mesenchymatous tissue and few blood vessels were observed. According to the literature, the
107 tumor usually has a poorly circumscribed pattern and is composed of proliferating stellate to
108 spindle cells arranged in long fascicles or whorling patterns with bland nuclear features and
109 dense keloid-like collagen in areas [10]. Ultrastructural studies indicate that the spindle cells
110 have features of both fibroblasts and myofibroblasts. Architecturally, the tumor cells are
111 typically arranged in long, sweeping fascicles, and vague whorls in a background of
112 eosinophilic, collagenized stroma with prominent thin-walled vessels. The stroma can also
113 show myxoid features, reportedly more common in the breast and the mesentery and can be
114 seen in other more common sites [11].

115 Immunohistochemistry, specifically β -catenin, and more recently, molecular diagnostics can
116 play an important role in its diagnosis. The presence of actin and vimentin is useful for the
117 diagnosis of desmoid tumor. Desmin is rarely positive, S100 and CD34 are usually negative;
118 β -catenin nuclear staining is an option for diagnosis, which may be only focally
119 positive. Sporadic DTs are commonly associated with somatic mutations of the codons 41, 45
120 of exon 3 of the beta-catenin gene (CTNNB1). As such, antibody of β -catenin is useful in
121 distinguishing desmoids from its histologic mimics, which generally lack this feature [5, 10,
122 11, 18].

123 The differential diagnosis of desmoid tumor vary from benign reactive lesions such as a
124 hyperproliferative scar, to a more sinister fibrosarcoma [11]

125 The treatment of this patient was surgical. Radiotherapy was not directly demanded. The
126 management of breast fibromatosis includes surgical excision with clear margins, Systemic
127 therapy and radiation therapy [6, 11]. Surgical therapy remains the cornerstone of desmoid

tumor management. The mastectomy can be indicated for vast fibromatosis or for too large recurrences. In case of parietal invasion, the surgery can be extremely decaying going as far as taking away the breast, the pectoral muscles, the thoracic wall, and the parietal pleura [15]. Obtaining microscopically negative margins (R0 resection) is the preferred objective in oncologic surgery, including resection of desmoid tumors. Not all positive margins were recurrent; the true impact of surgical margin negativity as well as the role of adjuvant therapies i.e., radiation, antiinflammatory drugs, antiestrogen agents, and cytotoxic chemotherapy in preventing tumor recurrence remain uncertain [11]. Recurrent disease has been observed in negative margins, some authors proposed the algorithm of a more conservative wait-and-see approach in a less critical site with an asymptomatic lesion [19]. If the lesion progresses by RECIST (response evaluation criteria in solid tumor) criteria, then treatment is needed. Desmoid tumors respond to systemic medical therapies, from anti-inflammatory drugs to standard chemotherapy agents. Liposomal doxorubicin, doxorubicin-based regimens, methotrexate in combination with vincristine and vinorelbine have demonstrated notable responses in this disease. Molecular-targeted agents, antiinflammatory drugs, hormonal agents, and interferons have all shown various degrees of activity in DT. However, the mechanisms through which these compounds conduct their antitumor activity are not completely understood [20].

Radiotherapy can be used in patients who have recurred after initial surgery and as primary treatment in those patients who are medically inoperable or those to whom resection presents unacceptable morbidity [21]. Some studies have shown a beneficial effect of the radiotherapy with 60% of decreasing cases non-accessible to the surgery. Therefore, the radiotherapy could be curative in recurrence cases when the surgery is impracticable [22, 23]. The use of radiotherapy must be considered carefully in light of the potential long-term side effects of radiotherapy including the risk of secondary malignancy, particularly in younger patients. Treatment decisions are individualized to each patient depending on a range of patient and tumor characteristics [24].

Conclusion

Breast fibromatosis is a benign affection which can be mistaken clinically, radiologically, cytologically as a malignant lesion. The bilateral form is extremely rare. A well-coordinated, multidisciplinary approach involving the input of surgical and nonsurgical specialists is needed to develop an individualized treatment strategy appropriate for each specific patient.

Conflict of interest

There is no conflict of interest.

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