Fibromatosis, a benign breast disease mimicking carcinoma.
A case report

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A R T I C L E   I N F O

Article history:
Received 31 July 2017
Received in revised form 8 November 2017
Accepted 9 November 2017
Available online 11 November 2017

Keywords:
Breast disease
Benign breast lesion mimicking cancer
Benign breast disease
Case report
Fibromatosis

A B S T R A C T

INTRODUCTION: Fibromatosis is an uncommon breast lesion that can mimic breast carcinoma in its clinical presentation.

CASE SUMMARY: We present a clinical case in which a diagnosis and treatment dilemma existed, in terms of ultrasound findings that were not clear and suspicious, as well as results of fine needle aspiration cytology. Our findings are compared with previous published cases. Also, literature review regarding fibromatosis presentation and diagnosis has been discussed, as well as treatment options.

CONCLUSION: Management of breast fibromatosis remains controversial because of the low incidence and further efforts needed to establish evidence-based treatment guidelines.

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1. Introduction

Fibromatosis, a locally aggressive but non-metastasizing neoplastic proliferation of fibroblastic cells. It is commonly encountered in the abdominal wall and extra-abdominal sites but it rarely occurs in the breast [1]. They are distinctive lesions best defined as a group of non-metastasizing fibrous tumors which tend to invade locally and recur after surgical excision. Fibromatosis arising from the breast, also referred to as desmoid tumor, is a rare benign entity, accounting for only 0.2% of all breast tumors, and 0.3% of all solid tumors [2].

Our case of recurrent fibromatosis in a young lady presented at the European institute of oncology in Milan, a well know reference center of breast cancer in Europe.

This work has been reported in line with the SCARE criteria [21].

2. Case summary

31 years old lady presented to the breast surgery clinic with dimpling in the left breast, with no history of nipple discharge. The patient had regular menses, she denied any genetic familial history or family history of breast cancer or any other breast conditions, any previous breast procedures; including Aesthetic ones. No drug history including hormonal therapy. The patient’s psychological status was optimum with no past history. No smoking history

On physical examination, a nodular density around 2 cm in the LOQ (lower outer quadrant) of the Lt (left) breast was noted, which was firm on palpation with irregular borders, no other lesions were noted in the same breast or in the other side. Bilateral axillary examination was unremarkable.

Ultrasound examination of the Lt breast showed a solid hypo-echoic irregular vascularized nodule around 20 mm in size at the LOQ, with suspicious characteristic. Axillary examination of the same side documented the presence of ovalar 9 mm lymph node with central hyperecogenesity and minimal cortical thickening. The report was staged as BIRADS 4C with a high suspicion of malignancy. MRI was done and showed background enhancement already evident in early phases that limits the diagnostic sensitivity.

In the Lt breast at the LOQ (upper outer quadrant), a coarse distortion is noted about 23 × 10 mm which was suspicious. Noted as well at the OCQ (outer central quadrant) of the same breast a distortion of 15 mm with a doubt significance. At the Rt (right) breast, in the LOQ (lower outer quadrant), another suspicious area of distortion is documented, which was difficult to measure [Fig. 1].

FNA aspiration was done for the lesions noted at the Lt breast which shows negativity for malignant cells in the LOQ, staged C2 according to the European Guidelines – 1997. The same results were noted also at the LOQ lesion of the Rt breast (the lesion was also suspected in MRI). The FNA for the lesion in the OCQ of the Lt breast was inadequate for diagnosis, as well as those at the Lt axilla.
Giving the discrepancy between the suspicious findings in the US for the lesion noted in the LOQ of the Lt breast and the result of the FNA which was C2, a Core biopsy was performed, which revealed a proliferation of spindle cells of a fibroblastic type with slight atypia in a background of dense connective tissue. Embedded mammary ducts in apocrine metaplasia without atypia and microcalcifications. The lesion tended to be a mesenchymal one with an uncertain potential of malignancy, suggesting the need for surgical excision.

The patient after that underwent a surgical excision by a senior breast surgeon of the suspicious lesion which in the LOQ of the left breast. Intraoperative frozen section (Macroscopic Examination) for the lesion revealed a 2 cm nodule without signs of malignancy, compatible with a benign mesenchymal lesion. A radioguided excision of the other non palpable lesion in COQ of the same breast was done and sent for definitive histological exam. The patient tolerated the procedure well in day surgery and was discharged home in good condition with No post operative events.

The final histopathology for the LOQ lesion revealed; Proliferation of Spindle cells with a slight focal atypia in a background of dense connective tissue, [Fig. 2], immunophenotyping was positive for ACTIN of the smooth muscles, and focally for B-CATENIN, negative for CYTOKERATIN, DESMIN, CD34, CD99 & BCL-2.

Fibrocytic disease with non proliferative type with stromal fibrosis and apocrine metaplasia were the findings for the COQ lesion. So, the final diagnosis was Fibromatosis, which is a benign condition.

Then, the patient was reassured of the benignity of the lesion and was scheduled for close follow up.

After 3 months of follow up the patient presented for the reappearance of dimpling in the lower part of her Lt breast at the position of the previously excised lesion. And the patient reported episodes of pain as well at the same site.

Physical Examination showed a hard nodule, moderately mobile, with an overlying dimpling of the skin [Figs. 3 and 4]. Repeated follow up ultrasound of the breast showed a hypoechoic area with dyshaemogenic structure with a maximum diameter of 23 mm, the same characteristics of the previous lesion in the LOQ which was excised for a Fibromatosis [Fig. 5]. At the site of the excised lesion, there was another hypoechogetic area, with an irregular margin with the same characteristics of the previous lesion and a maximum diameter of 14 mm. A third lesion morphologically identical of that at the LOQ of the Rt Breast (previously documented in the MRI) about 17 mm which resulted previously to be C2 in the FNA. Bilateral Fibrocytic lesions. Axillary regions were clear. The stage of the US was BIRADS 3, a probably benign finding.

The patient was referred after that to a Sarcoma specialist for further evaluation, and repeated MRI was done, which showed in comparison with the previous one, that at the Rt breast two newly developed lesions about 5 mm & 5.5 mm in size respectively, with irregular margins and early and progressive enhancement which was suspicious (considering at that time the menstrual cycle of the patient). The same findings were found in a 5 mm lesion noted at the Lt UQO of the other breast.

Again in the Rt breast at the LOQ, a distorted lesion around 20 mm with efigarly and persistent enhancement with the same characteristics of the previously excised fibromatosis. Meanwhile, a less distortion was evident at the site of the excised lesion at the Lt COQ of the left breast, while persisted particularly at the late signaling at a 24 mm lesion noted at the Lt LOQ adjacent to the surgical wound. The report of MRI was staged as BIRADS 3 with a benign probability.

Comparing the characteristics of the lesions in MRI with the previous histological exam and literature review, the diagnosis was made that of a recurrent Fibromatosis.
After that, a revaluation for the receptors status of ER, PR and Her-2 of the previously excised fibromatosis was made and resulted to be negative.

Giving the above all data and after the discussion with the sarcoma specialist and the reassurance of benignity noted in the follow up US, the decision was to follow up the patient in a 6 months period with a new MRI.

The patient was explained in details about her condition and the exclusion of malignancy in her case. She understood that her condition is a special kind of a disease that has a tendency to recur and she understands the need of a close follow up. The patient was happy to share her information in this case report for the sake of education and for other patients.

3. Discussion

Among the benign mesenchymal lesions of the breast masquerading as clinical carcinoma, fibroblastic and fibrous proliferations such as fibromatosis are fairly rare. Their inflammatory, reactive, or truly neoplastic nature is often difficult to determine histologically, and the cytological features of fine needle aspiration of these lesions may also be misleading [4]. As it was seen in our case with the FNA results that were inconclusive [5].

The disease usually occurs in the trunk and upper extremities, but its occurrence in the breast is rare [3].
Aggressive fibromatosis of the breast is rare, in some cases the pectoral muscle and fibroaponeurotic fascia are involved and are presumed to be the site of origin. In others, the tumor is clearly discrete from pectoral muscle. In all described cases, presentation has mimicked breast carcinoma clinically and radiologically [6].

Their specific genetic alterations have not been elucidated. However, their occasional occurrence in patients with familial adenomatous polyposis (FAP) and their morphologic identification with other deep fibromatoses (desmoid tumors), suggest that alterations of the APC/-catenin pathway might be involved in the pathogenesis of sporadic and FAP-associated breast fibromatosis [7].

Breast fibromatosis is a rare benign lesion that should be included in the differential diagnosis of malignant local tumor recurrence after lumpectomy in cases with no good correlation between clinical and imaging findings as it was also presented in a case report of early tumor recurrence after conservative cancer surgery [8].

It can occur at any age and virtually in any anatomical location but it is more commonly seen in the abdominal wall as a desmoid tumor, in the proximal limb girdles, or in the muscles and fascia of the shoulders of adolescents and young adults. Fibromatosis has a tendency to recur locally after surgical excision [5].

Presentation of fibromatosis in the breast is usually associated with an area of skin depression, dimpling or retraction and usually no previous surgeries are done to the breast [4]. Clinically, fibromatosis of the breast are movable, firm masses. Lesions close to the nipple may present as well with nipple retraction [9,10].

The etiology of mammary fibromatosis is still a subject of debate. An association with Gardner’s syndrome, silicone breast implants, or surgical trauma has been reported [16]. Trauma, particularly resulting from surgical intervention, has been emphasized as an etiological factor [11,12]. Association with breast Implant breast fibromatosis can also develop in association with the capsule around a breast implant, although only a few cases of fibromatosis associated with breast implants have been reported thus far. As the demand for breast augmentation had increased, it is important to understand the diseases associated with breast implants. [9].

Although its occurrence is more common in young and fertile women, desmoid tumors also have been reported in men [4]. In male patients, the growth rate has been reported as low. Estrogen dominance, as is the case during pregnancy, is considered to be a significant predisposition for developing those tumors [10].

Ultrasound finding are characterized usually with what we have found in our case. Yamaguchi.H et al., had also noted in a review of the Japanese literature, describing the characteristic ultrasonographic findings of breast fibromatosis which have been reported to include hypoechoic mass with an irregular shape and un even interior texture [3]. Fine needle aspiration cytology is often unhelpful but it may provide a pre-operative diagnosis and allow treatment planning [5]. True Cut Core Biopsy usually aids in establishing a diagnosis and guidance for treatment planning. It usually shows fragments of cellular fibrous tissue composed of uniform spindle cells [5].

C. Yiangou et al., mentioned that frozen sections are unreliable in making the correct diagnosis and in assessing the margins and should be avoided [13]. In spite of that, a wide local excision was done in our case using Intraoperative frozen section which revealed a mesenchymal lesion with benign features.

Wide local Excision is the treatment of choice for these lesions giving the impression of their benignity although mastectomy was performed in the old days because of their presentation mimicking carcinoma [1,4]. Nowadays wide local excision is usually done, and mastectomy may be required for extensive or locally recurrent disease. When “tumor” is adherent to fascia, muscle or skin, excision should be extended to include the affected area [5].

Histologically the lesion is composed of spindle cells arranged in interlacing bundles and fascicles. Cellularity varies from uniformly cellular lesion with minimal collagenization to densely hyalinized collagenous lesions with keloid like appearance. The lesions have irregular margin, with spindle cells infiltrating the parenchymal elements. The entrapped ductal and lobular units may show mild cellular atypia; however, the spindle cells are usually uniform with a low or absent mitotic index [6,9,14].

An important diagnostic tool is B-catenin nuclear staining, which was done also in our case. Noted that High-grade staining is restricted to a small number of mesenchymal tumors. Among these lesions, the only one that has an important morphologic similarity with Desmoid tumors is Solitary fibrous tumor. A simple way to differentiate it from breast fibromatosis is with CD34 immunohistochemical staining [15].

Differential diagnosis of spindle cell lesions of breast is challenging, which can vary between reactive, benign and malignant. In addition, clinical, radiological, and immunohistochemical similarities can be seen in these lesions [16]. Pathologically, fibromatosis must be differentiated from fibrous histiocytoma, fibrosarcoma, cystosarcoma phylloides, spindle-cell (metaplastic) carcinoma and benign processes such as radial scars, nodular fasciitis and post-
irradiation fibrosis [13]. Its distinction from fibrosarcoma, with the absence of nuclear pleomorphism, high mitotic index, abnormal mitotic figures, extreme cellularity, necrosis and vascular invasion generally serve to distinguish fibromatosis from fibrosarcoma [1]. Spindle cell carcinoma must be excluded by looking for cohesive foci and ductal carcinoma in situ, and by performing immunohistochecmistry with a panel of antibodies to CKs. The opinion of a specialist soft tissue pathologist is useful in such cases, and our case was managed collaboratively with a sarcoma specialist. Typically, the mitotic rate does not exceed 1 per 10 HPF, and usually no mitoses can be found. Another neoplasm included in the differential diagnosis is fibrous histiocytoma. Although mammary fibromatosis may have storiform areas, this is rarely a prominent pattern. Epithelioid, histiocytic, and multinucleated cells often found in fibrous histiocytoma are not a feature of fibromatosis [2]. Scars from healed fat necrosis, remote trauma, and surgery must be distinguished from fibromatosis. Calcifications are more likely to be associated with fat necrosis but they can occur in fibromatosis. Foreign body granulomas, sometimes with partially absorbed suture material, are an indication of prior surgery. If the patient has recurrent fibromatosis, reparative changes caused by an earlier operation may mingle with recurrent tumor, further complicating the diagnosis. Lymphoid infiltrates that commonly occur in fibromatosis should not lead to the erroneous diagnosis of an inflammatory condition such as nodular fasciitis. The inflammatory component of fibromatosis is typically limited to isolated separate lymphoid aggregates at the periphery of the lesion. In fasciitis, the inflammatory cells are dispersed more diffusely at the periphery and within the lesion although localized areas of inflammation also occur. “Myoid” and multinucleated cells characteristically found in nodular fasciitis are not a feature of fibromatosis [2].

The simplified approach is to evaluate the spindle cells and the accompanying epithelial cells. In the biphasic lesions with predominance of spindle cells with benign epithelial component, fibroepithelial lesions including fibroadenomas and phyllodes tumors are the most common, followed by pseudoangiomatosus stromal hyperplasia, hamartoma and adenomyoepithelium. For biphasic lesions with predominance of spindle cells with malignant epithelial component, the biphasic metaplastic carcinoma is likely. For monophasic lesions with pure pleomorphic spindle cell only, the monophasic metaplastic carcinoma is more common than the rare primary sarcomas like malignant fibrous histiocytoma, angiosarcoma, and other high grade sarcomas. In monophasic lesions with pure bland spindle cells only, the possible lesions include fibromatosis, fibrosarcoma like metaplastic carcinoma and other unusual conditions like dermatofibrosarcoma protuberance [14].

An important point to mention is the high rate of recurrence of this tumor. C. Yangou et al., highlights the frequent problem of multiple recurrences of this disease, which are normally treated with wider excision, and describes a non-operative approach as a treatment option in recurrent fibromatosis, giving an example of a reported case of recurrent fibromatosis for 3 times with a history of multiple previous breast biopsies [13]. He also emphasized the importance of follow up in the short term and in the long term interval as recurrence has been occurred shortly within one year and up to 11 years post excision. Others has suggested that recurrence usually recurs within 3 years post excision [10]. And this goes with our approach of follow up within 6 months.

There is also evidence to suggest that radiation reduces the volume of gross tumor in extramammary fibromatosis and may be effective in controlling gross disease. Radiation added to those patients with positive margins following resection has also been shown to significantly reduce the rate of recurrence [9]. Erguvan-Dogan B. et al., reported that postoperative radiation therapy can improve the 10-year recurrence-free survival rate [17]. Tumor regression has been reported in inoperable fibromatosis with conventional low-dose chemotherapy [17].

Some studies have reported stabilization or regression with antiestrogen and combination of antiestrogen/nonsteroidal anti-inflammatory drug (NSAID) regimens in sporadic extra-abdominal fibromatosis [18].

It is unclear whether antiestrogen therapy will show a benefit in fibromatosis of the breast, given the typical lack of ER and PR positivity in these lesions [19] which was clear as well in our case.

4. Conclusion

Fibromatosis is an infiltrating histologically low grade spindle cell proliferation, composed of fibroblastic cells with variable amount of collagen. It is characterized by locally aggressive but not metastasizing behavior and high recurrence rate. Although it has been described in many site, relatively few instances of fibromatosis originating in the breast have been reported [20].

With the advent of more accurate imaging methods, together with proper histopathologic interpretation and the judicial use of ancillary methods like immunohistochemistry, most of the entities making up this spindle cell lesion can be identified with certainty, facilitating treatment planning [14].

Management of breast fibromatosis remains controversial because of the low incidence and, in consequence, the limited data. The treatment of choice for breast fibromatosis; is primary surgical excision with clean margins. Tumor edges frequently microscopically extend further than noticed macroscopically; therefore, wide, clean margins become a goal to reduce the recurrence rate. Recurrence seems not to be related to age, sex, tumor size, or location. Conservative therapies (radiotherapy, chemotherapy, and hormonal therapy) should be taken into account when important neurovascular structures are involved or where there is a poor clinical status of patient. A “plateau phase” with growth rate depression or spontaneous regression had been noticed in some desmoid tumors; this could be confused with a false benefit from conservative treatment options, and the question if the cosmetic or oncoplastic surgical approach could be applied given the high rate of recurrence. Although treatment should be individualized based on the patient’s status, location, and extent of these tumors, there is a lack of accurate evidence-based treatment options for patients with breast fibromatosis. And obviously the lack of consensus guidelines with regard to treatment and management of Fibromatosis. Subsequent studies for breast fibromatosis should be performed for a better understanding of its etiology and for the establishment of the most effective diagnostic and treatment choices for individualized cases [18].

Conflict of interest

The authors declare no conflict of interest and no source of funding.

Consent

The consent was obtained for publishing these data.

Funding

No source of funding.

Ethical approval

No ethical approval was needed from the institute.
CASE REPORT – OPEN ACCESS

A. Ashoor et al. / International Journal of Surgery Case Reports 41 (2017) 392–397

Author contribution

Arwa Ashoor: Study design, data collection, literature review, writing the paper.

Simonetta Monti: Study design, data collection, manuscript review, literature review.

Modestino Pezzella: Collection of images and slides, data collection, literature review.

Guarantor

Arwa Ashoor; Simonetta Monti; Modestino Pezzella.

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