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Case Report

Breast Sarcoma: A Case Report

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Abstract

Objective: Breast sarcoma is a rare but serious form of cancer and the surgery represents the only potentially curative therapy. Due to the low incidence of the disease, not enough studies have been conducted on its clinic-pathological characteristics so to define an adequate and consolidated therapeutic approach. The differential diagnosis in cases of primary soft tissue sarcomas of the breast is important for treatment as well as for prognosis.

The objective of this paper is to report a case of breast sarcoma to highlight the importance of differential diagnosis for an early and targeted intervention.

Methods: We report a case of a 47 year-old woman with breast sarcoma previously diagnosed as a fibroadenoma on the left breast.

Results: A core biopsy was performed, the patient was diagnosed with stromal sarcoma with features of fibroblastic differentiation of the left breast. Despite the poor prognosis, the patient is free from disease and her follow-up is ongoing.

Conclusion: This case report underlines the importance of differential diagnosis for early and targeted intervention and the added value that the Breast Unit offers in terms of multidisciplinary approach.

Keywords: Breast Sarcoma; Fibroadenoma; Mastectomy

Introduction

Breast-onset sarcomas are a histologically heterogeneous group of neoplasms. These are very rare tumours accounting for less than 1% of breast cancers and 5% of soft tissue sarcomas [1,2]. The annual incidence is about 4.5 cases/million. Breast sarcomas can develop de novo (primary), or arise on tissue previously radio-treated for another neoplasm and is defined as secondary radiation-induced secondary breast sarcoma. The biological differences from other primary breast tumours necessitate a corresponding unlike approach to diagnostic and management strategies. These tumours present specific histologic and cytogenetic characteristics, clinical implications, and development dissimilar from breast carcinomas [3]. They exhibits accelerated growth, more aggressive behaviour, poorer response to adjuvant treatment, and higher tendency to both local recurrence and systemic metastasis. Primary breast sarcoma can be subdivided into three distinct subgroups: malignant phyllodes tumours, sarcomas arising in the post-irradiation breast and primary breast sarcomas. Both phylloides tumours and primary breast sarcoma may be easily confused with fibroadenoma on cytological analysis, and the consequences of being falsely reassured by negative cytology are potentially serious. Clinically it can be difficult to distinguish from fibroadenoma because it is a

mobile mass without skin thickening, nipple discharge or palpable auxiliary lymph nodes. Ultrasound, mammography and MRI have a low sensitivity, in order to establish the sarcomatous nature of a breast neoformation being, at least in the initial phase, very similar to fibroadenomas. The definitive diagnosis is offered only by core or excisional biopsy with histological examination by inclusion.

In the case of breast sarcomas, radiotherapy or chemotherapy are not indicated, due to the low sensitivity of the sarcomatous tumour cells to these treatments. The therapeutic "gold standard" is constituted by surgical therapy even if it is not standardized and is evaluated case by case, as well as its possible association with chemotherapy and radiotherapy. The histologic results may be the leading factor for the management of these tumours, in cases of high KI in which chemotherapy may benefit the treatment of the tumour. Some studies on radiotherapy and chemotherapy report effect on local recurrences and survival respectively [4,5]. Overall, the prognosis of breast sarcoma is poor. Tumour grade and size of the lesion impact on prognosis; early diagnosis and treatment influence the survival. Unfortunately, due to the rarity, there is still a lack of knowledge; there is no evidence of data capable of fully guiding clinical decisions. For the reason it is necessary that studies be conducted starting from retrospective ones and the broadest collaborations developed between researchers to be able to have valuable information sources to improve treatment strategies for patients. Here we present a case of a 47 year-old woman, who had non-family history of breast cancer, neither history of breast trauma, nor previous radiation exposure, attended to our hospital after noticing an increase in size of a mass previously diagnosed as a fibroadenoma on the left breast.

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A 47 year-old woman attended to the Breast Unit of the Fatebenefratelli Hospital in Rome after noticing a rapidly growing mass on the left breast, previously diagnosed as fibroadenoma. The mammographic finding showed a single radiopaque nodule with circumscribed margins without microcalcifications (Figure1). The echography showed a nodular formation with a diameter of 10 cm, occupying left breast with substantially regular margins, ultrasound heterogeneous echo structure due to the presence of a fluid component similar corpuscular collection (Figure 2). The MRI imaging confirmed an oval and complex mass with irregular margins, a hyperintense signal T2-weighted imaging probably related to central necrosis. Contrast enhancement T1- weighted looks heterogeneous after initial rapid wash in (Figure 3 a,b). CT total body performed to evaluate the staging of disease and did not reveal the presence of secondary lesions (Figure 4 a,b). We decided to perform a core biopsy, from which morphological picture deponent for spindle cell mesenchymal tumor. The treatment for breast sarcoma was planned by the multidisciplinary team. Surgery consisted in nipple sparing mastectomy with skin expander implant and Tiloop mesh. A lozenge incision was made to include the nipple-areola complex and the previous surgical scar; the nipple was de- epithelialized, removed and placed in physiological solution. The nipple-areola complex is positioned in the new site after de-epithelialization of the new site. Sentinel lymph node biopsy and axillary dissection were not indicated due to the absence of evident clinically/radiologically lymph node involvement. Gross examination revealed a nodular tumor of 9 cm (Figure 5). At one pole of the neoplasm, there is a portion of breast tissue measuring 9x3x3 cm. The neoplasm appeared well defined, circumscribed, and consisted of fleshy, firm, pale tan tissue with large area of haemorrhage and necrosis (over 50% of the neoplasm) and areas with a myxoid appearance. Microscopic examination showed interlacing bundles of fusiform cells, with fibroblastic appearance, mixed with collagen, in a storiform arrangement and surrounded by many blood vessels. Myxoid areas were present. Extensive areas of necrosis and haemorrhage were present. Expansive and focally infiltrative growth was present with minimal sarcomatous infiltration in the surrounding breast tissue. Excision margins were negative (distance: >1 cm).The histopathological diagnosis was stromal sarcoma with features of fibroblastic differentiation (G2 sec FNCLCC, score 5). Mitotic Index: 8-9 for 10 HPF – Ki 67%: 25 %. In relation to the histological characteristics and the size of the lesion, the patient was subsequently subjected to chemotherapy and treated with Epirubicin: 60 mg/m2; days 1 and 2, Holoxan 3000 mg/m²; 1 -2 -3, Uromitexan 3000 mg/m²; day 1 – 2 – 3, q 21. Radiotherapy treatment was not taken into consideration. The high rate of local recurrence after surgery (up to 33% of operated patients) would justify the role of adjuvant radiotherapy. However, the available studies report conflicting results with satisfactory loco-regional recurrence rates, but uncertain impact on survival [6]. The patient and her family members were followed during the diagnostic and therapeutic process with interviews and brief psychotherapy with the aim of promoting understanding of therapeutic needs, improving adherence to treatment and maintaining, as far as possible, an acceptable quality of life. The phases of the intervention included: a) interviews before surgery aimed at containing preoperative anxieties and anxiety: individual; group of three/four women; b)administration of a battery of tests that explore resilience and tolerance to uncertainty; c) during the chemo and/or radiotherapy process, the psychologist remains available for individual, couple or family interviews at the request of the woman and/or health professionals; d) at the end of the medical therapies, group psychotherapy is planned for the women and their families, neuropsychological evaluation.

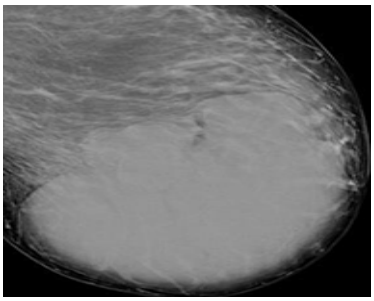


Figure 1: Mammographic profile shows a single radiopaque nodule with circumscribed margins.

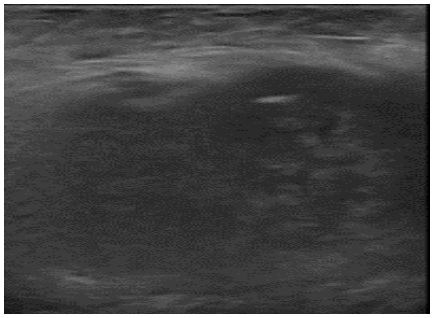
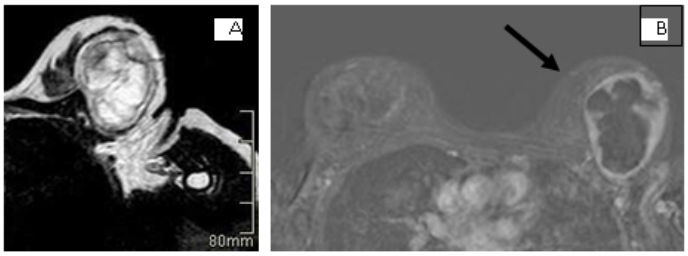


Figure 2: Ecographic profile shows nodular formation in left breast with a diameter of 10cm.



A B

Figure 3: A) MRI imaging confirms an oval and complex mass with regular margins, a hyperintense signal T2-weighted imaging probably related to central necrosis. B) Contrast enhancement T1-weighted looks heterogeneous after initial rapid wash.

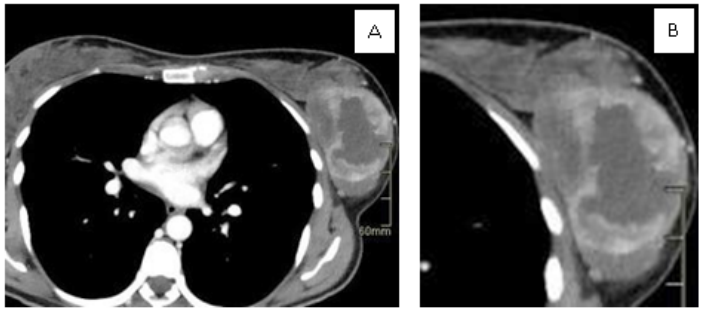


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Figure 4: A) CT reveals the absence of secondary lesions. B) Absence of certain plane of cleavage with the underlying chest wall.

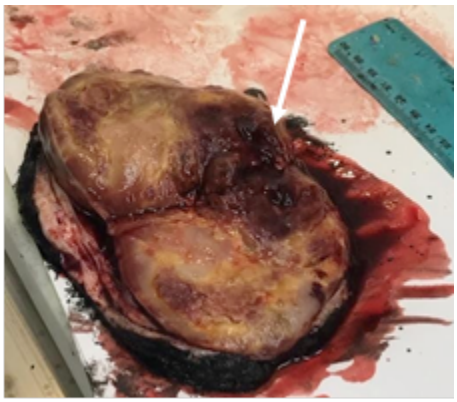

Figure 5: Sarcoma of the left breast with necrotic central portion.

Discussion

Primary breast sarcoma is an extremely rare but aggressive neoplasm with few hundred cases reported in literature until today [1]. Due to the low incidence of the diseneoplasmase, not enough studies have been conducted on its clinic-pathological characteristics so to define an adequate and consolidated therapeutic approach. The exact aetiology is unknown and although women who receive radiotherapy have an increased risk of presenting secondary breast sarcoma, the absolute incidence is small. These tumours present a high possibility of recurrence

and have poor prognosis. They tend to spread hematogenously involving lungs, bone marrow and liver [7]. Although imaging tools are nonspecific they may be indicative of sarcoma but core biopsy is required for diagnosis [8]. With regard to imaging, either breast mammography, ultrasound or MRI can be useful. In mammography it appears as a smooth, well-defined lesion, occasionally with a perilesional halo. Ultrasonography more often shows an abnormality than mammography: a hypoechoic, oval, lobular mass. MRI appears to be more useful in delineating the disease but still has disadvantages similar to other modalities. Adjuvant chemotherapy and radiotherapy should be considered in high risk cases when the size is >5 [9]. The role of chemotherapy is disagreeing and might be proposed in cases with worst prognosis [10]. A partial effect has been observed for some combination therapies but without significant improvements [11]. Complete surgery represents the best treatment of the breast sarcoma, being the only curative approach and at the same time the most important prognostic factor for long-term survival. The local therapy should be aggressive, with radical mastectomy and resection of the axillary lymph nodes. Since lymph node metastases occur in only 5% of cases, axillary lymph node resection is recommended only when lymph node positivity is confirmed [12]. The differential diagnosis in cases of primary soft tissue sarcomas of the breast is important for treatment as well as for prognosis [13,14]. It includes phylloides tumours, metaplastic carcinoma, triple-negative carcinomas, juvenile fibroadenoma and squamous cell carcinoma of the periprosthetic capsular tissue of the breast, a rare entity that affects the inner skin area where the breast implant is placed [11]. In the present case, a 47-year-old woman showed a granular formation in the external quadrants of the left breast, already present two years earlier, during which the mass increased from 1 to 10 mm, diagnosed both clinically and with instrumental examinations as fibroadenoma in the absence of histological typing. The comparison with the previous controls and the exponential increase in size made it necessary to evaluate the lesion with needle biopsy. Histological typing showed proliferation of spindle cells (B4) and subsequently histopathological confirmation of stromal sarcoma with features of fibroblastic differentiation. The patient was also able to benefit from the psychological support present in the Breast Unit. The psychological intervention in the multidisciplinary team of the Breast Unit is important in order to modulate the intervention through sharing with the team. The transversal function to several operating units allows the psychologist to link up with the work of different specialists and the patient to feel accompanied in a highly personalized path. In our case report, the patient and her family were followed up during the diagnostic and therapeutic process with interviews and brief psychotherapy with the aim of promoting understanding of therapeutic needs, improving adherence to treatment and maintaining, as far as possible, an acceptable quality of life. The psychological journey shared with the treating team promoted the emotional and cognitive process of "re-signification" of internal and interpersonal experience and the possibility of reconstructing beliefs about oneself and the world. This process led to a better adaptation to the traumatic event and a reduction of psychological distress over time. Despite the unfavourable prognosis and the size of the tumour mass, the surgery and chemotherapy, after 5 years, resulted in the patient being free of disease and her follow-up is ongoing.

Conclusion

Breast sarcoma is a rare but serious form of cancer and the surgery represents the only potentially curative therapy. Tumour size is an important prognostic factor, with tumour less than 5 cm in diameter, the overall survival rate is better. The early and correct diagnosis is extremely relevant because it's severe course and poor prognosis. The biopsy is necessary to have a definitive diagnosis because of the imaging findings are not pathognomonic. This case report underlines the importance of differential diagnosis for early and targeted intervention. Besides, our case highlights the added value that the Breast Unit offers in terms of multidisciplinary approach, which lets face the problem in its entirety without underestimating the importance of psychological support throughout the course of the disease.

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