Breast sarcoma. A case report and review of literature

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ABSTRACT

INTRODUCTION: Breast sarcomas are rare with an annual incidence of 4.6 cases/1,000,000 women. They can appear as primary forms or secondary to radiation therapy or chronic lymphedema.

PRESENTATION OF CASE: A 41 year old woman attended our hospital after having noticed an increase in the size of her fibroadenoma. The examination revealed a 7 cm retroareolar nodule. Breast sonography described a hypoechoic bilobulated lesion and MRI showed a large size polinodular image, suggesting a Phyllodes tumor. A core needle biopsy was performed with a histological result of low-grade fusiform cells sarcoma on Phyllodes tumor so we proceeded to surgical treatment with a mastectomy.

After two years and a half she noticed a tough nodule over the mastectomy scar, which was resected with a histological result of fusiform cells sarcoma. Considering the diagnosis of recurrence of the disease, surgery was undertaken.

DISCUSSION: Breast sarcoma is a rare but aggressive entity. Core biopsy is the procedure of choice for the diagnosis. Lymphatic spread is uncommon so nodal status in breast sarcoma is less informative. Staging study differs from other breast tumors and chest computed tomography is helpful since lungs are the predominant metastatic sites. The use of radiotherapy or chemotherapy is controversial and will depend on the risk of tumor recurrence.

CONCLUSION: Surgery represents the only potentially curative therapy for breast sarcoma. Tumor size and adequate resection margin are the most important prognostic factors. Approximately 80% of recurrences appear in the first two years.

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1. Introduction

Breast sarcomas, which develop from mesenchymal tissue, are rare and their annual incidence is approximately 4.6 cases/1,000,000 women, representing less than 1% of all breast malignancies. They can appear as primary forms (de novo) or secondary to chronic lymphedema or radiation therapy on the breast or chest wall, with the two forms presenting different features. The primary forms appear histologically as heterogenous subtypes and their mean age of diagnosis is around 40 years. In contrast, the secondary forms typically present later at around 45–50 years and the most common histological subtype is angiosarcoma.

Lymph node metastases are uncommon in breast sarcomas and surgery represents the only potentially curative therapy, always with an adequate resection margin.

They can share some clinical features with breast carcinomas but therapy and prognosis can differ substantially.

We present this case because of the low incidence of breast sarcomas with few cases being reported, especially of primary forms, and the clinical imagery obtained.

2. Presentation of case

A 41 year old woman, who had been diagnosed previously with a fibroadenoma on the left breast, attended our hospital after having noticed an increase in its size. Her personal history did not feature other important details. The examination revealed a 7 cm retroareolar nodule, well-defined and tough. Breast ultrasound described the previously known fibroadenoma and a hypoechoic bilobulated lesion with thick margins (Fig. 1) and the result of the citology we obtained by fine needle aspiration was connective-adipose tissue. The MRI showed a large size polinodular image, without signs of necrosis or haemorrhage, suggesting a possible Phyllodes tumor (Fig. 2). The images revealed a low diffusion and a rapid contrast-enhancement with a posterior plateau. We decided to perform a core needle biopsy with a histological result of low-grade fusiform cells sarcoma on Phyllodes tumor. As a consequence of...
After two and a half years of close monitoring, she came again to our hospital because of a tough nodule over the mastectomy scar and the breast ultrasound revealed a heterogeneous epidermic nodule of 19 mm (Fig. 4). It was resected and the histological result showed fusiform cell sarcoma. Considering the diagnosis of recurrence of the disease, surgical treatment was performed with an enlargement of tumor margins, resecting the pectoral muscle and removing the prosthesis.

3. Discussion

Breast sarcoma is a rare but aggressive entity. Due to its rarity, there have not been sufficient studies of its clinicopathological features and adequate treatment approach to achieve a consensus in terms of management of the disease, with published articles usually being small retrospective case reports and reviews.

In most cases, the etiology is unknown and although women who receive radiotherapy have an increased risk of presenting secondary breast sarcoma, the absolute incidence is small.

Clinically it appears as a unilateral, well-defined and large mass; it often grows faster than epithelial breast carcinoma. It can be suspected with a physical examination or through imaging tools, but core biopsy is required for diagnosis [1].

With regard to imaging, either breast ultrasound, mammography or MRI can be useful. The feature usually found in the imagery is the presence of a mass with irregular margins with or without calcifications. It appears to be frequently hypoechoic at ultrasound and T2 hyperintense at MRI [2,3].

Lymphatic spread is uncommon in this entity, and dissemination usually occurs haematogenously and the principal organs to be metastasized are lungs, bones and liver. This is why sentinel lymph node biopsy has not been studied in these cases, as it does not seem to offer sufficient benefit to the patient or to prompt a change in clinical management. Consequently, this means that staging studies will differ from other breast tumors due to the fact that nodal status in breast sarcoma is less informative, so in these cases chest computed tomography is helpful for staging since lungs are the predominant metastatic sites.

Surgery is the standard treatment. Tumor size and adequate resection margin are the most important prognostic factors; in fact, an adequate resection margin is the most important determinant of long-term survival. Routine lymphadenectomy does not seem to improve outcomes [4,5].

There is some controversy about the use of radiotherapy or chemotherapy in these patients, so there is no consensus as to the use of adjuvant therapy and it will depend mainly on the risk of tumor recurrence. Taking this into account, the use of radiotherapy
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