

Carcinosarcoma of the Breast: A Case Report and Review of The Literature

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Citation: Mohamed BS, Lamiaa D, Miriem M, Mustapha B, Mohamed E, et al. (2021) Carcinosarcoma of the Breast: A Case Report and Review of The Literature. Annal Cas Rep Rev: ACRR-278.

Received Date: 26 August, 2021; **Accepted Date:** 31 August, 2021; **Published Date:** 06 September, 2021

Summary

Carcinosarcomas of the breast are a rare entity, representing less than 1% of invasive breast carcinomas. They are defined by a double tumor proliferation associating classical carcinomatous foci and areas of mesenchymal differentiation. They belong to the heterogeneous group of metaplastic carcinomas. We report a case of carcinosarcoma of the breast in a 44-year-old married mother of two children, followed for bronchial dilatation, treated by eight cycles of neoadjuvant chemotherapy followed by radical surgery. The patient is in complete remission after 12 months. The aim of our work is to report the clinical, anatomopathological, therapeutic and prognostic characteristics of this entity.

Keywords: Carcinosarcoma, diagnostic difficulty, treatment, prognosis.

Introduction

Carcinosarcoma of the breast, often referred to as metaplastic carcinoma of the breast, is a rare malignancy that is composed of two distinct cell lineages: classic carcinomatous and spindle cell sarcomatoid of mesenchymal origin, with no transition zone between the two elements.

As metaplastic carcinomas of the breast do not generally express estrogen or progesterone receptors and do not overexpress the HER2/neu oncogene, these tumors tend to be more aggressive as a result of this "triple negative" phenotype. The epidermal growth factor receptor protein HER-1/EGFR is expressed in the majority of metaplastic carcinomas and therefore may potentially serve as a therapeutic target for EGFR inhibitors such as gefitinib and cetuximab.

Observation

The patient was 44 years old, married with two children, with a pathological history of bronchial dilatation for 10 years under treatment, and no history of breast cancer in her family. The onset of her symptomatology was 7 months before her first consultation with the autopalpation of a left breast nodule rapidly increasing in volume.

The physical examination of the patient on admission to the department found her to be in good general condition.

Examination of the left breast revealed a 4 cm cystic mass in the upper-external quadrant with inflammatory signs opposite, which fistulated to the skin a few days after the first consultation. The lymph node examination showed an axillary adenopathy. The rest of the physical examination was normal.

Mammography showed the presence of a homogeneous opacity with blurred and spiculated contours in the superior-external quadrant of the left breast.

Breast ultrasound revealed a parietal mass at the level of the left QSE that was liquid with fine echogenic content and vascularized wall on Doppler. This mass extends deep into the mammary gland to the contact of the pectoral muscle at 45 mm depth, continuing with a solid-cystic mass at the periphery of the left QSE of 30 x 45 mm, with spiculated wall in places and whose fleshy portion is vascularized on Doppler, with the presence of a centimetric and infra-centimetric left axillary ADP some of which are hypoechoic.

Microbiopsy of the mass showed tumor cells expressing vimentin and CKA1/AE3 and not expressing the vascular markers CD31 and CD34 or PS100 as a melanin marker. Overall, the biopsy suggested an undifferentiated malignancy with an immunohistochemical profile suggestive of a carcinosarcoma.

As the mass was deeply adherent, neoadjuvant chemotherapy was indicated, and our patient had received 8 courses of FEC100, after an extension workup that was negative. The clinical response to chemotherapy was excellent.

A mastectomy with axillary curage was performed. The histological study of the mastectomy specimen showed a fibrosing inflammatory remodeling after breast treatment without residual neoplasm. The lymph node metastases concerned a lymph node invaded by a carcinomatous tumor proliferation without necrosis foci or sarcomatoid component or capsular effraction on a dozen of lymph nodes taken. The immunohistochemical study was performed on the metastatic lymph node as there was no tumor residue in the breast. It showed the absence of hormone receptor expression with a KI67 cell proliferation index estimated at 40%.

Discussion

Breast carcinosarcoma (metaplastic, biphasic metaplastic, metaplastic sarcomatoid carcinoma, sarcomatoid carcinoma) is a rare tumor, it presents 0.08 to 0.2% of all breast cancers [1,2].

It is defined by a double tumor proliferation combining classic carcinomatous foci and areas of mesenchymal differentiation, [2].2010 with cytological and histological features identified on light microscopy images and immunohistochemical tests [2,3].

In his definition of carcinosarcoma, Rosen et al [8] use more restrictive morphological criteria and consider that carcinosarcoma is constituted by the association of a breast carcinoma and a mesenchymal tumor without the presence of transition zones between the two neoplastic proliferations. 2007 Contrary to current literature, the term carcinosarcoma is used for tumors where the transition line between the carcinomatous and sarcomatous components is well respected on all microscopic fields 2013.

The histogenesis of carcinosarcoma has long been a matter of controversy. Several theories have been proposed to explain its development. Kauffman's hypothesis suggests the particular phenotypic transformation of epithelial cells into myoepithelial cells and then into sarcoma; a theory confirmed by the detection of epithelial characters by electron microscopy and immunohistochemistry in mesenchymal cells [13].

The histogenesis of carcinosarcoma is discussed. Some authors [9] suggest the possibility of a dedifferentiation of the carcinomatous component, supported by molecular biology studies that found an identical clonality for both components, confirming that the cells are of myoepithelial origin.

In addition, molecular biology studies showed that the tumor cells of both tumor components had the same translocation at the p53 gene, reinforcing the theory that the tumor cells derive from the same totipotent stem cell 2010. These tumors occur in postmenopausal women over the age of 55. [9, 10].

The age of our patient was similar to the results of the study done at the Orangers maternity hospital in Rabat, where the average age was 44 years.

Clinically, carcinosarcoma is distinguished by its large size with an average between 3 and 4 cm and by its faster growth than in other classical infiltrating carcinomas. [1, 3, 4, 11]. 2007 Our patient's tumor size was large with an aggressive course. Mammography is not very specific; it is most often a poorly limited opacity, sometimes with spiculated contours. Breast ultrasound is more sensitive, showing the heterogeneous character of the lesion, alternating solid and cystic areas, hence the need to evoke the diagnosis of metaplastic carcinoma in front of a breast nodule with a cystic component [12] 2011. The clinico-radiological characteristics of our patient's lesion correspond to those of the literature.

The macroscopic appearance of carcinosarcoma of the breast is characterized by a larger size than that encountered in infiltrating ductal carcinoma. On section, the tumor is firm and solid with the presence of cystic remodeling. [2].

Histologically, carcinosarcoma has a dual epithelial and mesenchymal component, the proportions of which vary from patient to patient: one carcinomatous of ductal type, the other sarcomatous of variable differentiation.

The immunohistochemical study confirms the diagnosis of carcinosarcoma by showing the particular immunophenotype of the mesenchymal component: a frequent negativity of broad spectrum and high molecular weight cytokeratins and a positivity of vimentin. In contrast, markers of myoepithelial differentiation can be expressed [12], namely low molecular weight cytokeratins (CK5/6, CK14) in less than 50% of cases, p63 and SMA.

Carcinosarcoma is an aggressive tumor [13], and rare, which explains the difficulty of establishing the survival rate. The establishment of prognostic criteria remains difficult in carcinosarcoma because the SBR grade is not applied to them and the lymph node invasion is lower for the same size than in the common form. As for the grade of the sarcomatoid component, it does not seem to have a prognostic impact [11]. In addition, the proportion of carcinomatous and sarcomatous components could interfere, but has not been evaluated in the literature. 2007 Some authors cite mitotic activity and high nuclear grade as unfavorable histopronostic factors.

The evolution of carcinosarcoma is marked by local recurrences, including axillary metastases in 26% of cases and a high frequency of visceral metastases, mainly pulmonary, ranging from 41% [14] to 46% [11]. The prognosis of carcinosarcoma is poor with a mean survival of 67% at 5 years [8] and 40% at 10 years [1], compared with 68% and 64% at 10 years for metaplastic carcinoma with matrix production and sarcomatoid carcinoma, respectively.

In this patient, is distant lymph node and liver metastases occurred one year after diagnosis. The evolution of our patient did not show any particularity even if the duration

is not sufficient to make a decision. Carcinosarcoma is known for its marked metastatic potential by lymphatic and hematogenous routes, knowing that lung metastases are more frequent than brain, bone and liver metastases, which requires a careful clinical examination in search of distant signs of call and an extension workup including liver ultrasound and chest X-ray even in the absence of signs of call before any therapeutic attitudes [3].

Treatment is not well codified due to the limited number of cases. Patients are usually treated by mastectomy and axillary lymph node dissection with or without radiotherapy. Furthermore, hormone receptors are rarely expressed by this type of tumor, which limits the contribution of hormone therapy. Given the negative evolution of this tumor, the main therapeutic issue is the type of adjuvant chemotherapy to be given. Most teams treat carcinosarcoma as invasive carcinoma without further indication (IAC); chemotherapies indicated in sarcoma, with reference to what is done in esophageal sarcoma, have also been proposed [14].

The treatment of carcinosarcoma of the breast is the same as for other types of breast cancer, and is based on mastectomy with or without axillary curage. Wargotz and Norris [12] reported a lymph node invasion rate of 26%, which indicates the usefulness of axillary curage. Various chemotherapy protocols including (CMF: Cyclophosphamide, Methotrexate, 5Fluorouracil) or Adriblastine-based chemotherapy, which appears to be more effective [13], and adjuvant radiotherapy have been used. The prognosis remains poor. Metastases are frequent, frequently involving the lung and pleura rather than the liver, skeleton or brain. Local recurrence is also frequent but can be controlled by surgical resection, intensive chemotherapy and radiotherapy, but given the rarity of cases reported in the literature, there is no therapeutic consensus for effective treatment of recurrence. [14,15].

Conclusions

Although carcinosarcoma of the breast is a rare subtype of breast cancer, it is of significant interest because of its clinical and pathologic heterogeneity from typical breast cancer.

Carcinosarcoma represents a single tumor type with a wide morphological spectrum, explaining the difficulties encountered by the pathologist in making the diagnosis of carcinosarcoma. Molecular biology studies could be an additional diagnostic tool by providing information on the histogenesis of these tumors.

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