

Carcinosarcoma of the Breast - Rare Clinical Case

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Abstract

True carcinosarcoma of the breast (BCS), occasionally defined as a subtype of a metaplastic carcinoma, is an extremely rare malignant tumor. It accounts for 0.08% - 0.2% of all newly found breast malignancies. BCS is composed of two distinct components- a carcinomatous and a non-epithelial component of mesenchymal origin, without a transition zone between them. Clinically, BCS has an aggressive progress. We report the case of a 44-year-old woman who presented with a very large tumor of the left breast, which started growing extremely fast after application of a non-traditional treatment. The optimal treatment regimens are not well established due to a limited number of cases. At present the best treatment is a complex one, both local and systemic. The surgical treatment, in view of organ preservation, with adjuvant chemotherapy gives results similar to those in respect to the invasive carcinoma of the breast. More often mastectomy is a treatment of choice. The optimal treatment of the carcinosarcoma has not yet been defined.

Keywords: Breast; Carcinosarcoma; Rare Malignancy; Diagnostics; Treatment

Introduction

True carcinosarcoma of the breast is an extremely rare malignant tumor. Its prevalence ranges between 0.08% - 0.2% of all malignant tumors of the female breast [1,2].

The proper definition of the true carcinosarcoma should include the two different cell components - a carcinomatous and a non-epithelial mesenchymal component, without a distinct transitional zone between these two components [2,3].

There is a controversy over the nature of this rare neoplasm, but the main opinion of most researchers leads us to believe that it arises from myoepithelial cells with a potential biphasic differentiation [4].

The pathological diagnosis of true carcinosarcoma of the breast is very difficult. The true carcinosarcoma should be well differentiated from the metaplastic carcinoma, the tumors with cartilaginous and bone metaplasia, the malignant phylloid tumors and from the different types of sarcoma [2,5]. The main difference of distinct metaplastic carcinoma from carcinosarcoma is whether or not there exists a transitional zone between these two components. This transitional zone is always present when it comes to the different types of metaplastic carcinomas - in the form of fibroplastic, chondrial, bone or osteoplastic [6,7].

The larger part of the true carcinosarcoma of the breast, shows no expression of estrogen and progesterone receptors and HER/2neu with the so- called “triple negative” pattern [8-10].

The treatment approaches are similar to those for other types of breast cancer [11-13]. True carcinosarcomas metastasize by the lymphogeneous and hematogeneous ways. Therefore, axillary examination and dissection is part of their surgical treatment.

The substantial characteristic of true carcinosarcomas are the hematogenic metastases usual in the lung and pleura, less often in the bones, liver and brain [11,14].

The five-year survival rate is 49%, worse than that in other types of cancers.

Case Presentation

We report the case of a 44- year- old female who was admitted with a large tumor occupying the entire left breast. Initially, the tumor started as a small ball. The patient chose non-traditional methods of treatment- she used food supplements, “bioresonance therapy”, and total starvation. During the last 2 months of treatment course, the tumor mass grew rapidly.

The clinical study found a large, lobulated, protruding tumor of the left breast with a size of 30/36 cm, which put this patient in a unique circumstance. The top layer of the skin was a reddish-violet colour, extremely stretched and in parts with the presence of hemorrhage zones. No regional lymph nodes were found (Figure 1a and 1b).



Figure 1a and 1b: Clinical example of a patient with carcinosarcoma.

A tumorotropic scintigraphy with ^{99m}Tc-MIBI was performed, followed by a SPECT-CT, to obtain an objective information for the operability of the tumor and its involvement of the underlying musculature (Figure 2).

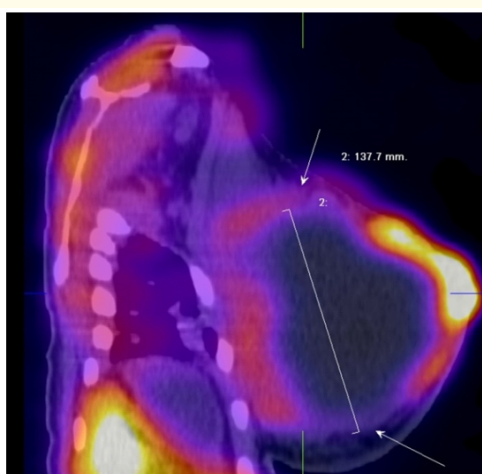


Figure 2: ^{99m}Tc-MIBI SPECT-CT image of the tumor.

The examinations showed the presence of a solid, fleshy formation, which had infiltrated the entire breast parenchyma, with infiltration of the skin and mamilla. There was central necrosis with a diameter of 15.5 cm and numerous suspect lesions of the lungs bilaterally. In addition, due to the long starvation, the patient was in a very low conditions and strongly impaired general condition and severe impairment of hemodynamics.

After the conducted reanimation procedures and hemotransfusions, the patient improved her overall health status and showed an improvement in her hematologic results, on October 19, 2017 the patient was ready to be operated.

Intraoperatively, a large, lobulated tumor formation was found with clinical dimensions of 30/39 cm. This tumor had infiltrated the underlying pectoral muscle (Figure 3 and 4). Based on these finding, we performed a radical mastectomy in the manner of Halsted. The tumor was excised in conjunction with the top skin layer and the partially infiltrated pectoral muscles, up to the point of healthy tissue on a macroscopic level. The operation was characterized by a histologically confirmed radicality. Eleven nonsuspicious lymph nodes were excised from the axilla.

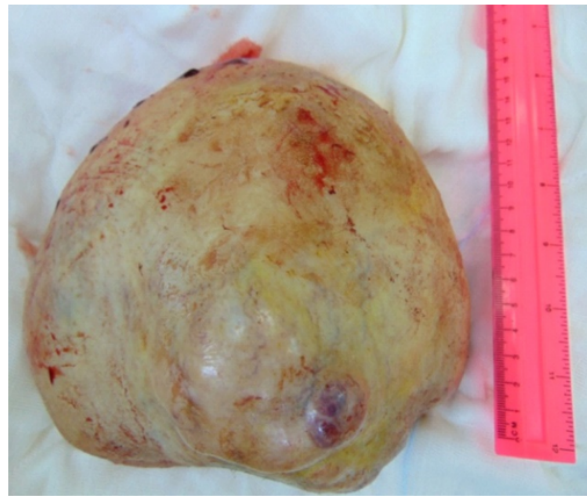


Figure 3: The operatively excised breast with the tumor.

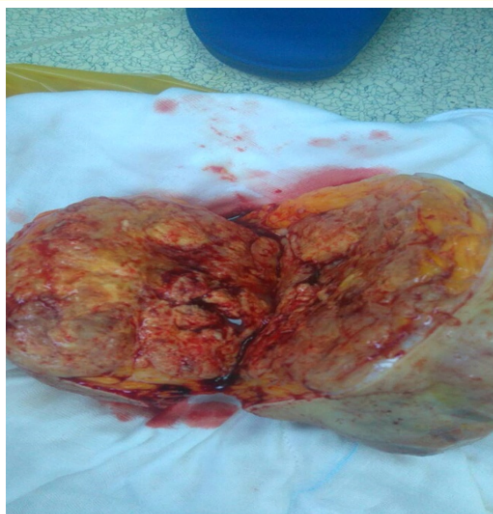


Figure 4: Axial cut of the carcinosarcoma.

The final histopathologic studies revealed carcinosarcoma 32/38/41cm, Grade 3. A residual tumor and metastatic invasion of all 11 examined lymph nodes were not found.

The immunohistochemical findings were as follows: HER-2/neu also (-). Hormone receptor assay stated the tumor to be negative for both ER-(neg.) and PR-neg. Ki 67 expression was 80%. CK (+), EMA (+) Vimentin (+), Desmin 0 (neg.) S 100 (+), HMB 45 -0 (neg.) The patient recovered fast and adjuvant chemotherapy was performed.

Discussion

The special pathological characteristics of the true breast carcinosarcoma are very important for its differentiation from the other types of sarcomas of the breast, such as fibrosarcoma, malignant histiocytoma, osteosarcoma, phylloid tumor and stromal sarcoma, which have different clinical behaviour and treatment response [6].

The clinical features of true breast carcinosarcoma are similar to those of the other invasive types of carcinoma [10-12]. It has often been demonstrated by the presence of local recurrences when an organ preservation operation was performed. Some local recurrences, particularly one with a short interval of appearance, can be more aggressive than primary tumor. Some recurrences can be surgically treated when it is possible, thereby improving survival. Usually distant metastases are primarily found in the lungs, as opposed to the brain, bones, or the liver. These patients have a poor prognosis [14].

The treatment scheme of the true carcinosarcoma is based on the accurate pathologic diagnosis, the main purpose of which is accurate definition. In our daily practice we follow the NCCN guidelines for patients with invasive breast cancer.

For the optimal treatment of this rare malignant tumor, a multidisciplinary treatment approach is needed. In vast majority of described cases, breast conserving surgery or radical mastectomy, followed by adjuvant chemotherapy and radiation therapy, was performed. The treatment scheme requires an oncology team, consisting of a surgeon oncologist, pathologist, chemotherapist, radiotherapist. It is assumed that new treatment opportunities will be developed in regards to target medications against specific biological markers like Gefitinib (Iressa) and Erbitux [11,13,14].

This case is of great scientific interest, due to the rare occurrence of the true carcinosarcoma, which has its own specific clinical course and a bunch of difficulties in regard to its treatment. In this particular case, the pathological and immuno-histochemical data show, that this is a case of a more aggressive subtype of the disease. This case also demonstrates an intensively fast progression of the tumor, which theoretically may be the result of the treatment with non-traditional methods. It is assumed that this "therapy" had had a stimulating effect on the growth of the tumor, which for a short time reached an enormous size. Therefore, this case arises a lot of medical and legal questions, in regards to the diagnosis and the treatment of rare tumors of the breast.

Conclusion

The case, mentioned above, gives us the basis to conclude that the diagnosis and the treatment of the rare tumors of the breast are a serious and a not yet resolved problem. The primary clinicians to meet these patients, to set the course of action and to show them the way of dealing with this problem are the family physicians. The hope of a better outcome in this case is the early diagnosis in a specialised oncological center and the development of targeting medications, such as Gefitinib (ZD 1839) and Cetuximab (Erbitux).

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