Journal of Cancer and Tumor International

12(4): 30-34, 2022; Article no.JCTI.89820
ISSN: 2454-7360

A Rare Breast Carcinosarcoma in a Young Female

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

DOI: 10.9734/JCTI/2022/v12i430185

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/89820

Case Report

Received 22 May 2022
Accepted 27 July 2022
Published 03 August 2022

ABSTRACT

Metaplastic breast cancer (MBC) is a rare aggressive type of breast cancer, as this form of cancer makes up only 1% of overall breast cancers. It mainly presents with a rapid-growing mass. Establishing the diagnosis is based on histopathology, as imaging studies show the same features as other types of breast cancer. The treatment protocol for MBC is similar to those for invasive ductal carcinomas owing to the lack of standardized management modality for metaplastic breast cancer per se.

Keywords: Metaplastic carcinoma; breast cancer; histopathology; mammogram.

1. INTRODUCTION

Metaplastic carcinoma of the breast was recognized by the World Health Organization as a unique pathologic entity; due to its rare and aggressive nature. It accounts for 0.2-5% of all breast cancers. It is characterized, histologically, by the presence of two or more cellular types; epithelial and mesenchymal components typically. Its diagnosis is complicated as it shares many similarities with invasive ductal carcinoma and benign lesions on mammography. Due to its rarity, standardized treatment guidelines are lacking, and little is known about its prognosis.

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Yet, studies suggest the greatest benefit is achieved with surgical removal and adjuvant radiation therapy [1, 2].

Here, we report a case of metaplastic breast cancer and provide our suggested treatment plan.

2. CASE PRESENTATION

A 31-year-old previously healthy female presented with a huge right breast mass. She first noticed the mass eight months prior to the presentation. As a result, the patient sought medical advice where her previous physician performed a fine needle aspiration of a large amount of brownish-colored fluid. Our patient did not follow up with her physician. However, when her breast started to get bigger and more painful; as she said, she came to our facility asking for medical advice. She is a grand multigravida; who has a history of giving birth to eight offspring by normal vaginal delivery. She denied a similar condition among family members; although there is a family history of breast cancer from the paternal side.

On clinical examination, there was an obvious asymmetry of both breasts, with a right-sided nodular huge mass occupying the whole breast, along with dilated congested-looking veins, as depicted in Fig. 1.

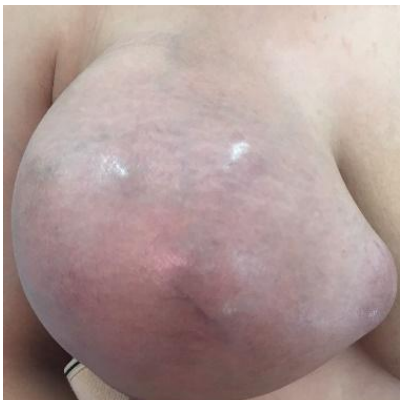

Fig. 1. The right breast on physical examination

Mammogram and Ultrasound were performed showing a mixed solid and cystic mass occupying most of the right breast measuring about 17.3 * 15.23* 16.4 cm with scattered calcification, internal color flow, and surrounding sub-areolar stellate masses. Moreover, multiple thickened cortices and compressed fatty hila of the right axillary lymph nodes were detected. These findings are highly suggestive of malignancy; BIRADS 5.

No doubt that core needle biopsy is an asset in the diagnostic process of breast cancer, which confirmed the presence of an MPC. A breast true cut core biopsy was carried out and revealed the presence of infiltration of invasive mammary carcinoma ductal type, however, ductal carcinoma in situ was not identified neither was lymphovascular invasion.

Nevertheless, few foci of the tumor showed infiltration by high-grade tumor sarcomatoid features representing the histological findings of metaplastic carcinoma. Fine needle aspiration of the right axilla showed clusters & single cells with atypia (high Ni ratio irregular Nuclear Membranes) admixed with reactive lymphoid cells and macrophages suspicious of malignancy.

On the other hand, the left FNA declared that a hemorrhagic smear revealed reactive lymphoid cells and macrophages with no malignant cells.

A staging study was an obligatory next step in the management of our patient; therefore, a CT scan was conducted to rule out distant metastasis. There were no radiological signs of infiltration. Moreover, the bone scan study showed no bone metastatic lesions.
Our treatment plan mainly focused on neoadjuvent chemotherapy and surgical removal of the mass hence the patient was referred to radiation oncology.

3. RADIO-IMAGING

On Ultrasound, a mass occupying most of the right breast measures 17.3 * 15.23* 16.4 mm with intra-mass scattered calcification, correlating with ultrasound complex solid and cystic mass. There is focal asymmetry at the upper outer left breast about 3.8 cm from the nipple.

A mammogram showed a huge mixed solid and cystic mass occupying most of the right breast with internal color flow and surrounding subareolar stellate mass. There are mildly dilated ducts with mobile internal debris in the left breast.

The right axillary lymph nodes had thickened cortices and compressed fatty hilum, while the left axillary lymph nodes had mildly thickened cortices and preserved fatty hilum.

4. CHEST ABDOMEN AND PELVIS CT SCAN

Right sided breast mass measuring 17.3*15.23*16.4 mm and right axillary lymph nodes.

No definite lung mass or nodule, no significant hilar or mediastinal lymphadenopathy, no pneumothorax, no pleural thickening, or pleural effusion.

The liver, spleen, pancreas, both adrenals and both kidneys appear unremarkable apart from a small cortical cyst seen in the left kidney.

No evidence of para-aortic or mesenteric lymph nodes enlargement.

5. LABORATORY FINDINGS

Serum level of AST was high 35.4 U\L. Also ALT was high 67.3 U\L, LDH was 285 U\L (135-214), creatinine was low 34 (44-79).

Other laboratory values of the patient were within the normal range.

6. DISCUSSION

Metaplastic carcinomas are a rare entity of malignant aggressive neoplasms. Metaplastic carcinoma can affect the breast, ovary, and uterus [3, 4]. Metaplastic breast cancers (MBC) account for 0.08–0.2% of all breast malignant tumors. The SEER database reported less than 10,000/ year cases of MBC in the USA between 1973 and 2015 [5].

To distinguish MBC from other types of breast neoplasms and specifically the rare tumor types, it is important to identify the clinical and pathological features of the mass. According to histopathology, these tumors have malignant fibroblastic and epithelial cellular components [3]. Many theories have discussed the MBC origin. Collision theory, combination theory, and the conversion/metaplastic theory were introduced. Collision theory reported that sarcomatous and carcinomatous cells develop from separate progenitor cells. However, the monoclonal combination theory suggests that both sarcomatous and carcinomatous cells were generated by common multipotent cells. On the other hand, the conversion/metaplastic theory conveys that the sarcomatous portion is made out of carcinomatous particles via a metaplastic process. The conversion/metaplastic theory had evidence of reported data concluding that both epithelial and mesenchymal components of the tumor have cytokeratin expression [5]. Some new studies convey that these tumors have an origin of a single stem cell that develops into myoepithelial cells with a biphasic differentiation [3], as myoepithelial markers including CD10, p63, and actin are positive in this entity [6].

Breast carcinosarcomas are subdivided – according to the WHO classification- as mixed metaplastic carcinoma, low-grade adenosquamous carcinoma, fibromatosis-like, squamous cell carcinoma, spindle cell carcinoma, and metaplastic carcinoma with mesenchymal differentiation. Wargotz used another subtype categorization that MBCs were divided into five major groups (carcinosarcoma, matrix-producing carcinoma, spindle-cell carcinoma, squamous cell carcinoma, and osteoclastic giant cell carcinoma) [4].

Our patient presented at a quiet young age taking into consideration that most MBC cases are diagnosed in the fifth decade. The clinical reports of the SEER database (from January 1998 to December 2016) showed that the majority (81.2%) of the MBC subjects were above 50 years [4] Nevertheless, the median age was 45.5 years according to a retrospective study that was done in Ankara hospital [7].

Assessing any breast mass demands the triple-modality approach; physical examination, radiological testing, and the histopathological features of the biopsy. The clinical picture of our patient was a well-defined, nodular, firm, concrete breast mass (Fig. 1). However, some patients may present with more benign-like clinical features such as a well-defined circular-shaped regular lump. This may be misdiagnosed by the similar benign presentation of the fibro-adenomas [4].

The next step would be radiological imaging. The imaging modalities we used included ultrasonography and mammography. At this point, these techniques are suboptimal when it comes to diagnosing breast neoplasms [8] Magnetic resonance mammography would add to the accuracy of the diagnosis but wasn't readily available at our institute. To overcome the challenging nature of diagnosing breast neoplasms, bedside biopsies including fine needle biopsy and/or core biopsy can be obtained preoperatively [8] The diagnosis of

breast masses by frozen section is accurate, with a sensitivity and specificity percentage of more than 90 and 99%, respectively [9]. Conversely, in MBC, there may be some limitations to frozen section diagnosis.

By the histopathological and hormonal tests this type of cancer is typically considered triple-negative breast cancer (TNBC). In other words, there is no expression of estrogen receptor (ER), progesterone receptor (PR), or human epidermal growth factor 2 receptor (HER2) [6]. In regards of our patient the histopathological testing revealed a negative response to receptors (TNBC).

In addition to that, in comparison to other TNBC, metaplastic carcinoma of the breast has a worse prognosis. Emphasizing the essential need to have more understanding of MBC. Despite MBC having a similar clinical presentation to invasive ductal carcinoma [3], MBC presents with large tumors, high histological grade, heterogeneity, overexpression of Ki-67 and p53 as well as less lymphatic invasion as the metastasis of the tumor is through a hematogenous route [6, 9]. As a consequence, it is far more aggressive than invasive ductal carcinoma, even when compared with the same age, stage, and tumor grade [7]. Most MBC patients present with an advanced stage of the disease at the time of diagnosis, with spread to the pleura and lungs then metastases to the bone and liver [4,8].

When compared to the triple-negative breast neoplasms whether lobular or ductal types, metaplastic carcinomas present with a more advanced and aggressive disease on a local basis. The management plan almost follows the guidelines of TNBC because there is no standard protocol for the treatment of such a rare tumor type [10]. The surgical approach could be used as an option for treatment as seen in some reported cases of a young age [8]. Systemic chemotherapy is a bad treatment modality since MBC is more chemorefractory than TNBC [6], besides the poor clinical side effects observed [6]. In addition to that, these patients have a higher recurrence rate, and a short remission period as well as less overall survival time [6].

7. CONCLUSION

Metaplastic carcinoma of the breast is one of the rarest breast neoplasms that has a bad prognosis compared to other malignant breast tumors. There is not enough data resources for the epidemiological distribution, staging guidelines or prognostic factors, hoping to conduct more research focused on the treatment options and the prognostic staging to improve the patients' outcomes.

ETHICAL APPROVAL AND CONSENT

As per international standard or university standard, patients' written ethical approval and consent has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/89820