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Review Article

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MUCINOUS CYSTADENOCARCINOMA OF THE BREAST: A CASE REPORT AND REVIEW OF THE LITERATURE

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ABSTRACT

Mucinous cystadenocarcinoma (MCA) is an unusual variant of primary breast carcinoma that bears a striking resemblance to MCAs of the ovary, pancreas and appendix. The metastatic lesion must be ruled out before the diagnosis can be determined. The differential diagnosis of MCA includes metastasis from other organs and cystic hypersecretory breast carcinoma. The incidence of breast mucinous cystadenocarcinoma is about 1-6% of primary breast cancers. It is usually diagnosed in postmenopausal women. The entity is usually triple-negative breast cancer. Herein we describe a case of 50-year-old female patient diagnosed histological with mucinous cystadenocarcinoma of the left breast. Immunohistochemistry profile showed triple negative and a high proliferation index. The biological behavior of MCA of the breast is reportedly favorable despite having a high proliferation index and triple-negative biomarker status. The importance of chemotherapy and radiotherapy is still debatable given the positive overall prognosis. The purpose of this study is to add to the body of literature by describing another instance of this uncommon case.

KEYWORDS: mucinous cystadenocarcinoma, breast, triple negative, high proliferation index.

INTRODUCTION

Mucinous cystadenocarcinoma (MCA) is an extremely rare variant of primary breast tumor which is histologically similar to mucinous cystadenocarcinoma of the ovary, pancreas and appendix.[1.2] It was firstly described in 1998 by Koenig and Tavassoli, who reported 4 cases.[3] Thus, the final diagnosis cannot be made before the metastatic origin of the lesion is excluded.[2] It was defined as a malignant tumor formed by intracellular mucin-rich columnar cells lining the wall of the cyst.[4] The incidence of breast mucinous cystadenocarcinoma is about 1-6% of primary breast cancers.[5.6] MCA of the breast is usually diagnosed in postmenopausal women.[2] It is only proved that the vast majority of these tumors are triple negative.[2] The aim of this study is to enrich literature with another single case diagnosed with this rare entity.

CASE REPORT

A 50-year-old female patient with history of ectopic pregnancy, menopausal 2 years ago, admitted to our hospital upon the palpation of a mass in her left breast. The history of the disease dates back to 6 months before her admission, with the appearance of a left breast nodule with no other associated signs. Clinical examination revealed a performance status of 1, an asymptomatic patient, a left breast nodule measuring approximately 3 cm, located at the union of the external quadrants; the contralateral breast was without abnormality. On mammography, the left breast showed a tissue nodule with microcalcification of irregular contours, measuring 28x22 mm, located at the junction of the external quadrants classified BIRADS V, left axillary nodule of 19x5mm suspicious; the right breast without abnormality. Micro-biopsy was performed on the left breast nodule, and cytologic examination revealed an invasive adenocarcinoma proliferation. It consists of glandular structures, often dilated and cystic, containing necrotic material with occasional calcifications. These cystic spaces are lined with columnar or cuboid cells whose cytoplasm contains mucosecretion vacuoles that stain with Alcian blue, and the diagnosis was type-specific invasive breast adenocarcinoma suggestive of grade III invasive breast mucinous cystadenocarcinoma. Immunohistochemical complement showed negative hormone receptors (RE 0%, RP 0%), negative HER2 (score 0) and proliferation index ki 67=70%. A thoraco-abdominal-pelvic CT scan showed a left breast mass classified cT2N0M0. The patient is currently undergoing neoadjuvant chemotherapy.

DISCUSSION

Breast cancer is a heterogeneous group of tumors consisting of different histological subtypes,

characterized by various biology, clinical course and prognosis. Among them there are some rarely diagnosed subtypes, including mucinous cystadenocarcinoma (MCA), occurring as a pure form or mixed with foci of other histological types of breast cancer[2] According to the WHO classification, the mucin-producing carcinomas of the breast are divided into four histologic subtypes, including mucinous carcinoma, mucinous cystadenocarcinoma (MCA), columnar cell mucinous carcinoma (CCMC), and signet ring cell carcinoma[7] The primary mucinous cystadenocarcinoma of the breast usually reported in postmenopausal females aged between 47 and 96 years[8], with median age of 61 years.[9] The entity is usually triple-negative breast cancer (TNBC), which is negative for estrogen receptor (ER), progestogen receptor (PR) and human epidermal growth factor receptor 2 (HER2) expression.[3,10] Our patient is a 50-year-old post-menopausal woman, diagnosed with triple-negative mucinous cystadenocarcinoma of the breast, which is perfectly in line with the literature. The most common sites of mucinous cystadenocarcinoma include the ovary, pancreas, and appendix.[8] When a patient develops this kind of tumor, we should claim that it is not a metastasis from distant organs (pancreas, ovary appendiceal cancer). The usual immunohistochemical profiles, to rule out metastasis from distant mucinous cystadenocarcinoma are the following: CK7 positive, CK20 negative, and CDX-2 negative. These results can help to exclude the possibility of metastatic mucinous cystadenocarcinoma from the ovary, pancreas, and gastrointestinal tract.[8] The differential diagnosis of MCA includes metastasis from other organs and cystic hypersecretory breast carcinoma due to the fact that they both show a cystic appearance,[3] and likewise cytologic image.[11,12] The differential diagnosis lay on the immunohistochemical profile in the former and on the presence of tall columnar cells in the latter.[1]

Microscopically, there are cystic spaces which are lined by mostly bland-looking columnar mucinous cells with some papillary formations. Nuclear atypia is evident in some foci, with some eosinophilic squamous cell differentiations.[3] It is difficult to exclude benign or borderline lesions and to make a definite diagnosis of malignancy based on morphology alone.[13] However, the absence of myoepithelium, frequent association with ductal carcinoma in situ (DCIS), and the capability of metastasis suggest malignancy.[14] The morphological spectrum of MCA ranges from pure MCA and MCA with DCIS to MCA with both DCIS and invasive ductal carcinoma (IDC).[3–10] The diagnosis of MCA without DCIS is challenging because overlapping morphological features are not uncommon among the entities, suggesting the use of broad immunohistochemical biomarkers. The combination of clinical history, morphology, and immunohistochemistry is helpful to confirm the diagnosis.[9] According to Lee and Chaung[15], the neoplasm is the result of an accumulation of mucin within intraductal papillary carcinoma with mucinous metaplasia and extracellular mucin production. This causes cystic dilation of the lumen with loss of myoepithelial cells and invasion of the adjacent stroma.

Lymph node metastasis is infrequent but is reported in the literature.[13] In our patient's case, the CT scan showed that there were no lymph node metastases, which is in keeping with the literature.

The tumor exhibited features like the association with ductal carcinoma in situ (DCIS), lymph node metastasis, and estrogen receptor positivity which are rarely reported in the literature.[13]

Because of low incidence of lymph node involvement, mucinous cystadenocarcinoma has a relatively good prognosis.[16] The biological behavior of MCA of the breast is reportedly favorable despite having a high proliferation index and triple-negative biomarker status.[8] However, because of the favorable overall prognosis, the significance of chemotherapy and radiotherapy remains doubtful. What is more, the triple negative molecular phenotype and high proliferative index of primary breast MCA can lead to false perceptions or overtreatment with redundant radiotherapy and/or chemotherapy in some patients.[10]

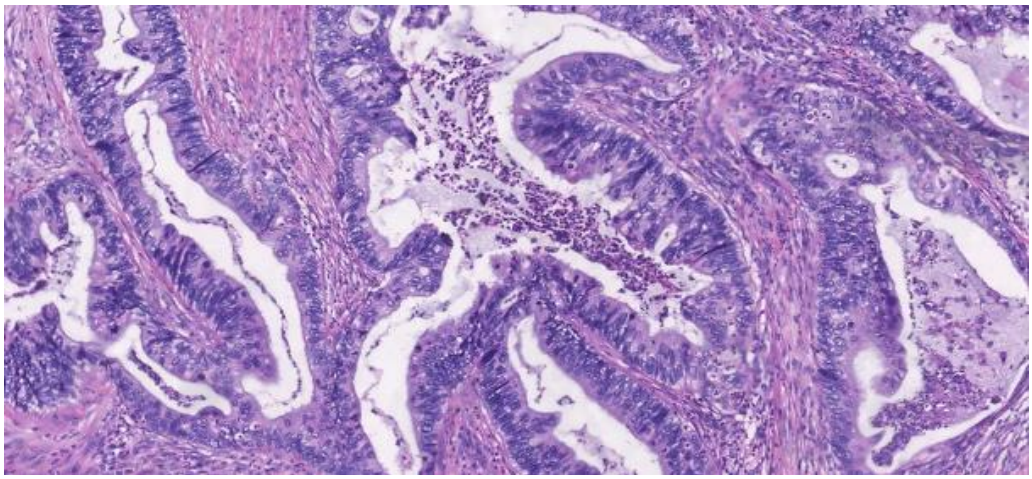

Figure 1: Microscopic pathology image showing mucinous cystadenocarcinoma of breast.

CONCLUSION

Mucinous cystadenocarcinoma of the breast is a very rare primary breast carcinoma belonging to the same histological family as the pancreatic, ovarian and appendix mucinous cystadenocarcinomas, which produce mucin. The combination of clinical history, morphology, and immunohistochemistry is helpful to confirm the diagnosis.

To create criteria for diagnosis and therapy, further studies on more cases are required.

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