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Case of Pure Mucinous Breast Carcinoma: A Rare Entity

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Abstract

Pure mucinous breast carcinoma is a rare subtype of invasive breast cancer characterized by the production of extracellular mucin. It typically demonstrates a distinctive histological appearance, higher hormone receptor positivity, and favorable clinical outcomes, with a good response to endocrine therapy. This case highlights the importance of recognizing pure mucinous carcinoma as a distinct pathological entity for optimal management and prognosis.

Summary

Pure mucinous breast carcinoma is a rare subtype of invasive breast cancer characterized by the production of extracellular mucin. It has a distinctive histological appearance, fewer chances of lymph node involvement, higher hormone receptor positivity, favourable clinical outcome, and good response to endocrine treatment. We report one such case in a 52-year-old female having a left breast mass of approximately 5 months duration with no nodal involvement. USG showed a hyperechoic lesion measuring 26 x 21 x 27 mm. FNAC showed mild to moderate pleomorphic epithelial cells with a moderate amount of mucin and 'chicken wire' blood vessels. Histopathology of the resected specimen showed clusters of tumour cells floating in pools of abundant extracellular mucin partitioned by fibrous septa containing blood vessels. Immunohistochemistry revealed ER and PR positivity and Her-2 Neu was negative. 6-month follow-up CT scan showed no recurrence. This case underscores the significance of recognizing this subtype as a distinct clinical entity.

Keywords: Malignancy, mucin, histology, hormone, receptors, fine needle aspiration cytology, Her-2 Neu, oestrogen, progesterone

Introduction

Mucinous carcinoma (MC) is an uncommon histological subtype and accounts for 1 to 7% of all breast carcinomas[1-3]. It is classified into two types: pure mucinous breast carcinoma (PMBC) and mixed mucinous carcinoma (MMC). PMBC expresses more than 90% mucin and MMC has a mucinous expression between 51-90% [4]. PMBC accounts for about 2% (1-6%) of all invasive breast cancers [1-3]. A recent Indian study reported the incidence of mucinous carcinomas to be 3.4% (14/420 patients) of all breast cancers with equal numbers of PMBC and MMC. The mean age of patients with PMBC was 52.1 ± 13.5 years while among MMC was 49.6 ± 11.2 years [5]. In the West, it occurs most

commonly in post-menopausal women aged 75 years or older, but in a Korean study, PMBC patients were younger than IDC patients [2]. Here we report a case of PMBC that is known to be rare and has a better prognosis than MMC [1, 2, 6].

Case Report

A 52-year-old female presented with a gradually increasing lump in the upper quadrant of her left breast for 5 months. It was not associated with pain, redness, warmth, discharge, nipple inversion, skin changes, or trauma to the breast. A firm nodular lump was palpable in the upper and outer quadrant of the left breast with areas of softness in it. It was fixed to the underlying structures; the overlying skin was normal with no signs of retraction of the nipple. No axillary lymph node was palpable. On USG, a well-defined hyperechoic lesion of 26 x 21 x 27 mm was seen at a 12-clock position.

Fine needle aspiration cytology (FNAC) showed cellular smears that comprised of mild to moderately pleomorphic epithelial cells in the form of small clusters, papillae as well as singly scattered cells. Nuclei had bland chromatin, indistinct nucleoli, and round, regular nuclear membrane. Binucleation was present. Nuclei were eccentrically placed and showed mild atypia with occasional mitosis. The cytoplasm was moderate to abundant and vacuolated. The background showed moderate amounts of mucin and 'chicken wire' blood vessels. These micropapillary structures were floating in pools of abundant extracellular mucin (Figure 1a, 1b, 1c). A diagnosis of malignant epithelial neoplasm suspicious for mucinous carcinoma was given and a biopsy was advised for confirmation and exact categorization.

Modified radical mastectomy and axillary lymph node excision were done and a skin-covered specimen measuring 16 x 12 x 4 cm was received. The overlying skin appeared normal. The cut surface showed a tumour measuring 2.5 x 2.5 x 2 cm having grey-white to grey-black solid and gelatinous areas (Figure 2a). On cutting, a gritty sensation was felt. Nipple-areola was identified on the anterior surface. No scar, ulceration, cysts, or nodules could be identified. Growth was 0.5 cm away from the posterior margin, 2 cm away from the superior margin, 7 cm away from the inferior, medial, and lateral margins. Another jar of specimen contained an axillary tail measuring 6 x 4 x 1.5 cm. On cutting, five lymph nodes were identified, the largest measured 0.8 cm. The cut surfaces of all nodes were grey-white to brown. Sections were taken from nipple-areola, growth, and all margins and lymph nodes. Histopathological examination showed clusters and nests of tumour cells exhibiting solid, acinar, and micropapillary structures. These clusters were floating in pools of abundant extracellular mucin partitioned by fibrous septa containing blood vessels (Figure 2b, c). Nuclei showed mild atypia with occasional mitosis. Lympho-vascular invasion was not seen. Posterior and superior margins were involved by the tumour, while the other margins were free. Five lymph nodes showed reactive hyperplasia. Components of IDC were not seen. Mucicarmine stain was positive (Figure 2d). A diagnosis of PMBC (Grade 1), with a pathological stage of PT2N0MX was made. Immunohistochemistry (IHC) revealed both ER and PR positivity and Her-2 Neu was negative (Figure 3 a, b, c). She was followed up until 6 months and was asymptomatic with no fresh complaints.

Discussion

PMBC is a special histologic subtype of invasive carcinoma, that is rare. The diagnosis is suspected by FNAC findings and confirmed by histopathology of the resected specimen. FNAC findings include low cellularity, mild pleomorphism of nuclei, free mucin presents outside the cell, tumour cells having

eccentrically placed nuclei and vacuolated cytoplasm due to intracellular mucin, mucinophages (histiocytes which have engulfed the extracellular mucin and having central to eccentrically placed nuclei with vacuolated cytoplasm), and branching vessels along with tumour cells and mucin material [5]. The present case was also suspected based on FNAC findings. Its gross appearance is soft, rubbery, and gelatinous having pushing or circumscribed borders. Microscopically, the tumor cells are arranged in clusters and small islands of cells within large lakes of mucin[4].

In a series of 16 cases of mucinous carcinoma out of 712 cases of breast carcinoma (2.2%) in women with mean age of 51.88 years ± 14.62 years, and left breast and upper outer quadrant being the most common site of involvement. Our case also falls in the same age group and had a similar location. Identification of PMBC through FNAC and its distinction from MMC holds significant diagnostic and clinical importance[7]. In comparison, MMC cases show a high Ki67 index and increased p53 expression [5,7].

A retrospective analysis of 11,400 cases of PMBC reported in the SEER database between 1973 and 2002 revealed a favourable outcome till 20 years after diagnosis. Its 10, 15, and 20 years survival were 89%, 85%, and 81% respectively compared to 82% (5 year), 72% (10 year), 66% (15 year), and 62% (20 year) for IDC. Multivariate analysis showed nodal status to be the most significant prognostic factor followed by age, tumour size, progesterone receptors and nuclear grade[6]. Thus, studies have confirmed that PMBC has less aggressive behaviour, smaller size, lower grade, lower mean Ki67 index, and lesser nodal involvement when compared to IDC [5,6,7]. Only 3-15% of the pure variety shows axillary node metastasis compared to 33-46% of the mixed type. Moreover, a higher frequency of ER and PR expression and a lower frequency of Her2/neu expression signifies a better biomarker profile and predicts a better response to hormonal therapy in the case of MC [5,6,7]. MC should be differentiated from fibroadenoma and mucocele-like lesions. 50% of MC's are oval shaped, so, circumscribed lesions on imaging may be misdiagnosed as fibroadenoma [8]. Mucocele-like lesions are characterized by cystic architecture and dilated cuboidal epithelium-lined ducts that are filled with mucin and can rupture to expel their contents into the periductal stroma, a key distinction from mucinous carcinoma [9].

Approximately 10% to 20% of women without axillary lymph node metastases recur with distant metastasis. In these patients, metastasis may occur via the internal mammary lymph nodes or hematogenously [10]. Our present case did not show any distant metastasis either at the time of diagnosis or during the 6 months of follow-up. Patients with PMBC who are older, hormone-positive receptor and HER-2 negative status may be treated with a less aggressive approach like breast conservation and endocrine therapy. However, HER2-positive PMC of the breast needs more aggressive oncologic and surgical considerations, such as mastectomy and chemotherapy along with adjuvant Her2-directed trastuzumab[10]

Conclusion

We have reported a case of PMBC whose early diagnosis was suggested by FNAC and averted further complications. There were no distant metastatic lesions at the time of diagnosis and at 6 months of follow up suggesting a good overall prognosis.

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