Lung Recurrence of Pure Mucinous Breast Cancer After 20 Years

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Abstract
Pure mucinous breast carcinoma is a rare histologic type of mammary neoplasm, representing 1-4% of all breast cancers. We present the case of a 45-year-old patient presenting with a painless lump on the upper outer quadrant of her right breast of 1 year duration. The mammogram demonstrated a 2.1 cm mass on the upper outer quadrant with irregular margins. Fine-needle aspiration results revealed pure mucinous breast cancer. The patient subsequently underwent a right modified radical mastectomy which included axillary lymph node clearance. All margins and axillary nodes were negative. The follow-up exams showed no signs of relapse or distant metastasis.

20 years after the initial breast cancer diagnosis the patient complained of cough, chest pain and fatigue. Thoracic computed tomography with intravenous contrast revealed a mass with soft tissue density on the left superior pulmonary lobe with 28 x 33 x 24 mm. The pathology of the endobronchial mass showed an adenocarcinoma with an immunohistochemistry profile compatible with breast carcinoma. The patient is presently under systemic chemotherapy treatment.

Keywords: Breast cancer; Lung recurrence; Pure mucinous carcinoma

Introduction
Mucinous carcinoma is categorized as a special type of breast cancer, presenting with a massive production of extracellular mucin. From a histologic perspective, mucinous carcinoma may be divided in pure mucinous and mixed mucinous entities, depending upon the quantification of cellularity. The pure type is very rare and consists exclusively of tumor tissue with extracellular mucin production, while the mixed form, also contains an infiltrating ductal epithelial component without mucin [1]. We present the case of a patient with a lung metastasis of a pure mucinous breast carcinoma occurring 20 years after the initial diagnosis.

Case report
A 45-year-old woman presented with a painless lump on the upper outer quadrant of her right breast of 1 year duration. She denied occupational or radiation exposures,
comorbidities, use of medications or history of breast cancer in her family. At physical examination the patient presented a palpable mass on the upper outer quadrant of the right breast with about 2 x 2 cm, mobile with non-defined margins and no apparent fixation to the skin or underlying structures. The overlying skin was normal and the patient had no evidence of nipple retraction or discharge. Bilateral axillary examination revealed lymphadenopathies on the right axillary zone. The opposite breast and axilla were normal. The mammogram demonstrated a 2.1 cm ovoid mass on the upper outer quadrant with irregular margins.

The mass was suspicious for malignancy and the patient underwent a fine-needle aspiration of the mass apart from other routine hematological and biochemical investigations. Fine-needle aspiration results revealed pure mucinous breast cancer. Other routine investigations were within normal limits. Metastatic work-up revealed no evidence of metastasis.

The patient subsequently underwent a right modified radical mastectomy which included axillary lymph node clearance. The final surgical pathology revealed a mass with 2.5 cm corresponding to invasive pure mucinous breast carcinoma. All margins were negative. Eighteen lymph nodes from the right axilla were analyzed, and were all negative for metastasis. There was no vascular or perineural invasion.

The patient did not undergo subsequent radiation or endocrine therapy. An annual follow-up with physical examination, mammogram, and chest radiography and serum cancer biomarkers was performed. The follow-up exams showed no signs of relapse or distant metastasis.

20 years after the initial breast cancer diagnosis the patient complained of cough, chest pain and fatigue. The patient denied anorexia or weight loss. The physical examination revealed only wheezing on the pulmonary auscultation.

The chest radiography showed a nodular mass on the left hemitorax. On the same day a thoracic computed tomography (CT) with intravenous contrast revealed a mass with soft tissue density on the left superior pulmonary lobe with 28 x 33 x 24 mm, irregular margins, peripheral contrast capturing and a central non-capturing zone suggesting necrosis. The mass contacted with the adjacent pleura and leaded to obstructive atelectasis of the lingula. The apical segment of the same lung presented a mass with soft tissue density and irregular margins with 11 x 14 x 8 mm, strongly suggestive of atypia. No suspicious lymphadenopathies were identified in the thoracic CT.

The bronchofibroscopy showed a voluminous hyper vascularized exophytic mass with implantation on the pulmonary lingula. As a profuse hemorrhage occurred after only touching the mass with the biopsy needle, a rigid bronchoscopy was scheduled.

The rigid bronchoscopy showed a mass on the final portion of the left main bronchus which was removed with laser. The bronchial secretions analysis was positive for adenocarcinoma. The pathology of the endobronchial mass showed an adenocarcinoma with an immunohistochemistry profile compatible with breast carcinoma. The lesion was estrogen and progesterone receptor positive. The abdominopelvic CT and the brain CT showed no other signs of metastasis. The patient is presently under systemic chemotherapy treatment.

**Discussion**

Mucinous carcinoma is one of the rarer breast neoplasms accounting for 1% to 7% of all invasive breast carcinomas. A prevalence as high as 7% is found in women over the age of 75 years, while a prevalence of 1% is found in those younger than 35 years [2].

Pure mucinous breast carcinoma (PMBC) is a rare histologic type of mammary neoplasm, representing 1-4% of all breast cancers and usually occurs in women aged over 60 years. It is generally associated with a better prognosis, longer disease-free interval and higher estrogen receptor and progesterone receptor expression [1,2,3]. Axillary lymph node involvement is rare and was detected in 12% of the patients in the review by Saverio and 14% in the series by Komenaka [1].

In PMBC mucin comprises the majority of the tumor volume. The volume of mucin contributes to an overestimation of tumor size, and thus early detection could contribute to the very good prognosis seen in PMBC. The overexpression of mucinous proteins, results in the production of excess gel-forming types of mucins, which might act as a barrier to cancerous extension. Mucinous carcinoma tends to grow slowly, but can sometimes reach a large size by the time of diagnosis since its mucinous content does not feel solid or firm upon examination [1].
Most mucinous carcinomas are readily detected on mammography. They appear as low-density, well-defined or microlobulated oval masses and generally belong on the category of “well-circumscribed” breast carcinomas. Microlobulated margins have been associated with higher mucin content, while irregular or speculated margins correspond to lower percentages of mucin and infiltrating margins histologically [2].

Sonographically, mucinous carcinomas typically present as complex masses of mixed echogenicity with solid and cystic-appearing components. However, 20% of these lesions may present as homogenous masses on ultrasound. They are isoechoic or hypoechoic to subcutaneous fat, with posterior acoustic enhancement being a common finding [2].

On magnetic resonance imaging mucinous carcinomas frequently present as lobulated tumors, with dynamic time-signal intensity curves that exhibit a gradual contrast enhancement or plateau-type pattern after the initial upstroke. They classically demonstrate high signal intensity on T2-weighted images due to intrinsic mucin component. Signal intensity on T1-weighted images varies from low to high, a feature that is largely dependent on the protein composition of the tumor.

Fine-needle aspiration has been described to yield copious amounts of mucinous material in mucinous carcinoma with a variable proportion of tumor cells. The tumor cells are described as being generally small and fairly uniform with minimal atypia [4].

Despite advances, 20% to 30% of patients with early breast cancer will experience relapse with distant metastatic disease. Risk of recurrence is influenced by stage at initial presentation and the underlying biology of the tumor [5].

Patients with mixed mucinous breast carcinoma have been described to have a significantly higher recurrence rate than those with PMBC [6]. PMBC has been associated with outstanding overall and disease-free survival in the literature. However, because of its rarity, most studies on PMBC have few case numbers, and information after long-term follow-up is limited.

The literature recommends that this subtype of cancer should be treated less aggressively than infiltrating ductal carcinoma [3]. A study by Anan et al suggested that patients with PMBC, except for those invading the local skin, are suitable candidates for breast-conserving therapy, probably even in the presence of large tumors up to 5 cm in diameter [1]. Axillary staging by sentinel lymph node biopsy, administration of adjuvant radiotherapy and endocrine therapy after breast-conservation are recommended. However larger data samples with longer follow-up are necessary to gain a better understanding of PMBC [1].

The fact that the tumor displays the ability for late recurrence shows that despite its indolent local behavior, it might become systemic. This indicates that it needs longer to proliferate and manifest clinically, and hence it should be followed-up over a long period of time. However this case highlights that even with a close follow-up and normal routine exams the disease can become systemic.

References