

# Pure Mucinous Breast Carcinoma Mimicking a Benign Phyllode Tumor: Case Report and Review of Literature

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## Abstract

Pure mucinous breast carcinoma is a very rare histopathological type of breast cancer. Despite its invasive nature, mucinous carcinoma is most often described as a benign lump. It can be misdiagnosed as a phyllode tumor, which has certain similarities both clinically and radiologically. Surgery is the cornerstone of therapeutic management: lumpectomy or mastectomy depending on the size of the tumor, associated with axillary lymph node dissection or with sentinel lymph node biopsy procedure for early stage tumors. The treatment is often associated with adjuvant chemotherapy and radiotherapy. We describe an uncommon case of pure mucinous breast carcinoma, which clinical and radiological characteristics, occurred as a benign phyllode tumor presenting a real diagnostic challenge.

**Keywords:** Pure mucinous carcinoma, phyllode tumor, breast cancer.

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## INTRODUCTION

Mucinous carcinoma of the breast was first described in 1826 by the World Health Organization (WHO), defining it by the presence of malignant mucosecreting tumor cells floating in mucin [1]. Also called colloid or gelatinous or mucoid carcinoma, it constitutes a rare histological type of breast neoplasms, representing 1 to 4% of all breast cancers. The pure mucinous breast carcinoma subtype is extremely rare and account for about 2% of all primary breast carcinomas [2]. We report an original case of pure mucinous breast carcinoma, discovered in a 70-year-old patient with a large breast lump, which clinically and radiologically appeared as a phyllode tumor causing diagnostic difficulties. The diagnosis of mucinous breast carcinoma was only confirmed by histopathological examination.

## CASE REPORT

A 70 years old, single lady, nulligravida, menopausal for 20 years, with no personal or family history of breast cancer, who had noticed a painless lump in the right breast, discovered on self-examination, and was gradually enlarging.

Clinical breast examination revealed a smooth, well defined, firm, retro-areolar mass of 10x8 cm, non-fixed to the neither superficial nor deep tissue plane, with no inflammatory features or nipple discharge. The axillary hollow was free.

Mammography and ultrasonography showed two opacities with well circumscribed margins in the right breast, and no micro-calcifications. One was retro-areolar, with multi lobulated borders, that measures 7cm, the second was located at the upper inner quadrant measuring 06 x 3,5cm. No significant axillary lymphadenopathy was demonstrated either clinically or radiologically. The benign radiological appearance would favor the diagnosis of two phyllod tumors (Fig-1). A surgical biopsy was performed, revealing a mucocele-like lesion versus a mucinous carcinoma (Fig-2). Immunohistochemistry identified a specific subtype of pure mucinous carcinoma grade II SBR and showed estrogen and progesterone receptor positivity (ER & PR), receptor (Her 2) negative and Ki-67 (+) on 10% of tumor cells. The initial extension assessment was negative. A mastectomy with lymph node

dissection was performed to complete the surgical treatment. The final histological study identified a pure mucinous carcinoma, 16N-/ 16N, pT3N0Mx; the

patient received also an adjuvant chemotherapy. The evolution after 6 months was favourable, the extension assessment remains negative.

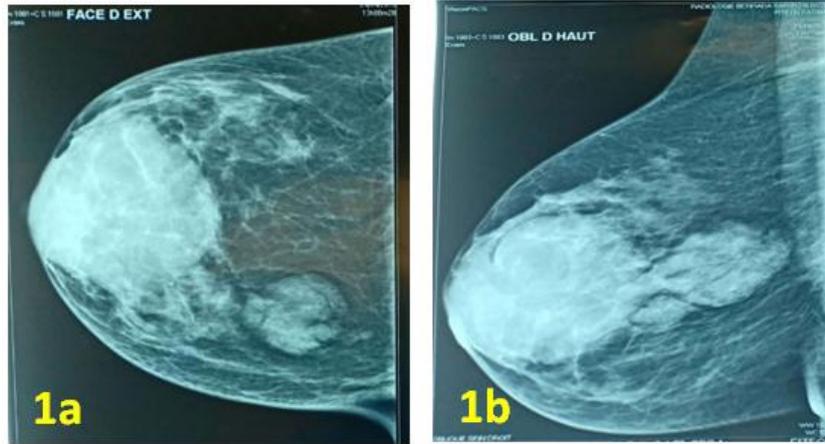


Fig-1: Mammography of the right breast showing two well circumscribed opacities with no microcalcifications (Fig 1a= frontal incidence; Fig 1b= oblique incidence)

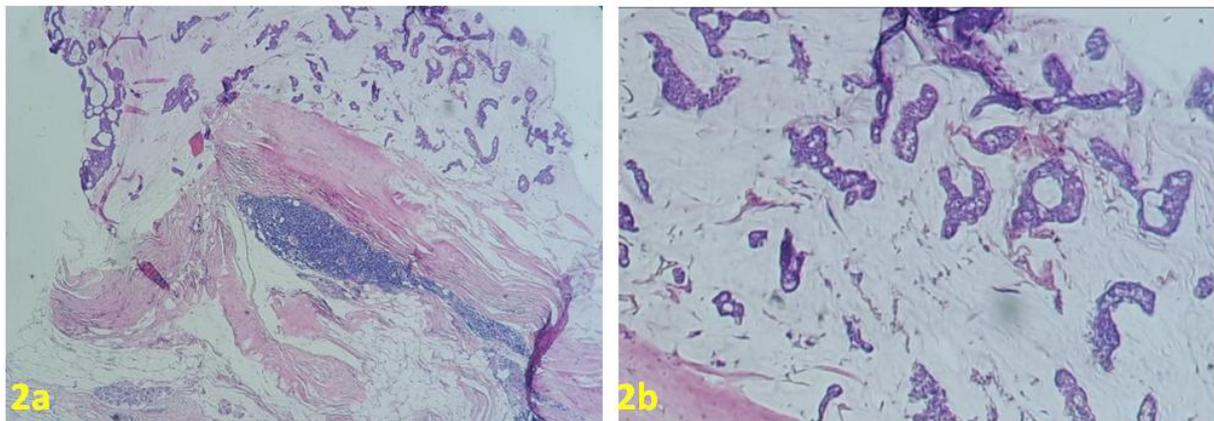


Fig-2: Histological features of mucinous breast carcinoma showing breast parenchyma site of an invasive carcinomatous proliferation (Fig 2a= microscopic magnification x4; Fig 2b= microscopic magnification x20)

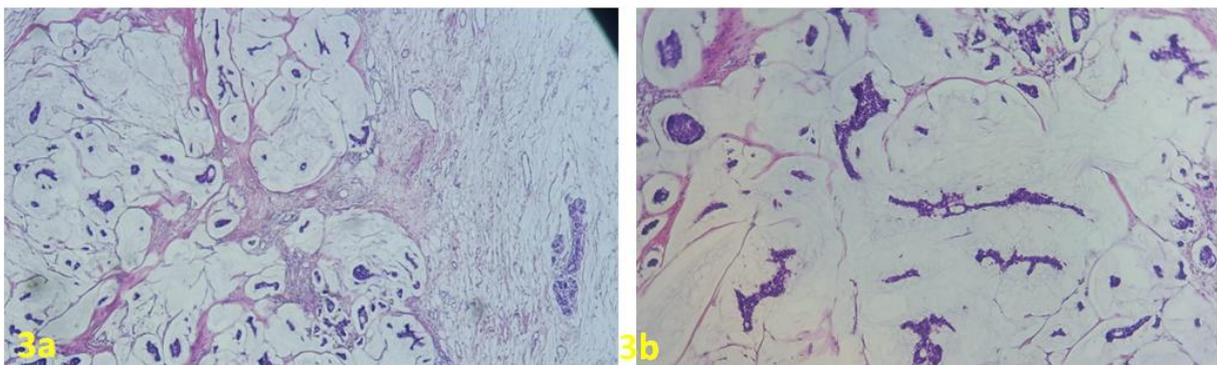


Fig-3: Light microscopy (x4 Fig 3a & x20 Fig 3b) demonstrates proliferation made of large pools of mucus occupying > 90% of the tumor, glands and clusters of tumor cells of varying sizes

## DISCUSSION

Mucinous carcinoma of the breast is a rare histological entity which preferentially affects women over sixty years. Its prevalence is 3% [3, 5]. This carcinoma is revealed in 80% of cases by auto palpation of a well-defined lump, classified T1 or T2 according to the classification Tumor Node Metastasis (TNM) [4, 5]

with extremes of 0.3 and 19 cm reported from the literature [6]. Axillary lymphadenopathy is rarely found (<25% of cases) [6, 7]. Barkley and DiSaverio have shown a correlation between tumor size and lymph node metastases. Indeed, the presence of a lymph node is associated with a larger tumor size [1, 8]. Nevertheless, we noticed in our patient, the absence of

lymph node invasion despite the large size of the tumor which reached 10 cm, orienting more towards a benign tumor.

According to Tan, Bode and Memis [9-11], pure mucinous carcinoma appears on mammography as a well circumscribed opacity similar to the aspect of a phyllode tumor, with regular contours, rarely associated with micro calcifications [12]. Falsely reassuring, the ultrasound complement shows in 71% of the cases posterior acoustic enhancement explained by the presence of cystic component in the pure type of mucinous carcinoma [11]; leading to misdiagnosis as a phyllode tumor or a reshaped cysts as it was for our patient. Therefore, the presence of a posterior enhancement should not lead to a wrong conclusion of a reshaped cyst, or a benign tumor. The mixed type of mucinous carcinoma appears as hypochoic masse with spiculated margins and a higher frequency of intra or peri-tumor calcifications, showing the aggressiveness of this subtype. Tumor vascularization can also be found in one third of the mucinous carcinomas [13]. The bilateral character is rarely described, while multifocality remains exceptional [7]. Our patient had the particularity of having a bifocal aspect of the tumor revealed on ultrasound.

MRI has higher sensitivity for detection of mucinous carcinomas, the main characteristic is a homogeneous hyperintense signal on T2 [10-12]. This is probably due to the specificity of the mucinous component. The diversity of the imaging findings calls for caution. No radiological criteria seem to allow an etiological diagnosis. Therefore, the diagnosis of mucinous carcinoma is considered as an imaging challenge.

Histologically, mucinous carcinoma is built up of rich extracellular mucin in which a cluster of tumor cells float as separate groups. It may occur in two forms [14]: the pure form in which all the tumor cells, even at the invasive margins, are surrounded by extracellular mucin constituting a real mechanical barrier attenuating cellular invasion. The fibrous reaction is weak or absent and the transition between mucus and surrounding connective tissue is abrupt.

The mixed form contains invasive areas not surrounded by mucin, often identical to the usual type of invasive ductal carcinoma or some of the special histologic types. The fibrous reaction is predominant and the transition between extracellular mucus and adjacent carcinomatous tissue is gradual. Most pathologists agree that a diagnosis of pure mucinous breast carcinoma should be reserved for tumors with at least 90% mucinous component [7]. The pure subtype is often well-differentiated with positive hormone receptors and human epidermal growth factor receptor (HER-2) negative.

The distinction between both subtypes is crucial due to its prognostic value. Neuroendocrine differentiation gives a favourable prognosis to mucinous carcinoma, in particular a lower nuclear grade, a greater expression of oestrogen receptors (91% of cases) [11] and a low incidence of lymph node metastases. In our case, it was the histological examination of the lumpectomy that confirmed the diagnosis, objectifying a mammary carcinoma of specific type: pure mucinous subtype SBR II, HER2-negative.

Studies show that HER2 status is an independent prognostic indicator of overall survival in pure mucinous breast carcinoma (PMBC). Given the unfavourable prognosis and aggressive clinical features of HER2-positive PMBCs, the determination of HER2 status in PMBC should warrant special attention [15, 16].

The therapeutic management is surgical [7] by lumpectomy or mastectomy depending on the size of the lump associated to an axillary lymph node dissection or a sentinel lymph node biopsy. Breast-conserving therapy must be carcinological and aesthetic in view of the favourable prognosis. As for the other therapeutic pillars, namely chemotherapy, radiotherapy and endocrine therapy, their indications are similar to those of other breast carcinomas [5, 6].

During the first assessment of extension, the discovery of metastatic locations is rare [8]. Several authors have agreed that lymph node status is the main significant prognostic factor, followed by age, size, hormonal and HER2 status [1, 2]. The low incidence of lymph node metastases in the pure forms of colloid carcinoma (2 to 14% compared to 45 to 64% in mixed forms) explains the favorable prognosis of the latter [5, 6], with 90 to 100% survival in ten years against 60% in mixed forms [17].

## CONCLUSION

Pure Mucinous carcinoma of the breast is a rare variety, occurring mainly in elderly postmenopausal women. Its clinical and radiological diagnosis can be confused with benign tumors, as the phyllode tumor. Caution is advised and histological proof is essential before any therapeutic treatment.

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