Mucinous carcinoma of the breast with neuroendocrine differentiation- a rare case report

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Abstract: Mucinous carcinoma of breast is one of the rarer forms of breast neoplasms which usually presents as a lobulated, well circumscribed mass with very low incidence of lymph node metastasis. It accounts for 1 -7% for all breast carcinomas. It is more common in elderly females with prevalence of 7% and in younger females the prevalence is as low as 1%. The mucinous neoplasms are characterized by the production of abundant extracellular and/or intracellular mucin. The definition requires a mucinous component >50% of the lesion. Mucinous carcinoma may be divided into pure mucinous and mixed mucinous entities. Herewith we present a case of breast carcinoma in a 65 year old female with mucinous carcinoma with neuroendocrine features.

Keywords: Mucinous carcinoma, neuroendocrine differentiation, chromogranin

INTRODUCTION
Breast carcinoma is a heterogenous disease with mucinous carcinoma being an uncommon variant with good prognosis. The mucinous Carcinoma requires 50% mucin component by definition[1]. However pure and mixed variants are identified. The pure type contains mucin in all the areas of the invasive duct cell carcinoma. Mucinous tumours can be divided into two groups: mucinous A (or paucicellular) and mucinous B (or hypercellular). Mucinous B cancers display histological features that significantly overlap with those of neuroendocrine carcinomas[2,3].

CASE STUDY
A 65 year old female presented with a mass in the left breast in the upper inner quadrant since one year gradually increasing in size with a sudden noticable increase in size since 2 months. The sudden increase in size was preceded by blunt trauma and attained the present size of 13X8 cm. There was no skin or nipple involvement and no axillary lymphadenopathy. Fine needle aspiration cytology yielded very few ductal epithelial cells with mild pleomorphism and stromal cells with a proteinaceous background. In view of the large size of the tumour and with above findings a diagnosis of phyllodes with cystic degeneration was offered. With this cytological diagnosis, simple mastectomy was done.

Gross: We received a simple mastectomy specimen measuring 15X8X6cm with skin flap measuring 6X3cm with an ulcer measuring 2X2cm. Cut section showed tumor mass measuring 14X9X6cm with variegated appearance with solid, haemorrhagic and cystic areas with mucoid material (Fig.1).

Microscopy:
Sections from tumour mass revealed large areas of necrosis with lakes of mucoid material. Tumour tissue was arranged in nests and trabeculae, some floating in pools of mucin (Fig.2).

Tumour cells were round to oval, pleomorphic with moderate cytoplasm, few showing vacuolated cytoplasm. Nuclei were oval vesicular with powdery chromatin with nucleoli in some Mitotic activity was 2-3 per 10 HPF. Nipple and areola were free from tumor infiltration. Deep resected margin showed tumour infiltration. However other margins were free. With these features we suggested mucinous carcinoma of breast. (MBRF score of 6).

In view of the nuclear chromatin nature we also thought of neuroendocrine differentiation and immunohistochemistry for Neuron Specific Enolase (NSE) and chromogranin in addition to routine estrogen receptor, progesterone receptor and Her2 neu were
done. Estrogen receptor and Her2neu were positive. Progesterone receptor was negative. We noticed that NSE and chromogranin showed diffuse cytoplasmic positivity. So a final diagnosis of pure mucinous carcinoma type B with neuroendocrine features was offered.

![Fig-1: gross picture showing abundant micin (white arrow) and extensive areas of hemorrhage.](image)

![Fig-2: H&E stained section showing tumour cells floating in abundant mucin pools.](image)
DISCUSSION

Worldwide, breast cancer is the most common form of cancer in women and listed as second leading cause of cancer death among women[4]. Breast cancer is the second most common cancer in the Indian female population[5]. Breast cancer is a heterogenous disease with many variants possessing different presentation, natural history, pathology and clinical outcome. Some may be indolent whereas some may be aggressive. Knowledge of special histologic subtypes like tubular and mucinous types which are low risk aids in clinical management. Mucinous or colloid carcinomas are the rare low risk breast malignancies with an incidence of 1-7%, more common in post menopausal women with low axillary node involvement[1]. Histologically MC is classified into 2 subgroups based on the degree of cellularity, which are pure type and mixed mucinous-ductal type [6]. However, cut-off values of the mucinous component for defining pure type MC was not standardized, and later, others divided MC into type A and type B[7]. They are grossly well circumscribed with cut section showing abundant mucin and hemorrhagic areas. These are mostly well-differentiated lesions frequently associated with positive ERs (>90%) and PgRs (81.5%) and HER-2-negative disease. However, rare cases with HER-2-positive staining have been reported[8]. Tumor size in the AJCC staging system may not be a significant factor because mucin comprises the majority of the tumor volume[9]. It is important to identify the neuroendocrine differentiation in mucinous carcinoma as it has bearing on the treatment of the tumor. According to the NCCN Clinical Practice Guidelines in Oncology [10] adjuvant endocrine therapy is indicated for tumors more than 3 cm in diameter and for MC patients with axillary lymph node metastasis, adjuvant endocrine therapy plus or minus chemotherapy should be considered[11].

Fig-3: Immunohistochemistry showing ER-nuclear Positivity, PR-nuclear Negativity, Her2neu-membrane Positivity, NSE-Cytoplasmic Positivity and Chromogranin-Cytoplasmic Positivity
REFERENCES


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