Mucinous Cystadenocarcinoma: A Rare Variant of Breast Carcinoma: Case Report

Müsinöz Kistadenokarsinom: Meme Kanserinin Nadir Bir Varyantı

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ABSTRACT A variety of carcinomas of the breast are characterized by production of abundant extracelluler and/or intracelluler mucin. Among these are mucinous (colloid) carcinoma, mucinous cystadenocarcinoma, columnar cell mucinous carcinoma and signet ring cell carcinoma. This carcinoma is usually composed of tall, columnar cells with basally located nuclei and abundant intracytoplasmic mucin and appears either cystic (mucinous cyst adenocarcinoma) or solid (columnar cell mucinous carcinoma). We report a case of progesteron receptor positive mucinous cystadenocarcinoma of the breast in a 49-year-old women. Primary mucinous cystadenocarcinoma is an extremely rare tumor of the breast. Only eight previous cases have been reported. All of them were hormon independent tumors.

Key Words: Breast neoplasms; cystadenocarcinoma, mucinous

ÖZET Memede bir grup kanser bol ekstrasellüler ve/veya intrasellüler müsin yapımı ile karakterlidir. Bunlar müsinöz (kolloid) karsinom, müsinöz kistadenokarsinom, kolumnar hücreli müsinöz ve taşlı yüzük hücreli karsinomdur. Bu kanser genellikle bazal nücleuslu, uzun kolumnar hücrelerden oluştuğu ve bol intrasitoplazmik müsin içerdiği için, kistik (müsinöz kistadenokarsinom) veya solid (kolumnar hücreli karsinom) şeklinde görülür. Kırk dokuz yaşında kadın hastada, memede progesteron reseptörü pozitif bir müsinöz kistadenokarsinom sunuyoruz. Memenin primer müsinöz kistadenokarsinomu son derece nadirdir. Bu güne kadar sadece sekiz vaka yayınlanmıştır ve bu yayınların hepsi hormon bağımsız tümörlerdir.

Anahtar Kelimeler: Meme kanseri; müsinöz kistadenokarsinom

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There are a variety of breast tumors with obvious mucin production. Mucinous carcinoma is the most common type. Mucinous cystadenocarcinoma (MCA) is a rare form, first described by Koenig and Tavassoli in 1998.[1] This unusal variant of primary breast carcinoma bears a stringing resemblance to MCA's of ovary and pancreas.[2]

Only eight primary MCA's of the breast have been reported previously. All of them were hormon independent tumors.[3] We wish to document an additional and hormon dependent case. The informed consent of the patient was obtained for this case report.

CASE REPORT

A 49-year-old postmenopausal woman referred to Ankara Oncology Educational and Research Hospital because of a malignant epithelial tumor in her left breast. Patient's mammogram obtained before the biopsy showed smoothly bordered 2 cm-diameter mass in the upper left quadrant of the left breast. Ultrasonography of the mass defined a 20 x 15 mm, irregular, hypoechoic lesion. Excisional biopsy was performed in the hospital that the patient was admitted previously. In the initial pathologic report of the patient, macroscopic examination showed a cystic tumor, 3x1.8x1.5 cm in size. Some white solid areas arising from the cyst wall were found. The cyst lumen was comprised of abundant transparent to gelatinous material with many small papillary structures. The final diagnosis in the first center was "malignat epithelial tumor, suspicious for the metastases from gastrointestinal tract". The whole tumor was submitted for histopathological re-evaluation.

Clinical examinations of the case ruled out the possibility of metastases in our hospital. There were no proven lesions in gastrointestinal or genitourinary systems with endoscopic examination, abdominal ultrasonography and CT scans. Tumor markers were within the normal ranges. (Ca 125:24 U/ml; Ca 19-9:5.03 U/ml). No palpable axillary lymph node was noticed. There were no suspicious metastases in the axillary lymph nodes by ultrasongraphy.

Re-excision and axillary lymph node sampling was performed.There were neither residuel tumor nor axillary metastases. The parafin blocks of the patient were also consulted. In hematoxylin-.eosin stained slides, there were dominant cystic lesions which hade many microcysts and small papillary structures with delicate fibrous cores. These were extracellular eosinophilic material. Lining epithelial cells were tall columnar and their cytoplasms were vacuolated. Some of these were similar to goblet cells (Figure 1, 2). Epithelial cells showed nuclear pleomorphism, ranging from minimal to moderate atypia. Mitotic figures were rare. There was no ductal insitu carcinoma (DCIS) inside or near the tumor or surgical margins, so that the surgical margins were tumor-free.

Histochemically, there was abundant mucin in the cytoplasm of the columnar cells and the endocellular areas were stained with periodic acid-Schiff (PAS), with or without diastase digestion.

Immunohistochemically, there were strong positivity for cytokeratin 7 (CK7) (Neomarker's,dilution 1/100), mammoglobulin (Dako, dilution 1/100) and progesterone receptor (PR) (50% and moderate severity) (Figure 3). Cytokeratin 20 (CK20) (Immunovision, dilution 1/100), estrogen receptor (ER)(Neomarker's, dilution 1/200) and c –erbB-2 (Score 1) were negative (Neomarker's, dilution 1/800 for IHC and Ventana silver insitu hybridization for insitu hybridization) (Figure 3).

DISCUSSION

Breast carcinoma comprising of tall columnar cells with nuclei located at the base of the cell and abundant intra and extracytoplasmic mucin with cystic appearence is extremely rare.[4]

A review of the literature, revealed eight previous reports of breast MCAs.[1,2,5,6] The ages of the patients' at the diagnosis ranged from 49 to 96 years, with a mean of 68 years. Our case was a 49 year- postmenopusal woman and her age was similar to other patients reported in the literature. Most of the reported cases were postmenopausal.[1,2,5]

Because of the rarity of this tumor, the possibility of metastases from other organs is a subject of debate in the literature.[1,6] Metastatic mucinous carcinoma, particularly originating from the ovary or pancreas, should be ruled out before primary mucinous cyst adenocarcinoma of the breast is diagnosed.[7] Breast MCAs are CK 7 (+) and CK 20 (-) while both markers are positive in ovarian and pancreatic mucinous adenocarcinoma. In our case, CK 7 was positive and CK 20 was negative, and the carefuly examinations of the gastrointestinal and genitourinary systems did not show any primary malignancies. Additionally, mammoglobulin was strongly positive in our case. Since mammoglobu-

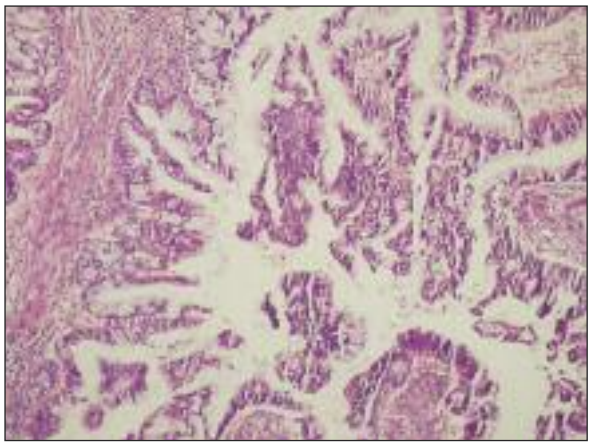

FIGURE 1: Cyst wall consisted of tall columnar, mucinous epithelial cells showing papillary projections (H&Ex40)

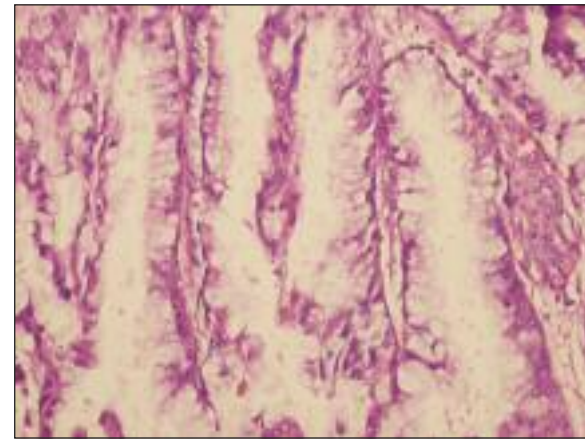

FIGURE 2: Epithelial tumoral cells show abundant intracytoplasmic mucin with basally located nucleus (H&Ex100).

lin is a spesific marker for the breast cancer, this tumor should primarly arise from the breast.[8]

According to Koenig and Tavassoli, the tumor cells are negative for ER-alpha and PR.[1] Houna et al. also reported that tumor cells were negative for ER-alpha and PR, suggesting that MCA's of the breast develop independentl of estrogenic stimulation.[2,9] Similar to the previuos reports ER was negative in our case, but in contrast, we found that PR was positive and because of this we thought this tumor was hormon dependent.

It is better to observe ductal carcinoma insitu (DCIS) areas in and out of the tumor mass. Of the eight previously reported cases, one case was associated with adjacent foci of DCIS, mucinous cystadenocarcinoma in situ and invasive ductal carcinoma.[8] Another case was associated with adjacent DCIS and a focus of ordinary mucinous carcinoma.[6] Two cases had foci of mucinous cystadenocarcinoma in situ and one had DCIS adjacent to the tumor.[1] Three cases, similar to the one presented here showed no association with DCIS.[1,3]

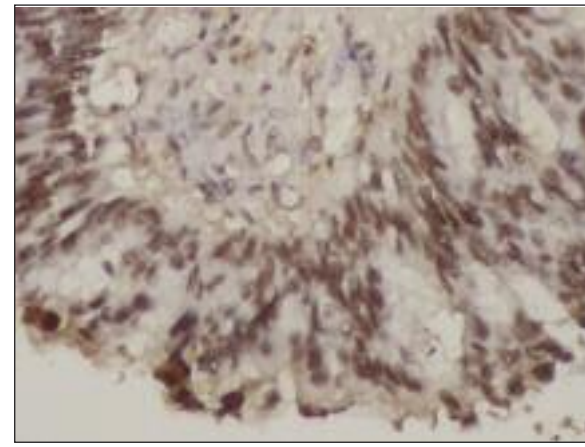

FIGURE 3: Immunohistochemically, epithelial tumoral cells were positive with PR(x200).

Because MCA is an extremely rare variant of breast carcinoma, some cases may have been misclasified as other breast carcinomas. We want to present a case who was hormon dependent in contrast to previously reported cases. This was the first PR positive case. We do not know how the positivity of PR will affect the prognosis, longer follow-up duration and more cases needed to be evaluated to have more information on this issue.

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