

SHORT COMMUNICATION

Pure mucinous carcinoma of the breast: a single center experience

E. Dogan, S. Aksoy, O. Dizdar, C. Arslan, D.S. Dede, Y. Ozisik, K. Altundag

Department of Medical Oncology, Hacettepe University Institute of Oncology, Ankara, Turkey

Summary

Purpose: Mucinous breast carcinoma is rare subtype of breast cancer. Histopathologically, it is classified into two forms, pure and mixed type. It recurs late, metastasis to axillary lymph nodes is less common and is more hormone receptor positive. We herein present the data of our patients with pure mucinous breast cancer (PMBC) treated in our institution.

Methods: Among 1211 breast cancer patients with breast cancer diagnosed and treated in Hacettepe University Institute of Oncology, 20 patients (1.6%) with PMBC (defined as having mucinous component of more than 90%) were identified. Patient demographics, tumor characteristics and patient outcomes were assessed retrospectively.

Results: The median age at diagnosis was 52.5 years (range 27-80). The majority of the patients presented with stage II disease (n=15; 75%). One of 20 patients recurred with bone metastasis 50 months after diagnosis. Median follow-up was 39 months (range 3-137). Estrogen receptors (ER) were positive in 16 (80%) patients and HER-2 positive in one (5%). Twenty-five percent of the patients had positive axillary nodes.

Conclusion: PMBC is a rare entity with favorable prognosis. Lymph node metastasis is rarely seen even in large-sized tumors.

Key words: breast cancer, clinical, pathological, pure mucinous

Introduction

Mucinous carcinoma of the breast is one of the uncommon subtypes of breast cancer which is also historically called colloid or gelatinous carcinoma of the breast and makes up 1-6% of all breast cancers [1,2]. It is an invasive type of adenocarcinoma which is characterized by abundant accumulation of extracellular and/or intracellular epithelial mucus secreted by tumor cells. The mucinous component of the tumor must be >50% [3]. Histopathologically, mucinous carcinoma is classified into two subtypes, pure and mixed [3]. It is known that mixed mucinous carcinoma has more aggressive behavior than the pure form, so it is important to differentiate the two forms regarding their different prognosis.

Methods

Among 1211 breast cancer patients with breast cancer diagnosed and treated in Hacettepe University

Institute of Oncology, 20 patients (1.6%) with PMBC (defined as having mucinous component > 90%) were identified. The patients were retrospectively evaluated regarding demographics, tumor characteristics (biologic markers, tumor size, grade, stage and nodal status) and outcomes.

Results

Patient characteristics are shown in Table 1. The median age at diagnosis was 52.5 years (range 27-80). All of the patients were women, of which 55% were postmenopausal, 40% premenopausal, and 5% perimenopausal. The tumors were right-sided in 40% (8 of 20) and left-sided in 60% (12 of 20) of the patients.

Mastectomy was performed in 15 patients (75%) and the remaining 5 (25%) patients underwent breast-conserving surgery. Eighteen patients (90%) underwent axillary lymph node dissection. Of these 25% (5 of 20) had positive nodes. Two patients had stage I disease, 9

Table 1. Clinical and pathological characteristics of the PMBC patients

Characteristics	n (%)
Patients	20 (100)
Age, years, median (range)	52.0 (27-80)
Grade	
I	8 (40)
II	7 (35)
III	0 (0)
Unknown	5 (25)
ER	
Negative	3 (15)
Positive	16 (80)
Unknown	1 (5)
PR	
Negative	6 (30)
Positive	7 (65)
Unknown	1 (5)
HER-2	
Negative	13 (65)
Positive	1 (5)
Unknown	6 (30)
Stage	
Localized	17 (85)
Advanced	3 (15)
Tumor size	
T1	2 (10)
T2	14 (70)
T3	4 (20)
Nodal status	
N0	13 (65)
N1	3 (15)
N2	1 (5)
N3	1 (5)
Unknown	2 (10)
Adjuvant radiotherapy	
Yes	11 (55)
No	9 (45)
Adjuvant chemotherapy	
Yes	10 (50)
No	10 (50)
Adjuvant hormonotherapy	
Yes	17 (85)
No	3 (15)

PMBC: pure mucinous carcinoma of the breast, ER: estrogen receptor, PR: progesterone receptor

stage IIA, 6 stage IIB, 1 stage IIIA, and 2 patients had stage IIIC disease. Ten patients received adjuvant chemotherapy. Sixteen patients were ER and/or progesterone receptor (PR) positive and received endocrine therapy. The majority of the patients (65%) were HER-2 negative. The median follow-up was 39 months (range 3-137). Only one patient relapsed with bone metastasis 50 months after the initial diagnosis.

Discussion

Invasive ductal carcinoma is the most common

histological subtype of breast cancer and comprises approximately 80% of the cases. The remaining 20% of the cases are special types of invasive carcinomas. Invasive lobular carcinoma (8%) and mixed type (ductal/lobular 7%) are the other common histologies [4].

Mucinous carcinoma of the breast is one of the uncommon special types of breast cancer. Histopathologically, it is classified into two forms, pure and mixed type [5]. PMBC is a rarely seen tumor which represents 1-4% of all breast cancers [6]. In our institute, the incidence was 1.6%. The low incidence is most probably due to in-depth histopathological evaluation criteria of our center. The classification of mucinous carcinoma as pure or mixed type is important because PMBCs have a much better prognosis than mixed mucinous carcinomas.

Tumor size was reported to have no prognostic significance on survival because mucin forms the best part of tumor volume and the cellular component is less [7,8]. Therefore, tumors can appear bigger because of mucin component accumulation. Most of the tumors in our patients were > 2 cm. Komenaka and coworkers stated that the majority of patients presented with palpable mass, a reflection of delayed presentation. They also emphasized that less than one fifth of the mucinous lesions had microcalcifications [7]. André et al. [1] also reported that one third of the mucinous lesions had microcalcifications. Probably the large amount of mucin and less microcalcifications in the lesion prevent early diagnosis via mammography. Furthermore, the most common mammographic appearance in mucinous carcinoma is a lesion with well-circumscribed margins, resulting in misdiagnosis as a benign lesion [9].

Lymph node metastasis is an important prognostic factor for breast cancer. PMBC may be associated with a low risk of axillary node metastases. In the present study, 5 patients (25%) had lymph node metastasis, which was consistent with literature [8,10]. Komenaka et al. found a low rate of nodal metastasis; 7 of 50 patients (14%) had nodal metastasis and lymph node positivity was the only predictor of disease-specific survival [7]. Therefore, if the diagnosis is PMBC, breast-conserving surgery without axillary dissection might be the most appropriate treatment modality. PMBCs mostly express ER and PR receptors. The high rate of hormone receptor expression suggests indolent behavior and good prognosis [7,11]. In our study, 80% of the patients were hormone receptor positive.

Mucinous carcinomas can also be classified by their nuclear grade which also is another significant prognostic factor. Predominantly PMBCs are low grade tumors and are associated with good prognosis. In our study, many patients (75%) had low grade (grade I and II) carcinoma, in concordance with previous studies [11,12].

Many authors state that PMBCs are indolent tumors and have better survival rates than mixed tumors because of late recurrence pattern. The 10- year survival rate among patients with PMBC is 87-90.4% and among patients with mixed tumors it is 54-66% [13,14]. Komenaka et al. reported that the median survival in their patient was approximately 11 years [7].

PMBC is an indolent, slow growing, mucin containing carcinoma and has favorable prognosis. This tumor typically presents as a large tumor, more commonly in elderly patients, and nodal metastasis is the most significant prognostic factor for overall survival.

References

1. André S, Cunha F, Bernardo M, Meneses e Sousa J, Cortez F, Soares J. Mucinous carcinoma of the breast: a pathologic study of 82 cases. *J Surg Oncol* 1995; 58: 162-167.
2. Silverberg SG, Kay S, Chitale AR, Levitt SH. Colloid carcinoma of the breast. *Am J Clin Pathol* 1971; 55: 355-363.
3. Yerushalmi R, Hayes MM, Gelmon KA. Breast carcinoma-rare types: review of the literature. *Ann Oncol* 2009; 20: 1763-1770.
4. Conzen SD, Grushko TA, Olopade OI. Cancer of the breast. Ch 43, pp 1576-1654. In: DeVita VT, Hellman S, Rosenberg SA (Eds). *Cancer: Principles & Practice of Oncology*. Lippincott Williams & Wilkins, Philadelphia, PA, 2008.
5. Lee BJ, Hauser H, Pack GT. Gelatinous carcinoma of the breast. *Surg Gynecol Obstet* 1934; 59: 841-857.
6. Barkley CR, Ligibel JA, Wong JS, Lipsitz S, Smith BL, Golsan M. Mucinous breast carcinoma: a large contemporary series. *Am J Surg* 2008; 196: 549-551.
7. Komenaka IK, El-Tamer MB, Troxel A et al. Pure mucinous carcinoma of the breast. *Am J Surg* 2004; 187: 528-532.
8. Avisar E, Khan MA, Axelrod D, Oza K. Pure mucinous carcinoma of the breast: a clinicopathologic correlation study. *Ann Surg Oncol* 1998; 5: 447-451.
9. Wilson TE, Helvie MA, Oberman HA, Joynt LK. Pure and mixed mucinous carcinoma of the breast: pathologic basis for differences in mammographic appearance. *AJR Am J Roentgenol* 1995; 165: 285-289.
10. Paramo JC, Wilson C, Velarde D, Giraldo J, Poppiti RJ, Mesko TW. Pure mucinous carcinoma of the breast: is axillary staging necessary? *Ann Surg Oncol* 2002; 9: 161-164.
11. Bal A, Joshi K, Sharma SC, Das A, Verma A, Wig JD. Prognostic significance of micropapillary pattern in pure mucinous carcinoma of the breast. *Int J Surg Pathol* 2008; 16: 251-256.
12. Tse GM, Ma TK, Chu WC, Lam WW, Poon CS, Chan WC. Neuroendocrine differentiation in pure type mammary mucinous carcinoma is associated with favorable histologic and immunohistochemical parameters. *Mod Pathol* 2004; 17: 568-572.
13. Komaki K, Sakamoto G, Sugano H, Morimoto T, Monden Y. Mucinous carcinoma of the breast in Japan. A prognostic analysis based on morphologic features. *Cancer* 1988; 61: 989-996.
14. Fentiman IS, Millis RR, Smith P, Ellul JP, Lampejo O. Mucoid breast carcinomas: histology and prognosis. *Br J Cancer* 1997; 75: 1061-1065.