Neuroendocrine carcinoma of the breast: a case report and review of the literature

Kemal Eyvaz¹, Arsenal Sezgin Alikanoğlu², Evrim Hamamcı Özak¹, Murat Kazım Kazan¹, Tuğrul Çakır¹

¹Department of General Surgery, University of Health Sciences, Antalya Training and Research Hospital, Antalya, Turkey; ²Department of Medical Pathology, University of Health Sciences, Antalya Training and Research Hospital, Antalya, Turkey

ABSTRACT

Objectives: Neuroendocrine carcinoma (NEC) is a very rare condition among other types of invasive cancer of breast. Whole-body screening should be performed in order to detect any metastatic or primary disease localization. A 58-year-old patient admitted to hospital with palpable mass on the right retro areolar region and with bloody nipple discharge. Due to axillary positivity with a diagnosis of neuroendocrine carcinoma neoadjuvant chemotherapy was performed. The pathological stage was reported T2N2 with a 2 cm tumor and 5 of 11 lymph node positivity after modified radical mastectomy following neoadjuvant therapy. Tumor cells were stained with positive for neuroendocrine markers. She received adjuvant hormonal treatment with aromatase inhibitors and on regular follow-up with a free of disease to date. The neuroendocrine tumor of the breast is a diagnosis of exclusion. Primary or metastasis distinction is compulsory for the planning of appropriate treatment. There is still a debate on how neuroendocrine differentiation affects the clinical outcome.

Keywords: Neuroendocrine carcinoma, breast cancer, immunohistochemistry

The term neuroendocrine tumor was first described by Cubilla et al. [1] in the late 70’s which reports ‘neurosecretory’ granules were seen under electron microscopy. Neuroendocrine tumors (NET) arise from the neuroendocrine cells. The gastrointestinal tract and the lungs are the most commonly observed locations of neuroendocrine tumors [2]. Pure neuroendocrine tumors of breast are very rare among all types of breast cancer (0.1% of all breast cancers), but neuroendocrine cells can be detected much more in other types of breast cancers [3, 4]. Here we would like to present a 58-year-old patient’s management of treatment and review of the literature with a diagnosis of a neuroendocrine differentiating tumor of the breast.

CASE PRESENTATION

A 58-year-old generally in good health condition woman was admitted to policlinic with a palpable mass on her right breast. The patient has only mild hypertension. A palpable 3×2 cm mass examined on the right upper retro areolar region. Ultrasonography and bilateral mammography performed. A BIRADS category 4c was reported by both sonographic and mammographic examination. Enlarged lymph nodes were detected in the axilla. A tru-cut biopsy was performed from the palpable mass under sonographic guidance. Pathology has resulted in a suspicious neuroendocrine tumor whether primary or metastatic. Immunohisto-

Received: February 13, 2021; Accepted: March 25, 2021; Published Online: January 25, 2022


Address for correspondence: Kemal Eyvaz, MD., University of Health Sciences, Antalya Training and Research Hospital, Department of General Surgery, Kazım Karabekir Cad., Varlık Mah., 07100 Muratpaşa, Antalya, Turkey. E-mail: drkemal07@gmail.com, Tel (Mobil): +90 506 388 31 60

©Copyright © 2022 by Prusa Medical Publishing
Available at http://dergipark.org.tr/eurj

The European Research Journal • Volume 8 • Issue 4 • July 2022
Neuroendocrine carcinoma of the breast

There was no additional focus observed except axillary involvement. Due to the locally advanced stage, neoadjuvant chemotherapy was planned by the oncologic tumor board.

After neoadjuvant therapy control positron emission tomography revealed partial response and persistent axillary status. Modified radical mastectomy performed to avoid locoregional relapse. The final pathology result was invasive carcinoma of the breast with neuroendocrine differentiation. Five positive lymph nodes out of 15 were detected. Pathologic grade was T1N2Mx. Adjuvant hormonotherapy with aromatase inhibitors and taxane based treatment was given. She is still on regular follow-up at our clinic.

DISCUSSION

Neuroendocrine carcinoma (NEC) of the breast is a rare condition that represents 0.1% of breast cancers and less than 1% of other neuroendocrine tumors. Mainly, neuroendocrine tumors observed in the gastrointestinal system and pulmonary system [3]. It was also reported in a retrospective study reported by Wang et al. [5] a total of more than 380 thousand invasive carcinomas of the breast only 142 of them were diagnosed as neuroendocrine carcinoma which refers to 0.1% of invasive carcinoma of the breast, and also was observed relatively older ages (mean age was 64). NEC of breast could also be observed younger ages and among men [6, 7]. Palpable mass, bloody nipple discharge can be the first signs observed on admission to hospital [7]. Almost there were no specific or pathognomonic sign presents in breast ultrasonography and mammography but Park et al. mentioned that some radiological characteristics such as high-density round, oval, or lobular noncalcified mass with nonspiculated margins suggest neuroendocrine tumor of the breast [8, 9]. Chromogranin and synaptophysin and neuron-specific enolase are known as specific immunohistochemical markers of neuroendocrine differentiation. They could be stained with argentaffin histochemically and neurosecretory granules of the tumor could be observed under electron microscopy. NEC diagnosis could be made whether these marker occupy more than 50% of tumor cells [10-12].

Estrogen receptor (ER) and progestrone receptor (PR) positivity and human epidermal growth factor re-
ceptor 2 (HER-2) negativity mostly observed in neuroendocrine tumors of breast [13-15]. Positive ER status may not contribute positive prognostic benefit as it does in other types of invasive carcinomas of breast [5].

Ki 67 protein is a proliferation antigen, which is present in the different phases of the cell cycle and accepted as a poor prognostic factor and high Ki 67 index observed in poorly differentiated tumors [15, 16]. Although neuroendocrine carcinoma is a rare condition observed in the breast, routine systemic work-up should be performed to rule out any other primary or metastatic focus with positron emission tomography and bone scintigraphy [17, 18].

Surgical approach to neuroendocrine carcinoma of the breast is not different in any other type of invasive carcinoma of breast. The location of the tumor and the clinical stage are the main determinants of the surgical procedure [19, 20]. Neo-adjuvant chemotherapy is indicated in case of locally advanced disease or malignity which is not suitable for surgery. Adjuvant therapy regimens including anthracyclines and/or taxanes, hormonal and HER-2 status should be evaluated for endocrine and anti HER-2 treatment [19].

CONCLUSION

In conclusion, the neuroendocrine tumor of the breast is a diagnosis of exclusion. A biopsy followed by appropriate immunohistochemical staining could help for diagnosis. Primary or metastasis distinction is compulsory for the planning of appropriate treatment. There is still a debate on how neuroendocrine differentiation affects the clinical outcome.

Authors’ Contribution

Study Conception: KG; Study Design: KG; Supervision: KG; Funding: KG; Materials: KG; Data Collection and/or Processing: KG; Statistical Analysis and/or Data Interpretation: KG; Literature Review: KG; Manuscript Preparation: KG and Critical Review: KG.

Conflict of interest

The authors disclosed no conflict of interest during the preparation or publication of this manuscript.

Financing

The authors disclosed that they did not receive any grant during conduction or writing of this study.

Informed Consent

Written informed consent was obtained from the patient for publication of this case and any accompanying images or data.
REFERENCES