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Case Report

Primary Neuroendocrine Breast Carcinoma: A Case Report

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Abstract

This case report describes a rare presentation of primary neuroendocrine breast carcinoma (NEBC) in an 86-year-old woman presenting with left breast areolar swelling. Diagnostic workup included breast ultrasound revealing a well-circumscribed hypoechoic mass, mammography showing a dense left areolar opacity, and core-needle biopsy confirming a poorly differentiated neuroendocrine carcinoma. Immunohistochemical staining was positive for synaptophysin and chromogranin. CT scan ruled out a metastatic origin. The patient underwent a modified radical mastectomy followed by adjuvant chemotherapy and radiotherapy. Six months later, the patient remains disease-free. This case highlights the diagnostic challenges of NEBC, emphasizing the need for immunohistochemical analysis to confirm the neuroendocrine nature of the tumor. It also underscores the importance of distinguishing primary NEBC from metastatic disease to guide treatment strategies.

Keywords: Neuroendocrine; Carcinoma; Breast; Immunohistochemistry

Introduction

Neuroendocrine tumors (NETs) are diverse neoplasms arising from neuroendocrine cells through the body. The gastrointestinal and respiratory tracts are the most common primary sites [1,2].

Primary Neuroendocrine breast carcinoma (NEBC) is a unique end exceedingly rare entity constituting less than 1% of all neuroendocrine tumors and less than 0.1% of all breast cancers. These tumors typically present in women between the ages of sixty and seventy [3,4].

The true incidence of NEBC remains unclear due to the limited number of reported cases and a lack of standardized diagnostic criteria. Reported incidence rates vary widely, ranging from 0.1% to 18% [4,5].

Furthermore, immunohistochemical markers for neuroendocrine tumors are not routinely employed during breast cancer diagnosis [6,7].

Clinically, these tumors often demonstrate an aggressive course and tend to have a higher risk for local and distant recurrence compared to other types of invasive breast carcinomas [4,8].

Due to their rarity, current knowledge regarding NEBC primarily stems from sporadic case reports or small retrospective studies, leading to ongoing debate in the literature concerning prevalence, prognosis, and optimal treatment strategies [4].

This case report details the sonographic, mammographic, CT scan, and histopathological findings of NEBC in an 86-year-old woman presenting with left breast areolar swelling.

Case Report

An 86-year-old woman with no significant past medical history presented with a progressively enlarging swelling in the left breast areola for several months. She denied any other associated symptoms, such as nipple discharge, pain, or skin changes.

Physical examination revealed a discrete area of asymmetry rather than a well-defined mass was palpated in the left breast areola. No palpable lymphadenopathy was noted in the axillary or supraclavicular regions.

Due to the clinical signs, the patient underwent complementary breast imaging studies.

The breast ultrasound (Figure 1) revealed A well-circumscribed, left peri-areolar subcutaneous node with slightly lobulated margins measuring 32 x 16 mm. This lymph node appeared homogeneously hypoechoic without posterior acoustic attenuation and demonstrated vascularity on Doppler interrogation. Bilateral axillary lymph nodes of sub-centimeter size with preserved architecture were also noted.

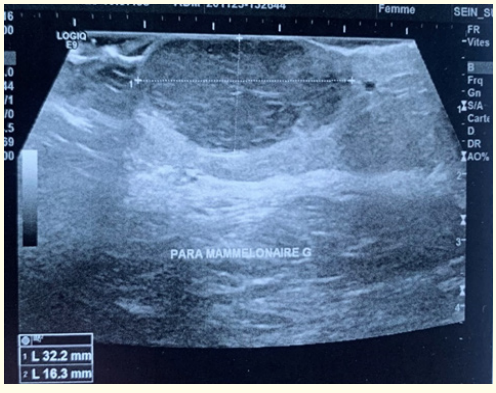

Figure 1: Ultrasound showing the mass.

Bilateral breast mammography (Figure 2) showed a dense left areolar opacity with circumscribed margins. No suspicious microcalcifications were identified. Benign bilateral vascular calcifications were present without evidence of axillary lymph node enlargement. Breast parenchyma density was normal with no architectural distortion.

To further characterize the identified mass, a core-needle biopsy was performed for pathological evaluation. Histological examination revealed an undifferentiated malignant tumor proliferation infiltrating the fibrous tissue. The morphological appearance was primarily suggestive of a carcinomatous origin.

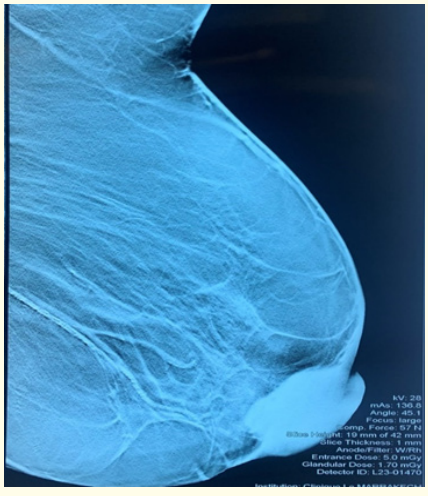

Figure 2: Mass appearance on mammography.

Immunohistochemical analysis was subsequently performed, revealing a poorly differentiated neuroendocrine carcinoma. Positive staining for synaptophysin and chromogranin confirmed the neuroendocrine nature of the tumor. Hormone receptor (ER, PR) and HER2 testing were negative. The Ki-67 proliferation index was 50%.

Before retaining the primary character of the carcinoma, it was crucial to rule out a metastatic origin from another primary site. To achieve this hematologic, biochemical evaluation and Computed Tomography (CT) Scan of Chest and Abdomen. All of the above came up negative thus the primary nature of the neuroendocrine carcinoma in the left breast areola was established.

The patient underwent a modified radical mastectomy (Patey procedure). Histopathological examination of the resected breast tissue revealed a 4 cm tumor in greatest dimension. The tumor was identified as a poorly differentiated carcinoma of neuroendocrine origin, infiltrating the skin but lacking an intraductal component or vascular emboli. The nipple remained uninvolved, surgical margins were sane of tumor involvement and no lymph node metastases were identified.

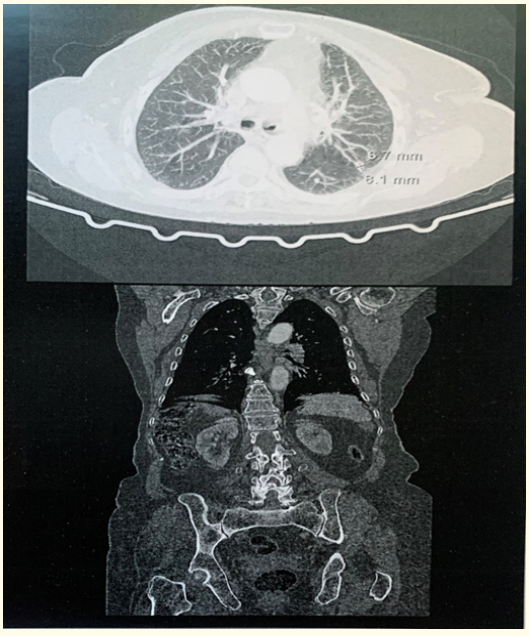

Figure 3: Full -body Computed Tomography (CT) showing absence of secondary lesion.

The patient subsequently received adjuvant chemotherapy and radiotherapy. Six months after the treatment, the patient is well with no evidence of disease recurrence.

Discussion

The diagnosis of primary NEBC presents a significant challenge. Clinical presentation can mimic other breast cancers, in addition to Neuroendocrine cancer-associated syndromes. These occur when the tumor secretes hormones into the bloodstream, causing symptoms such as flushing, diarrhea, abdominal cramping, wheezing, palpitations and peripheral oedema [1,9].

Imaging findings for NEBC are often non-specific describing a high-density mass with predominantly speculated or lobulated margins with no surrounding microcalcifications on mammography [4,10].

As for The sonographic appearance, NEBCs has been described as a hypoechoic or heterogeneous mass with irregular shape or micro-lobulated margins and normal sound transmission [10,11].

Definitive diagnosis of NEBCs requires a needle biopsy. Immunohistochemical staining is then performed to confirm the presence of neuroendocrine markers such as chromogranin A, synaptophysin, and neuron-specific enolase (NSE) [10,12].

The distinction between primary and metastatic neuroendocrine tumors of the breast is important since the two necessitate different treatment strategies [13,14].

A full-body imaging evaluation, including CT and PET scans, is crucial to identify the origin of the cancer, confirm its primary character and search for potential metastasis [1].

While there is no established standard treatment protocol for NEBC, the therapeutic approach is often similar to that for other invasive breast cancers, with surgery serving as the mainstay of treatment. The type of surgery performed is determined by the tumor location and clinical stage. Options include lumpectomy with sentinel lymph node biopsy for early-stage tumors. For more advanced stages, mastectomy with sentinel lymph node biopsy or modified radical mastectomy may be necessary, particularly if sentinel nodes contain metastatic disease [1,15,16].

Adjuvant chemotherapy may be indicated for patients with NEBC and a high risk of recurrence based on established prognostic factors. Neoadjuvant chemotherapy can be considered for patients with locally advanced disease not amenable to upfront surgery. Additionally, chemotherapy may be used for downstaging large tumors to facilitate breast-conserving surgery. The specific chemotherapeutic regimen selected will depend on individual patient factors and tumor characteristics [1,16].

NEBC exhibits a propensity for late recurrence, with metastases potentially occurring many years after initial treatment. Therefore, long-term follow-up is mandatory for patients with NEBC. The most common sites of metastatic disease include the liver, bones, lungs, soft tissues, pleura, brain, mediastinal lymph nodes, adrenal glands, ovaries, and pancreas [15,17].

Conclusion

In conclusion, this case report underscores the diagnostic challenges posed by primary neuroendocrine breast carcinoma (NEBC) due to its rarity. The clinical presentation may be similar to other prevalent breast cancers, necessitating a thorough immunohistochemical workup for definitive diagnosis. Given the limited knowledge of NEBC, the optimal treatment strategy remains undefined.

Currently, management of this disease still follows a similar approach as for conventional breast cancer, with surgery as the mainstay. Further research efforts are warranted to establish targeted therapies and ultimately improving clinical outcomes for this unique and aggressive tumor.

Conflict of Interest Statement

The authors declare no conflict of interest.

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