PAMJ Clinical Medicine
Pan African Medical Journals

Case report

Primary male neuroendocrine breast carcinoma: a case report and literature review

Leila Afani[1,&], Adjade Ganiou[1], Hind rachidi[2], Belbaraka Rizlane[1]

[1]Department of Medical Oncology, Center Mohammed VI University, Marrakech, Maroc, [2]Department of Pathology, ARRAZI Hospital, Center, Mohammed VI University Hospital, Marrakech, Maroc

[&]Corresponding author: Leila Afani, Department of Medical Oncology, Center Mohammed VI University, Marrakech, Maroc

Received: 28 Feb 2020 - Accepted: 14 Apr 2020 - Published: 14 May 2020

Domain: Oncology

Keywords: Neuroendocrine carcinoma, metastases, male breast cancer

Abstract

Breast neuroendocrine carcinoma is a rare tumor. It represents about 2 to 5% of breast cancers. The majority of cases have been described in women. Only a few cases have been reported in men. Due to the rarity of cases, there is no consensus on therapeutic modalities. We report a case of neuroendocrine carcinoma in a man with a review of the literature on histological features, clinical presentation, treatments, and prognosis.

Case report | Volume 3, Article 13, 14 May 2020 | 10.11604/pamj-cm.2020.3.13.22037

Available online at: https://www.clinical-medicine.panafrican-med-journal.com/content/article/3/13/full

Introduction

Primary neuroendocrine breast carcinoma is an uncommon disease. It represents about 2 to 5% of breast cancers [1]. It is recognized as a separate entity in 2003. The diagnostic criteria include neuroendocrine architecture and cytology combined with an expression of neuroendocrine markers in more than 50% of tumor cells. In 2012, the WHO classification divided neuroendocrine tumors into three groups: well-differentiated neuroendocrine tumors; poorly differentiated neuroendocrine carcinoma/small cell carcinoma invasive breast cancer with neuroendocrine differentiation [2]. The majority of cases have been described in women [3]. Only a few cases have been reported in men [4]. Due to the rarity of cases, there is no consensus on therapeutic modalities. We report a case of primary neuroendocrine carcinoma in a man with a review of the literature on histological features, clinical presentation, treatments, and prognosis.

Patient and observation

This is a case of a 70-year-old male patient, without medical history, who consulted for a left breast mass gradually increasing in volume. Clinical examination objectified a budding and ulcerated mass of 6 x 5 cm (Figure 1, Figure 2) in the left breast without axillary lymphadenopathy. Thoracic abdominopelvic CT scan showed two lung metastases. A biopsy of this mass was performed. Anatomopathological examination revealed a neuroendocrine carcinoma (Figure 3). Immunohistochemistry objectified positive hormone receptors and a negative HER2. This case was discussed at the multidisciplinary concertation meeting. The therapeutic decision was to put the patient on hormone therapy. Therefore, the patient received Tamoxifen as a first-line treatment. Assessment after 12 months of treatment showed stable disease.

Discussion

Primary neuroendocrine carcinoma of the breast is very rare. The largest series of reported cases include 142 cases registered from 2003 to 2009 in the Surveillance, Epidemiology and End Results (SEER) database. The incidence was less than 0.1% of all breast cancers [5]. The incidence of breast cancer in men represents less than 1% of all breast neoplasms [6]. Primary neuroendocrine carcinoma is a very rare variant of breast cancer in men and only a few cases have been reported [4]. It occurs mainly in the 6^{th} and 7^{th} decades with a female predominance [5]. The histogenesis of neuroendocrine breast tumors is still controversial. The theory adopted by WHO 2012 is that the tumor results from early divergent differentiation of breast cancer stem cells into epithelial and neuroendocrine lineages, respectively. The diagnosis of breast neuroendocrine carcinoma is confirmed by immunohistochemistry with neuroendocrine markers based on the diagnostic criteria reported by the WHO classifications [2]. The majority of neuroendocrine tumors express estrogen (ER) and/or progesterone receptors (PR). The expression of Her-2 is rare [5]. There is no difference in the clinical presentation and imaging characteristics of primary breast neuroendocrine carcinoma compared with other types of breast tumors [7]. It has been observed that breast cancers in men are usually high grade [6]. Due to the scarcity of cases, there are no clear recommendations regarding therapeutic modalities. The question remains: should this entity be treated like other breast carcinomas or other neuroendocrine carcinomas?

For localized tumors, surgery remains the main treatment. Postoperative irradiation remains controversial. It can adopt an algorithm similar to that used in invasive ductal breast cancer with certain benefits [3]. However, it seems that in small cell carcinoma, no benefit has been observed with radiotherapy [8]. No consensus on the choice of chemotherapy agents. The combination of Etoposide + Platinum is used in

patients with high ki67 index or small cell poorly differentiated carcinomas. Chemotherapy with Anthracyclines or Taxanes is used for a low ki67 index. Adjuvant endocrine therapy is indicated in patients with a positive hormone receptor [9]. For metastatic tumors, the choice of treatment depends on age, comorbidities, number, and site of metastasis and the biological behavior of the tumors. Chemotherapy is indicated in patients with metastases at multiple sites. The treatment regimens reported in the literature are derived from other metastatic breast cancers and small cell cancers [10]. Due to the high expression of hormone receptors, hormone therapy is an option for patients who do not have a visceral crisis or cannot undergo chemotherapy. Everolimus has been approved by the FDA for the treatment of pancreatic neuroendocrine tumors and hormone-receptor-positive advanced breast cancer in postmenopausal women after treatment failure with aromatase inhibitors. It could show a result in metastatic neuroendocrine breast cancer. Another treatment option is peptide receptor radionucleotide therapy for tumors expressing somatostatin receptors after the failure of conventional therapy [10]. The prognosis of neuroendocrine breast cancer is controversial. It depends on the grade, the ER/PR expression and the stage of the disease [3].

Conclusion

Primary neuroendocrine male breast carcinoma is a very rare entity. No consensus has been established. In localized disease, Surgery remains the main treatment. In metastatic settings, systemic treatment is derived from other metastatic breast cancers. Clinical trials are needed to define the best treatment for this entity.

Competing interests

The authors declare no competing interests.

Authors' contributions

LA contributed to the literature review and also wrote this article. AG helped to the literature research. H.R. performed the histological examination. RB analyzed the literature data. All authors read and approved the final manuscript.

Figures

Figure 1: a budding and ulcerated mass of 6x5cm of the entire left breast

Figure 2: aspect of salt and pepper chromatin (HEx40)

Figure 3: chromagranin A- positive tumor cells

References

1. Tavassoli FA, Devilee P. Pathology and genetics: Tumors of the Breast and Female Genital Organs. WHO Classification of Tumors Series. Lyon: IARC Press. 2003;32-4.

2. Lakhani S, Ellis I, Schnitt S, Tan P, van de Vijver MJ. Tumours Breast. IARC Press. 2012; 62-3.

3. Jurvcic Petra, Kruslin Bozo, Gatalica Zoran, Sanati Souzan, Vranic, Semir. Breast carcinoma with neuroendocrine features: a brief review. EOM. 2016;6:138-45. **Google Scholar**

4. Manupriya S, Aruna G, Suman S, Rashmi K. Primary Neuroendocrine Carcinoma of the Male Breast: a Rare Case Report. JOJ Nurse Health Care. 2017;5(3): 555661.

5. Wang J, Wei B, Albarracin C, Hu J, Abraham S, Wu Y. Invasive neuroendocrine carcinoma of the breast: a population-based study from the surveillance, epidemiology and end results (SEER) database. Biomed central Cancer. 2014;14:147. **PubMed** | **Google Scholar**

6. Ruddy KJ, Winer EP. Male breast cancer: risk factors, biology, diagnosis, treatment, and survivorship. Ann Oncol. 2013;24(6): 1434-43. **PubMed** | **Google Scholar**

7. Shin SJ, DeLellis RA, Ying L, Rosen PP. Small-cell carcinoma of the breast: a clinicopathologic and immunohistochemical study of nine patients. Am J Surg Pathol. 2000;24(9):1231-38. **PubMed** | **Google Scholar**

8. Hare F, Giri S, Patel JK, Hahn A, Martin MG. A population-based analysis of outcomes for small cell carcinoma of the breast by tumor stage and the use of radiation therapy. Springerplus. 2015;4:138. **PubMed** | **Google Scholar**

9. Yesim Yildirim, Sahende Elagoz, Ayhan Koyuncu, Cengiz Aydin, Kursat Karadayi. Managementof neuroendocrine carcinomas of the breast: a rare entity. Oncol Lett. 2011 Sep 1;2(5):887-890. **PubMed** | **Google Scholar**

10. Inno A, Bogina G, Turazza M, Bortesi L, Duranti S, Massocco A *et al*. Neuroendocrine carcinoma of the breast: current evidence and future perspectives. The oncologist. 2016 Jan;21(1):28-32. **PubMed** | **Google Scholar**

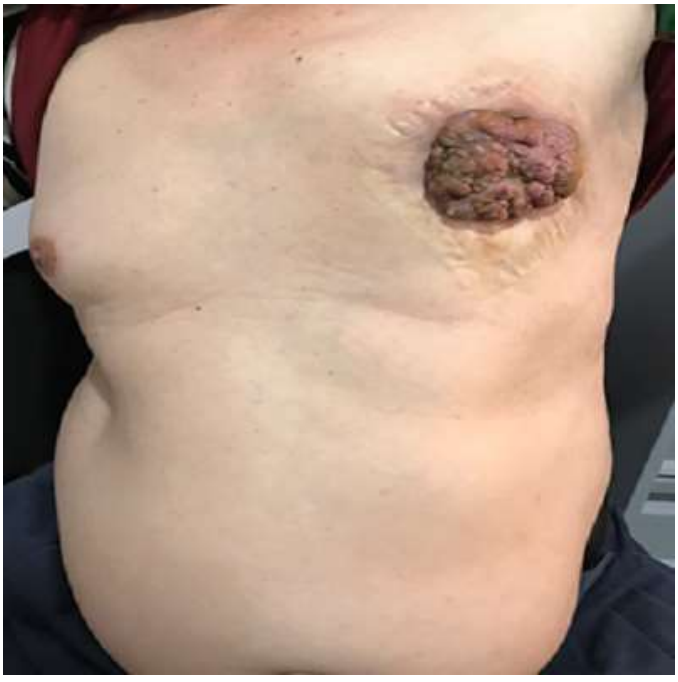

Figure 1: a budding and ulcerated mass of 6x5cm of the entire left breast

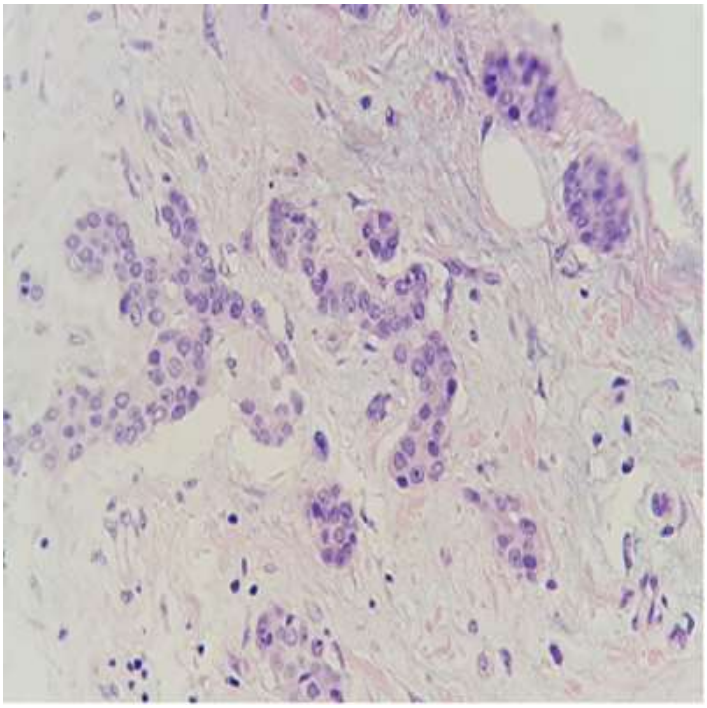

Figure 2: aspect of salt and peper chromatin (HEx40)

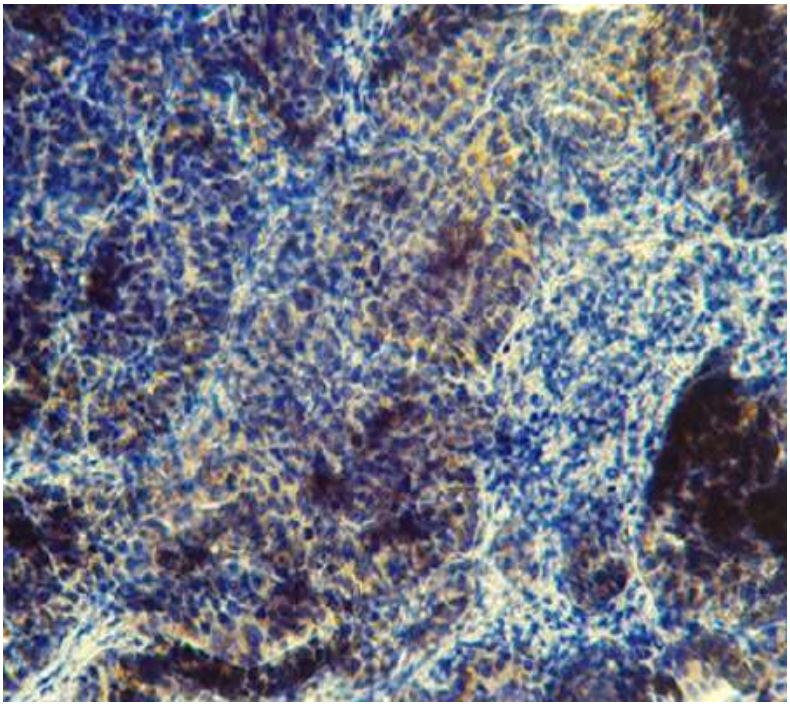

Figure 3: chromagranin A- positive tumor cells