BJCR
Brazilian Journal of Case Reports

PCAA
Publicações Científicas de Acesso Aberto

Case Report

Non-Hodgkin's Lymphoma of the Breast: An Unusual Case Report

Andreia Mandim [1,*], Maria Eduarda Couto [2], Raquel Oliveira [1], Raquel Santos [1]

[1] Department of Internal Medicine, Local Health Unit of Póvoa de Varzim/Vila do Conde, Portugal.
[2] Hematology Unit, Department of Internal Medicine, Local Health Unit of Póvoa de Varzim/Vila do Conde, Portugal.

* Correspondence: andreiamandim@hotmail.com.

Abstract: Primary breast lymphoma is a rare condition accounting for less than 0.5% of all breast malignancies. It typically presents as a solitary mass in women aged 50–60, with clinical and imaging features mimicking breast carcinoma. We present the case of a 60-year-old woman with a right breast nodule diagnosed as marginal zone lymphoma (MZL) by histology. The patient was treated with R-CHOP immunochemotherapy followed by radiotherapy, achieving complete remission with 24 months of sustained follow-up. This case highlights the importance of early histological diagnosis, the occasional diagnostic role of surgery, and the controversies surrounding treatment strategies for breast MZL.

Keywords: Non-Hodgkin lymphoma; Breast; Marginal zone; Immunochemotherapy; Differential diagnosis.

Citation: Mandim A, Couto ME, Oliveira R, Santos R. Non-Hodgkin's Lymphoma of the Breast: An Unusual Case Report. Brazilian Journal of Case Reports. 2026 Jan-Dec;06(1):bjcr 116.

https://doi.org/10.52600/2163-583X.bjcr.2026.6.1.bjcr116

Received: 7 August 2025
Accepted: 1 September 2025
Published: 29 September 2025

1. Introduction

Primary breast lymphoma (PBL) is a rare type of breast malignancy first identified in 1959 by Dobrotina, representing less than 0.5% of all breast cancers. Most of the information on PBL comes from small case series or individual case reports. Most cases are diffuse large B-cell lymphoma (DLBCL), while marginal zone lymphoma (MZL) is less frequent but clinically relevant. Recent reports have highlighted diagnostic challenges and heterogeneity in therapeutic approaches, particularly regarding the role of radiotherapy alone versus combined chemoimmunotherapy [1-3].

2. Case Report

A 60-year-old female, with history of diabetes, hypertension and depression, presented with a right breast mass. Mammography showed a 15.4x10mm hypoechoic nodule. An ultrasound-guided core needle biopsy revealed a lymphoid lesion of undefined subtype. Because the material was insufficient for accurate subclassification, an excisional lumpectomy was performed for diagnostic purposes. Histology revealed polymorphic lymphoid infiltrate with intermediate to large, atypical lymphocytes and Reed-Sternberg–like cells. This raised suspicion of lymphomatous granulomatosis or Hodgkin lymphoma. However, immunohistochemistry showed CD20 positivity, with negativity for CD10, CD30, BCL6, Cyclin D1, MUM1 and EBER, excluding Hodgkin lymphoma and EBV-driven disease, and supporting a final diagnosis of marginal zone lymphoma (MZL).

The patient reported weight loss but no other B symptoms. Laboratory tests showed mild anemia, with normal LDH. Bone marrow biopsy and staging CT scans revealed localized disease (Ann Arbor stage I). After multidisciplinary discussion, she received R-CHOP immunochemotherapy (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine,

Prednisone) for 4 cycles instead of 6, due to treatment-related neutropenic complications and institutional protocol for indolent histology. Consolidation radiotherapy (20Gy x 6 sessions) was administered. She achieved complete remission, sustained at 24-month follow-up.

3. Discussion

This case illustrates the diagnostic and therapeutic challenges of primary breast MZL. Core needle biopsy may fail to provide sufficient architecture for subtype classification, as occurred here, requiring excisional biopsy. While surgery has no role in treatment, it can be occasionally necessary for diagnostic clarification. The presence of Reed-Sternberg–like cells raised differential diagnoses (Hodgkin lymphoma, lymphomatoid granulomatosis), but the immunohistochemical profile supported MZL. This highlights the importance of thorough immunophenotyping in avoiding misdiagnosis.

Therapeutically, localized MZL is indolent, and radiotherapy alone is often considered curative. However, in this case, systemic treatment was chosen given the atypical histopathological features and diagnostic uncertainty at presentation. R-CHOP, although more commonly used for aggressive lymphomas, was administered in four cycles due to patient comorbidities, institutional practice, and treatment toxicity. Radiotherapy consolidation was added in line with some reported strategies. This approach achieved durable remission, consistent with outcomes described in recent literature for MZL of the breast [2, 4, 5].

Central nervous system (CNS) prophylaxis is not indicated for localized indolent lymphomas and was not considered here. Extensive discussion of CNS prophylaxis, more relevant to DLBCL, was avoided to maintain focus on MZL. Recent literature (2020–2023) supports the heterogeneity of treatment approaches in extranodal MZL, with radiotherapy being standard for localized disease, and systemic therapy reserved for selected situations [3-5]. Long-term monitoring is essential due to the risk of relapses or histological transformation.

4. Conclusion

Primary breast MZL is a rare disease that can mimic breast carcinoma clinically and radiologically. Diagnosis requires histological biopsy with immunophenotyping, and excisional biopsy may be necessary when core samples are inconclusive. Treatment strategies remain controversial; radiotherapy alone is often sufficient, but combined chemoimmunotherapy may be considered in selected cases. This case underscores the need for individualized management and highlights the importance of long-term follow-up given the risk of recurrence or transformation.

Funding: None.

Research Ethics Committee Approval: We declare that the patient approved the study by signing an informed consent form and the study followed the ethical guidelines established by the Declaration of Helsinki.

Acknowledgments: None.

Conflicts of Interest: The authors declare no conflicts of interest.

References

1. Liu Y, Wang J, Li X, et al. Primary breast marginal zone lymphoma: population-based analysis of incidence, treatment, and survival outcomes. Front Oncol. 2020;10:609512. doi:10.3389/fonc.2020.609512.
2. Deantonio L, Mazzola R, Trovò M, et al. Radiation therapy of extranodal marginal zone lymphomas. Ann Lymphoma. 2020;4:11. doi:10.21037/aol-20-37.
3. James S, Patel R, Thompson J, et al. Primary breast marginal zone lymphoma: a rare indolent entity with diagnostic and therapeutic challenges. JPRAS Open. 2022;33:16–20. doi:10.1016/j.jpra.2022.02.007.

4. Walewska R, Smith A, Brown K, et al. British Society for Haematology guidelines for the diagnosis and management of marginal zone lymphomas. Br J Haematol. 2024;205(1):45–63. doi:10.1111/bjh.19064.
5. National Cancer Institute. PDQ® Adult Treatment Editorial Board. Indolent B-cell non-Hodgkin lymphoma treatment (PDQ®): health professional version. Updated 2025. Available from: https://www.cancer.gov/types/lymphoma/hp/indolent-b-cell-lymphoma-treatment-pdq.