

A Diagnostic Surprise in a Breast Lump: Primary T-Cell non-Hodgkin's Lymphoma

Abstract

Unilateral primary breast lymphoma (PBL) is a rare entity usually occurring in elderly women and when present is of B-cell variety. We present a premenopausal female who presented with an asymptomatic breast lump whose initial investigations were inconclusive but for final histopathology and immunohistochemistry that revealed a non-Hodgkin's PBL of T-cell variety that is extremely rare. A detailed review has been presented.

Keywords: Breast, chemotherapy, lymphoma, primary, surgery

Introduction

Primary breast lymphoma (PBL) is a rare form of extranodal lymphoma, defined by the presence of a primary lesion within the breast with or without regional nodal involvement but no other extramammary sites of involvement.^[1] It is seen in elderly women and constitutes only about 0.04%–0.53% of malignant breast neoplasms: 0.38%–0.7% of all non-Hodgkin's and 1.7%–2.2% of extra-nodal non-Hodgkin's lymphoma (NHL) and is bilateral in 5% of women.^[2] B-cell lymphomas constitute more than 80% with diffuse large B-cell variety being the most common (50%) that are CD20 positive.^[3] Breast T-cell lymphomas are extremely infrequent and are reported mainly as isolated cases.^[4] The probable origin of breast lymphoma is mucosa-associated lymphoid tissue or the lymphatic tissue present adjacent to breast ducts and lobules.^[5] Clinical presentation and radiological features mimic breast carcinoma and lead to a diagnostic dilemma. Management requires a multimodality approach with chemotherapy, surgery, and radiotherapy (RT) based on the stage of the disease and histology.^[2] In general, the surgery has no role beyond obtaining a histologic diagnosis to guide definitive therapy. Anthracycline-containing chemotherapy followed by consolidative ipsilateral breast irradiation is standard of care.^[6]

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Case Report

A 45-year-old female presented with complaints of an asymptomatic lump in the left breast of 3 months duration. Examination revealed a 3 cm × 1 cm firm, nodular, nontender, mobile lump in the upper and central quadrant with no axillary lymphadenopathy. Examination of the right breast, axilla, supraclavicular regions, and systemic review was unremarkable. Mammogram revealed a well-defined thick-walled heteroechoic lesion with dense internal echoes at 10–11 o'clock position with surrounding diffuse inflammatory changes and few hypoechoic areas within representing mastitis [Figure 1]. A fine-needle aspiration cytology done was reported to be inconclusive. As the patient did not consent for a trucut biopsy, she was posted for a lumpectomy, frozen section (FS), and proceed to mastectomy, if breast carcinoma. FS of the lump was reported to be suspicious of lymphoma, and hence, mastectomy was deferred. Final histopathological examination revealed extranodal NHL: peripheral T-cell variety. Immunohistochemistry (IHC) showed positive CD3, focally positive CD30, negative CD20, CD79a, PAX5, GATA-3, MUM-1, CD-10, BCL-6, and EBV [Figure 2]. Ki-67 index was 53%. Positron emission tomography scan was done and did not show any residual or systemic disease. Hence, a diagnosis of a NHL: PBL of T-cell variety was arrived at.

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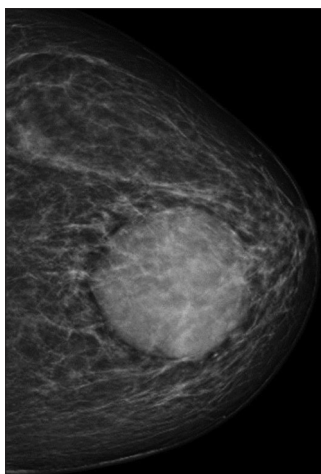


Figure 1: Mammogram showing well-defined heteroechoic lesion in the left breast

She deferred adjuvant treatment and is lost to follow-up. Attempts to contact her have been in vain.

Discussion

PBL is a rare, distinct clinicopathological entity, and a potentially curable disease. The presence of lymphomatous infiltrate in normal breast tissue with neither the presence of concurrent NHL at a different site nor a prior history of the same qualifies as PBL.^[1,4] Wiseman and Liao^[5] have laid down the specific criteria to diagnose PBL that include: (1) breast needs to be the clinical site of presentation, (2) a prior history of lymphoma or an evidence of metastatic disease needs to be ruled out at the time of diagnosis, (3) the pathologic specimen should reveal close association of lymphoma with the breast tissue, and (4) presence of ipsilateral axillary lymphadenopathy in case of concomitant disease.

PBL shows a bimodal peak with respect to age distribution with bilateral disease in younger population and unilateral disease in older patients. Bilateral disease is commonly seen during pregnancy or in the postpartum period due to the influence of tumor growth by hormonal variation.^[2] The most common symptom is a painless rapidly progressive breast lump. Less commonly, patients may also present as diffuse breast enlargement. Ipsilateral axillary lymphadenopathy may be present in 50% of the patients.^[3]

The radiological features remain nonspecific. On mammography, it typically appears as a solitary, circumscribed or indistinctly marginated mass, noncalcified with or without adjacent lymphadenopathy. On sonogram, it appears as a hypoechoic area with either microlobulated margins with increased vascularity or being well-circumscribed.^[7] Magnetic resonance imaging is more sensitive and accurate in detecting metacentric/multifocal lesions. The presence of large-enhancing breast lesions with skin thickening point toward a primary breast NHL.^[8] Distinguishing lymphoma from carcinoma based on either clinical features or radiological

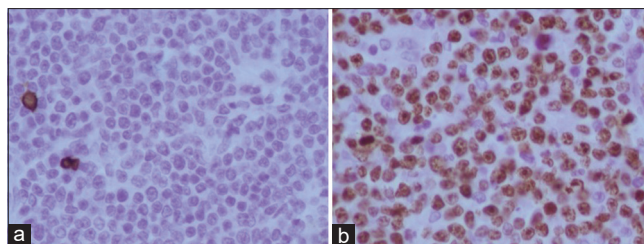


Figure 2: (a) Strongly positive CD-3 on immunohistochemistry; (b) Neoplastic cells showing high Mib-1 proliferation index

findings is almost impossible. Histopathology and IHC remains the gold standard for diagnosing a PBL.^[4] Multidisciplinary approach is advocated for these patients to attain complete cure, avoid local recurrence, and systemic metastases. Chemotherapeutic agents such as cyclophosphamide, doxorubicin, vincristine, prednisolone, and rituximab (R-CHOP regimen) are commonly used. In node-negative patients, RT has an important role as consolidation therapy.^[1,2] Important factors for relapse-free survival are as follows: (a) Ann Arbor stage, (b) International Prognostic Index, (c) LDH, and (d) RT. Over expression of Cyclin D1 is a poor prognostic factor in PBL. However, the optimal treatment remains unknown as there are reports showing recurrence following CHOP therapy alone.^[9]

Conclusion

Primary T-cell NHL in a premenopausal woman as was seen in our patient is extremely rare. Clinical and radiological features mimic breast carcinoma and can be differentiated only by histopathology and IHC. A multimodality approach (chemotherapy, surgery, and RT) based on the stage of the disease and histology is required in the management of these patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images, and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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