Primary Mammary (Non-Hodgkin) Lymphoma Presenting as Locally Advanced Breast Cancer

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Abstract:

Non-Hodgkin's lymphoma (NHL) of the breast can be either primary or secondary. Both are rare diseases. Primary breast lymphoma is an uncommon disease with poor clinical outcome. Breast lymphomas present less than 0.5% of malignant breast neoplasm and 2.2% of extra nodal lymphomas. We report a case of primary mammary non-Hodgkin lymphoma in a 70-year-old woman originally presented as locally advanced breast cancer with ulcerative skin changes. Clinical findings, diagnostic work-up, and follow-up are provided along with a review of the literature on primary mammary lymphoma.

Key words: Breast mass, NHL, PBL, IHC

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I. Introduction

Malignant lymphoma is a neoplasm which originates in lymphatic tissue. Primary non-Hodgkin's lymphoma of the breast is very rare and a distinct possibility in the diagnosis of breast malignancies, accounting for only about 0.1 to 0.5% of all reported malignant breast tumors and for 1.7–2.2% of extra nodal NHL. There are various kinds of breast lymphomas, but the most common is the B cell non-Hodgkin's lymphoma. At presentation, most patients are clinically thought to have breast carcinoma and the diagnosis of lymphoma is made at FNAC or at histopathology of the excised breast lump or mastectomy Primary and secondary lymphomas of the breast are defined according to the criteria used for other extra nodal lymphomas. Primary breast lymphoma (PBL) was diagnosed when the breast was the site of first or major manifestation of the lymphoma and there was no documentation of lymphoma elsewhere, excluding the presence of ipsilateral axillary node involvement by Wiseman and Liao's definition. However, these criteria are overly restrictive, since they limit the definition of PBLs to those exclusively localized to the breast. Patients with lymphoma of the breast that have disseminated elsewhere before diagnosis are not accepted. All lymphomas involving the breast but not including these criteria are considered as secondary breast lymphomas (SBL's). Pathologically there are no differences between primary or secondary lymphomas of the breast.

II. Case Report

A 70 years female presented with mass in the left breast for 1 year duration with ulceration of the skin and foul smelling discharge for the past 3 months and bleeding from the ulcer for past 10 days. No history of swelling elsewhere in the body. History of loss of appetite and loss of weight present. Patient is a known case of diabetes mellitus on oral hypoglycemic. No history of hypertension, tuberculosis, asthma. On examination patient was conscious, oriented, afebrile, hydration good, pallor +, no generalised lymphadenopathy. Systemic examination was normal. A ulcerative lesion of size 15*10 cm over the left breast with central eschar and slough with irregular borders, nipple areolar complex destroyed and surrounding skin erythematous. On palpation a 15*15 cm hard mass with irregular surface, moves along with breast tissue, not fixed to pectoralis muscle or chest wall and bleeds on touch. On examination of axilla 2*2 cm single central node, hard in consistency and mobile is palpable. Examination of right breast, right axilla, spine and cranium was found to benormal.



Fig 1: Clinical photograph showing left breast ulcerative mass

INVESTIGATIONS:

Mammography revealed a heterogeneously dense breast pattern with a marked degree of asymmetry between the right and left breast. An extremely dense, poorly defined mass measuring $12 \times 5 \times 10$ cm involving all the quadrants.On ultrasonography, a large heterogeneously hypoechoic mass occupied all the quadrants of the left breast. Multiple lymph nodes largest of size 1.8*1.3 cm present in left axilla. Ultrasonogram abdomen was normal. Ct chest revealed 7*3*7 cm illdefined heterogeneously enchancing soft tissue dense lesion in left breast with multiple nodes largest 1.8*1.4 cm in left axilla. Bone scan was normal. FNAC from the left breast lump showed possibility of phylloids tumor. Core needle biopsy was inconclusive.

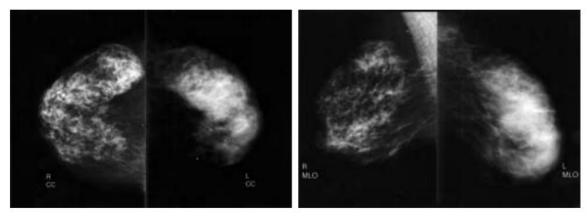


Fig 2: Bilateral mammography showing a large, high-density, poorly defined mass in the central and lateral aspects of the left breast

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Fig 3: FNAC left breast lump

We proceeded with mastectomy and specimen was sent for HP examination. It revealed sheets of medium to large sized lymphoid cells infiltrating the breast parenchyma. The cells are vesicular with prominent nucleoli with hyalinated fibrous tissue in between cluster of lymphoid cells. Lymph node shows medium to large cells with prominent nucleoli suggesting of Non Hodgkin lymphoma- large cell type. On immunohistocytochemistry(IHC) CD 20 wasdiffusely positive and CD 5 was focally positive. Patient was further evaluated to rule out the possibility of secondary lymphoma with CECT abdomen and pelvis showing no splenomegaly or intraabdominal lymphadenopathy. Bone marrow aspiration revealed normal study. Upper GI and colonoscopy revealed normal study. Hence we came with the diagnosis of primary mammary Non Hodgkin lymphoma and was planned for subsequent chemotherapy with R CHOP regimen and received 2 cycle and then lost follow up.

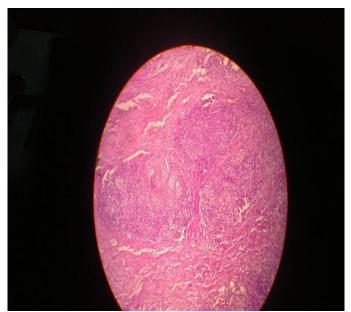


Fig 4: HPE showing lymphoid follicles in breast parenchyma

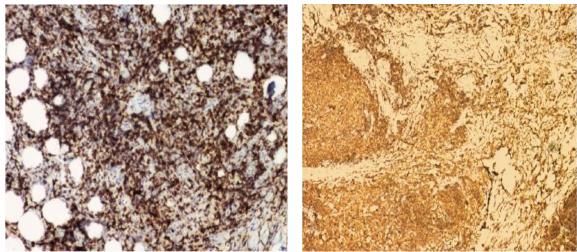


Fig 5: IHC showing CD5 and CD 20 positivity

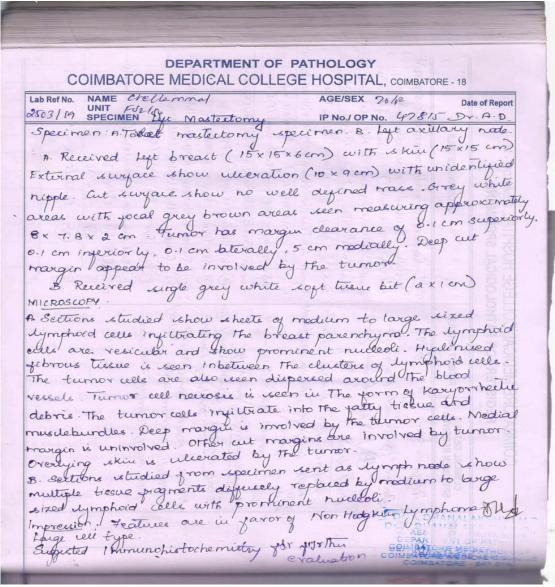


Fig 6: HPE report showing NHL Large cell type

III. Disscussion

PBL is a rare, potentially curable disease and has been considered a distinct clinicopathological entity. PBLs have a reported incidence of 0.04 -0.5% of all breast malignancies. PBLs account for less than 1% of all patients with NHLs and approximately 2.2% of all extra-nodal NHLs. The term primary breast lymphoma (PBL) is used to define malignant lymphomas primarily occurring in the breast in the absence of previously detected lymphoma localizations. Wiseman and Liao are credited with first defining the clinical criteria for the classification of PBL.

The usual clinical feature of a breast lymphoma is rapidly expanding, painless mass. There is a slight predilection for the right breast, but the explanation for this remains unclear. PBL is extremely rare in males. All histological types of lymphoma have been described. Primary breast lymphomas are most commonly B-cell lymphomas; approximately one-half are diffuse large B cell lymphoma.

The prognosis varies, as do the applied treatment modalities, which include surgery, radiotherapy and chemotherapy used alone or in combination. The prognosis of the lymphomas involving the breast either primary or secondary have been reported as poor; 5-year survival rates of 9 to 85% were reported in different series. Mastectomy has been a common component of PBL therapy for decades and remains a frequent treatment choice in some reports. Several studies found that mastectomy offered no benefit in the treatment of primary breast lymphoma. Ideally, surgery should be limited to a biopsy to establish the correct histological diagnosis, leaving the treatment with curative intent to radiotherapy and chemotherapy. Nodal status predicts outcome and guides the optimal use of radiation and chemotherapy. The choice of chemotherapeutic regimen should be based upon histological subtype, disease extent and the individual patient. The combination of anthracycline-based chemotherapy with rituximab should be considered standard, it appears that the addition of rituximab improves the outcome of all clinical and molecular subtypes of CD20-positive diffuse large B-cell lymphoma.

IV. Conclusions

Primary malignant lymphoma of the breast (PLB) appears to be a rare disease. A high degree of clinical suspicion is required for an early and prompt diagnosis of primary breast lymphoma so as to avoid unnecessary mastectomies. Fine needle aspiration cytology supplemented by immunocytochemistry is a reliable and cost-effective tool in the early diagnosis of primary breast lymphomas, while histopathology and immunohistochemistry are conclusive. No clear consensus concerning the therapy has emerged, although chemotherapy seems to be the more commonchoice, alone or in combination with other treatments. The choice of chemotherapy regimen and/or use of radiationtherapy (RT) is based upon the histological subtype, diseaseextent and individual patient. The prognosis and effectiveness of individual treatments are generally extrapolated from the rapy for other extra nodal lymphomas.

CONSENT

The authors would like to thank the patient's relatives for providing informed consent for the publication of this case report.

CONFLICT OF INTEREST

Authors have no conflict of interest to declare.

AUTHOR CONTRIBUTION

- 1.Patient management and treatment decisions.
- 2. Patient management, surgical treatment and manuscript writing.
- 3. Patient management, manuscript writing.

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