PRIMARY LYMPHOMA OF BREAST - UNUSUAL TYPE - CASE REPORT

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PRIMARY LYMPHOMA OF BREAST - UNUSUAL TYPE - A CASE REPORT (ABSTRACT): Introduction: Primary Lymphoma of the breast (PBL) is a rare entity, while secondary involvement of the breast with diffuse disease is more common. While Non Hodgkin Lymphoma type is the usual variant in PBL. We report a case primary Hodgkins lymphoma of the breast. Case Presentation: A 50 yr old woman presented three years ago with multiple lump in her left breast. Core biopsy revealed lymphoma like features. Excision biopsy of the upper outer quadrant lump revealed nodular lymphocytic predominant Hodgkin’s lymphoma (HL). Stage was II A. Chemotherapy resulted in complete remission. Patient was lost in follow up only to return with multiple local recurrences three years after primary treatment. Discussion: Primary NHL breast is rare but primary breast HL is rarer still. Nodular lymphocyte-predominant type of Hodgkin's disease is recognized as an entity distinct from classical Hodgkin's disease with a number of characteristics that suggest its relationship to non-Hodgkin's lymphoma. In histological terms, the differentiation of breast lymphoma cases from breast carcinomas and pseudo lymphoma is important. In our case, though no immunohistochemistry was available, the histological diagnosis was straightforward. Primary breast lymphomas behave similarly to lymphomas of similar histologic types. Treatment wise, the optimal sequence recommended is lumpectomy followed by Radiotherapy or radiotherapy alone for local control and standard anthracycline-based regimens. PBL tends to relapse to CNS; therefore, Computerized Tomography (CT) or Magnetic Resonance (MR) image of CNS is necessary during follow-up. Conclusion: PBL should be treated as lymphoma elsewhere with proper staging. While immunocytochemistry helps, when not available, treatment as per histological type should suffice. Proper staging and post treatment follow up is crucial.

KEY WORDS: PRIMARY LYMPHOMA OF THE BREAST, HODGKIN’S LYMPHOMA, NON HODGKIN’S LYMPHOMA..

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INTRODUCTION

Primary Lymphoma of the breast (PBL) is a rare entity, while secondary involvement of the breast with diffuse disease is more common. However, PBL is the most frequent haematopoietic tumor of the breast. Up to now, 700 Primary Breast Lymphomas (PBL) have been described as case reports or case series. Their incidence is growing; recently Aviles and coll. presented a review consisting of 96 patients [1], while the International Extra nodal Lymphoma Study Group (IELSG) has registered 204 cases [2].

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Diagnostic criteria for PBL of the breast are:
1. Presence of technically adequate pathologic specimens,
2. Close association of mammary tissue and lymphomatous infiltrate,
3. No prior diagnosis of an extra-mammary lymphoma and
4. No evidence of concurrent widespread disease, (except for ipsilateral axillary lymph nodes, if concomitant with the primary lesion).

The incidence of PBL lies between 0.04 and 1.1% for all breast tumors. Almost all descriptions of PBL is of Non Hodgkin type. We report a case primary Hodgkin’s lymphoma of the breast.

CASE PRESENTATION
A 50 yr old woman first presented three years ago with a lump in her left breast. On examination, there was bilateral multiple (five on the left and three on the right) lump noted with largest being a 3x 2 centimeter in the upper outer quadrant of the left breast. all the lumps were mobile and free overlying skin. No regional lymph node was palpable.

Mammography and ultrasound confirmed the clinical findings. The lumps were hypoechoic and ill defined. F.N.A.C from the lumps was inconclusive and core biopsy revealed lymphoma like features. No carcinoma was detected. Staging computerized tomography of chest and abdomen was normal. Diagnostic excision biopsy of the upper outer quadrant lump was done which revealed nodular lymphocytic predominant Hodgkin’s lymphoma. However, Immunohistochemistry was not available. Bone Marrow and blood investigations were normal.

After oncologist consultation, she was recommended for chemotherapy with endoxan and vincristin. There was a complete response and the patient was on follow up at four monthly intervals. Computerized Tomography (CT) scan was done at six monthly intervals. Patient was lost to follow up after two years only to return after another year with multiple ulcerating breast nodules along with supra clavicular and left axillary swelling (Fig. 1,2). On biopsy, it was confirmed to be loco regional recurrence. Chest and abdominal imaging and other work up confirmed that here was no distant spread of the disease. At present, the patient is being evaluated for further chemotherapy.

Fig.1 Ulcerating breast nodules  
Fig.2 Neck node enlargement
DISCUSSION

Primary NHL breast is rare but primary breast HL is rarer still. The incidence of primary breast lymphoma (PBL) lies between 0.04 and 1.1% of all breast tumors. For all extra nodal NHL, 1.7–2.2% occurs in breast [3]. It is usually of non-Hodgkin's type. Incidence for primary HD in breast is unknown. Secondary involvement of the breast in patients with diffuse disease is more common [4].

Nodular lymphocyte-predominant type of Hodgkin's disease is recognized as an entity distinct from classical Hodgkin's disease. Previous classification systems recognized that biopsies from a subset of patients diagnosed as having Hodgkin's disease contained a predominance of small lymphocytes and rare Reed-Sternberg cells. A subset of these patients has tumors with nodular growth pattern and a clinical course that varied from that of patients with classical Hodgkin's disease. This is an unusual clinical entity and represents <5% of cases of Hodgkin's disease. They usually occur in males, localized to peripheral lymph nodes, have frequent relapses and usually have good prognosis.

Nodular lymphocyte-predominant type of Hodgkin's disease has a number of characteristics that suggest its relationship to non-Hodgkin's lymphoma. These include a clonal proliferation of B cells and a distinctive immunophenotype; tumor cells express J chain and display CD 45 and epithelial membrane antigen and do not express two markers normally found on Sternberg-Reed cells, CD 30 and CD 15. This lymphoma tends to have a chronic, relapsing course and sometimes transforms to diffuse large B cell type, which is again the most common type of PBL.

In histological terms, the differentiation of breast lymphoma cases from breast carcinomas and pseudo lymphoma is important. In particular, poorly differentiated breast carcinomas and lobular carcinoma may cause differential diagnostic difficulties. In addition to the absence of in situ lobular neoplasm adjacent to infiltrative areas, leukocyte common antigen–positive and pancytokeratin-negative immunohistochemistry are helpful findings in differentiating lymphoma from lobular carcinoma [5]. In our case, though no immunohistochemistry was available, the histological diagnosis was straightforward.

Treatment of PBL is controversial. Some suggests that optimal sequence of full-dose anthracycline-containing regimens and radiation therapy should be the treatment of choice for patients with PBL [6]. Other studies suggest combination of surgery and radiotherapy for local control along with chemotherapy.

Primary breast lymphomas behave similarly to lymphomas of similar histological types and stages presenting at other sites. Most of these cases are NHL The majority is diffuse large B-cell lymphoma. Other common types are mucosa associated lymphoid tissue lymphoma and peripheral T-cell lymphoma. Primary breast lymphomas behave similarly to lymphomas of similar histological types and stages presenting at other sites.

PBL must be regarded as a kind of systemic disease in spite of its pathologic type and Chemotherapy should be included in the multi-modality treatment of PBL [7-9].

Treatment wise, the optimal sequence recommended is lumpectomy followed by Radiotherapy or radiotherapy alone for local control and standard anthracycline-based regimens. PBL tends to relapse to CNS, therefore, CT or MR image of CNS is necessary during follow-up [4,10]. Burkitt's or Burkitt-like lymphoma can bilaterally involve the breast of a young pregnant or lactating woman and typically behaves aggressively [11].

The role of surgery in PBL should be limited to get a definitive diagnosis while for the staging and the treatment, CT scan and chemo/radio therapy respectively are mandatory.
The surgical treatment must always be oncologically radical (R0); mastectomy must not be carried out as a rule, but only when tissue-sparing procedures are not feasible. Axillary dissection must always be performed for staging purposes, so avoiding the risk of under-staging II, due to the possibility of clinically silent axillary node involvement [12].

There is no suggestion regarding treatment of HD type of PBL. In general, treatment of patients with nodular lymphocyte-predominant Hodgkin's disease, which is the subtype in our case, is controversial. Some clinicians favor no treatment and merely close follow-up. Regardless of the therapy utilized, most series report a long-term survival of >80%.

Although our case did not have immunohistochemistry to confirm that it is more of Hodgkin’s type rather than Non Hodgkin types as usually occur, Nodular sclerosing subtype of Hodgkin’s disease has many immunocytochemical feature similar to NHL and could have been confusing as well. Histology should guide the treatment.

**CONCLUSION**

PBL should be treated as lymphoma elsewhere with proper staging. While immunocytochemistry helps, when not available, treatment as per histological type should suffice. Proper staging and post treatment follow up is crucial.

**REFERENCES**