Case Report

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Primary Breast Lymphoma in A 25-Year-Old Young Woman: A Case Report and Review of Literature

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Abstract

Primary breast lymphoma is a rare entity comprising 0.4-0.5% of all breast neoplasms. Diffuse large B-cell lymphoma is the most common identifiable type of primary breast lymphoma with the usual presentation being that of a painless palpable mass similar to breast carcinoma. A confirmed diagnosis is based on initial biopsy findings or post-operative histopathology and immunohistochemistry (IHC) reports. The treatment options vary from surgical interventions to chemotherapy and/or radiotherapy. However, there is no uptodate standard treatment guidelines present. In view of this rarity, we herein report a case of a 25 year-old young woman who presented with primary breast lymphoma of DLBCL variety.

Keywords: primary breast neoplasm, young woman, DLBCL, immunohistochemistry.

INTRODUCTION

Breast is a rare site of extra nodal involvement of lymphoma. Breast lymphoma represents approximately 0.5% of breast malignant neoplasms and comprises 1.7–2.2% of extra nodal lymphoma.[1,2] Primary breast lymphoma (PBL) is an unusual tumour accounting for 0.4–0.5% of all breast carcinomas.[3] Wiseman and Liao first described it in 1972.

The diagnostic criteria were: the breast as the site of presentation, breast tissue in close relation with lymphomatous infiltration, absence of disseminated disease beyond the ipsilateral axillary lymph nodes and no previous lymphoma diagnosis.[4] It is considered a rare neoplasm because lymphoid tissue is absent in the breast region.[5]

It occurs almost exclusively in females where the average age of diagnosis varies between 60 and 65 years with few cases of primary breast lymphoma reported in men.[6] There is a bimodal age distribution of PBL of DLBCL histology with a peak at 30–35 years and at 55–60 years.[7] Approximately 10% of cases are bilateral with a propensity for involvement of the right breast more than the left. A painless breast lump is the commonest presentation of PBL, mostly in the upper outer quadrant. Other symptoms like nipple discharge, skin retraction, peau d' orange appearance, erythema and oedema are quite unusual.[3]

Imaging findings are usually non specific with most lesions appearing hyperdense (91%) and oval (71%) in mammography and as single (75%), circumscribed (50%), micro lobulated (38%), oval

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(50%) and hypoechoic (87%) lesion in ultrasound. Calcifications or spiculated margins are infrequent.[8] PBL is usually non-Hodgkin's B-cell type, which accounts for about half of breast lymphomas with the most frequent subtype being diffuse large B-cell lymphoma (DLBCL).

The treatment options for PBL includes surgical intervention, chemotherapy and radiotherapy. However, the optimal management still remains unclear and controversial.[9] Mastectomy does not appear to improve survival or risk of recurrence in the treatment of primary breast lymphoma.[3]

Based on the success of chemotherapy in the treatment of nodal and other extra nodal non-Hodgkin's lymphomas, chemotherapy is considered as the mainstay of therapy. Prospectively the role of radiotherapy has never been explored.[9] The cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) regimen is the most accepted chemotherapy regimen.[11] A study by Jennings WC et al.[10] showed that nodal status predicts survival and outcome and guided optimal use of radiation and chemotherapy. Central nervous system (CNS) involvement has also been described, So some authors recommend CNS prophylaxis also.[12]

Here, we report a case of a 25-Year-Old Young woman with primary breast non-hodgkins lymphoma of DLBCL variety.

CASE REPORT

A 25 year-old young woman presented in our out-patient department with a 6 months history of lump in her left breast. There was no complain of pain or nipple discharge. On local examination a firm to hard mobile lump of about 2x2 cm was felt in the lower outer quadrant of the left breast. It was not fixed to overlying skin or underlying muscle. Contralateral breast examination and lymph node examination was unremarkable. Her systemic examination was within normal limits.

Ultrasonography (USG) was done which reported a lesion in the left breast of size 2.3x2.3 cm in 5 0'clock position. Fine needle aspiration and cytology (FNAC) was done which reported as round cell tumour. She then underwent a core needle biopsy which reported atypical round cell lesion. Immunohistochemistry of the tumour cells was done in which the cells were positive for CD-45, CD-20, BCL-2 and MUM-1. The cells were negative for CD-3, BCL–6, CD-10, CK, C-myc, tdT, synaptophysin and chromogranin. The proliferative index was 95% which gave a final diagnosis of high grade non – hodgkins B cell lymphoma – diffuse large B cell lymphoma – activated B-cell type. Her staging Computed Tomography (CT) chest was done which revealed a soft tissue density in the infero lateral quadrant of the left breast measuring 4x3.5x3.1 cm, not infiltrating skin and underlying muscle which was reported as neoplastic aetiology. Multiple enhancing foci was seen in both the breasts with sub centimetric lymph nodes present in the left axilla. No other abnormality was seen. Her CT abdomen and pelvis was unremarkable.

The patient was then given chemotherapy with R(rituximab) - CHOP regimen for 6 cycles and had excellent clinical response to it. A PET-CT examination was done later which showed complete radiological response too, following it she was taken up for involved field radiotherapy for 30 Gray (Gy) in 15 fractions. After radiation completion she is kept under follow – up with no clinical evidence of disease anywhere in the body till date.

DISCUSSION

Primary breast lymphoma (PBL) remains an infrequent neoplasm, so clinicians treating breast carcinomas should be aware of this entity in order to distinguish its clinical presentation, management, and prognosis from breast carcinoma cases .[13] It is defined pathologically as the presence of lymphomatous infiltrate in normal breast tissue in a patient with neither previous nor concurrent non-Hodgkin's lymphoma at another site, although involvement of ipsilateral axillary lymph node enlargement may be present.[14]

PBL shows a bimodal age distribution, with the younger population showing bilateral involvement and older population showing unilateral involvement.[15] It usually affects women in their fifth or sixth decade of life.[16] 95%–100% are reported among female patients and is very rare in males. Single breast involvement is more common, particularly in the right upper quadrant. Of all PBL cases approximately 1% to 14% have bilateral breast lymphomas.[5]

PBL most commonly presents as a palpable lump. Less often, it may also present as diffuse breast enlargement. The majority of breast lymphomas are of the non-Hodgkin's variety (PBNHL)[16] with majority having a B-cell lineage with DLBCL, which accounts for 40–70% of all breast lymphomas with features of non-germinal centre cells. The diagnostic approach includes radiological investigation (mammography, USG, magnetic resonance imaging

(MRI), and positron emission tomography), FNAC, and immunohistochemical biomarkers.[5]

On USG, it often presents as a hypoechoic area with circumscribed or micro lobulated margins demonstrating increased vascularity.[5] The typical mammographic appearance is of a solitary, noncalcified, circumscribed or indistinctly marginated mass with adjacent lymphadenopathy. Diffuse increased parenchymal density with skin thickening is observed less commonly. MRI may be more sensitive and accurate in representing and detecting multicentric lesions.[16] FNAC is ideal for the primary diagnosis of PBL as it helps in early detection and treatment.[5] Singh et al.[17] reported the importance of fine needle aspiration cytology (FNAC) in diagnosing breast lymphoma in a study of 13 cases over 20 years of which all except one were females. Chopra et al[18] reported an unusual case of synchronous mucosa associated lymphoid tissue (MALT) lymphomas involving bilateral orbits and breasts.

The immunohistochemistry profile consistent with diffuse large B-cell lymphoma, non-germinal centre-type shows cells that are positive for the B-cell markers CD20 and CD-79 and negative for T-cell markers CD3 and CD-5.

Although multimodality approach have been used in most studies earlier, mastectomy does not appear to improve survival or risk of recurrence in the treatment of PBL.[3] Several studies have recommended that surgery should be offered for diagnosis only and that minimally invasive surgery is the preferable option as extensive surgery may carry a high risk of morbidity. Axillary dissection adds no therapeutic advantage.[10,19,20] The management of PBNHL is based on histologic grade. Patients with low grade disease can be managed with local therapy alone. The role of chemotherapy in this group is unclear. Patients with intermediate or high grade disease have better out come if chemotherapy is included.[16]

In MabThera International Trial (MInT) trial, the addition of rituximab to chemotherapy has improved response rate, disease free survival and overall survival in patients with aggressive early stage nodal DLBCL.[19] SWOG 0014 trial has shown that patients with "limited" early stage disease have excellent outcomes with three cycles of CHOP with rituximab followed by involved field radiation therapy. Radiation therapy should be avoided preferably in young females due to the increased risk of breast cancer 10 to 20 years later. Stage IIE (node positive) and advanced DLBCL of the breast is treated with systemic chemotherapy (CHOP-R x 6 to 8 cycles).[3]

The role of central nervous system (CNS) prophylaxis in DLBCL of the breast is controversial. Nevertheless, given the high incidence of CNS recurrence, CNS prophylaxis should be considered.[16]

PBL is reported to exhibit a poor prognosis among extra nodal B-cell lymphomas. The overall survival rate of PBL with a B-cell phenotype is 43% at 5 years and even worse for DLBCL breast which has got a median survival of 36 months. Based on the studies on early stage PBL with low International Prognostic Index (IPI) score, the use of anthracycline containing chemotherapy and radiotherapy are significantly associated with longer overall survival.[3]

CONCLUSION

PBL is an extremely rare malignancy and its presentation is quite similar to breast malignancy. Hence, a high index of suspicion is need for the early diagnosis and treatment so as to avoid unnecessary mastectomies. FNAC along with immuno-cytochemistry is a reliable and cost-effective method in the early diagnosis while histopathology along with immunohistochemistry is conclusive. The management depends on the histological type and chemotherapy seems to be the most acceptable option alone or in combination with surgery and radiotherapy.

Conflict of interest: Nil

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