

Primary Lymphoma of Breast in a Male – A Rare Case

Dr Muvva Sri Harsha¹, Dr Srikanth Jakkula², Dr Rajesh Goud E³

¹Post graduate, Department of General surgery, SVS Medical college, KNR university of health sciences, Telangana

² Post graduate, Department of General surgery, SVS Medical college, KNR university of health sciences, Telangana

³ Consultant Surgical Oncologist, Department of General surgery, SVS Medical college, KNR university of health sciences, Telangana

Corresponding Author: Dr Muvva Sri Harsha

Abstract: Primary breast lymphoma is a rare disease accounting for 0.4-0.5% of all breast malignancies. They are even more rare in males. Amongst the various types of breast lymphomas, Most breast lymphomas are of B cell origin. Here we present a case of 65 year old male who presented with swelling and irritation over his left chest. Initial diagnosis of Carcinoma breast was made, followed by thorough investigations the patient underwent Modified radical mastectomy. Histopathology and immunohistochemistry confirmed the diagnosis of Diffuse lymphoma of the breast. Patient was kept on follow-up and advised adjuvant chemotherapy.

Keywords: Lymphoma, Male breast, carcinoma breast

Date of Submission: 28-02-2019

Date of acceptance: 18-03-2019

I. Introduction

The term “primary breast lymphoma” (PBL) is used to define a malignant lymphoma primarily occurring in the breast in the absence of previously detected lymphoma localizations^[1]. It is a rare disease, accounting for only 0.4-0.5% of all breast malignancies, 0.38-0.7% of all non-Hodgkin lymphomas (NHL), and 1.7-2.2% of extranodal NHL. PBL occurs mostly in females and they are very few male patients reported in the literature. More than 80% of PBL are B-cell lymphomas, mostly CD20+. The most frequent histopathologic types are: diffuse large B-cell lymphoma (DLBCL) which accounts for up to 50% of all PBL, follicular lymphoma (FL) – 15%, MALT lymphoma – 12.2%, Burkitt’s lymphoma (BL) and Burkitt-like lymphoma – 10.3%.The clinical and radiological presentations of the disease are very similar to those of breast cancer, however in majority of the cases, they mimick the inflammatory breast cancer. Multiple treatment strategies involving surgery, radiation, and chemotherapy (alone or in various combinations and sequences) have been reported. However amongst all, a combined therapy have shown better prognosis.^[3]

II. Case Report

A 60 year old male presented to us with complaints of swelling over right side of his chest since 3 months along with pain without any history of discharge from the nipple. On examination a 4*3 cm swelling was seen involving the nipple areolar complex, swelling is firm in consistency without any fixity to the underlying structure. 2 firm lymph nodes were palpable in the axilla. Ultrasound and mammogram were inconclusive. Fine needle aspiration cytology was done which is suggested inflammatory pathology. True cut biopsy was done and histopathological examination lead us to a provisional diagnosis of lympho-proliferative malignancy. A complete metastatic work-up was done and we found no evidence of secondaries. All the routine blood investigations were within normal limits.



Fig 1: Pre-operative clinical picture



Fig 2: Intra-operative specimen of MRM

Modified radical mastectomy was performed and the specimen was sent for Histopathological examination. All the margins were found to be free from tumor cells and the lymph nodes exhibited reactive hyperplasia. Immuno-histochemistry (IHC) markers were done which suggested a Diffuse Lymphoma in the view of CD45 Positive. Primary neuroendocrine tumor (PNET) was a remote since CD99 and FLI were negative. ER, PR and HER were negative ruling primary ductal cell carcinoma, rare exception of triple negative is a remote possibility but it has been excluded in the view of CD45 positive. Hence, a final diagnosis of Primary diffuse lymphoma was made. Patient was advised adjuvant chemotherapy. There was no recurrence for 2 years and the patient was kept on regular follow-up.



Fig 3: Cut-section

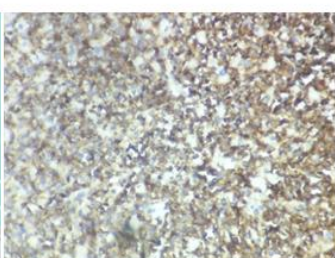


Fig 4: CD45+

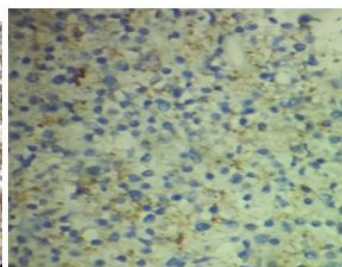


Fig 5: CD20+

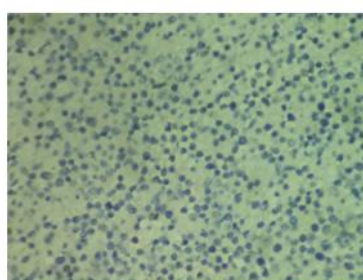


Fig 6: FLI negative

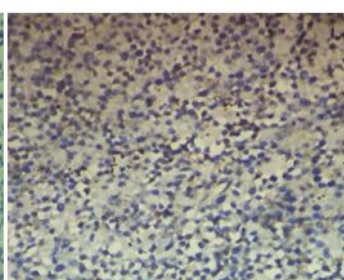


Fig 7: CD99 negative

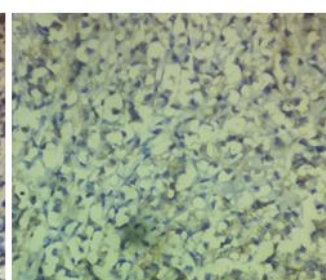


Fig 8: HER negative

III. Discussion

Primary breast lymphoma (PBL) 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^[4]. Wiseman and Liao are credited with first defining the clinical criteria for the classification of PBL.^[5] The specific criteria for the diagnosis of PBL include:

- 1.The clinical site of presentation is the breast.
2. A history of previous lymphoma or evidence of widespread disease are absent at diagnosis.
- 3.Lymphoma is demonstrated with close association to breast tissue in the pathologic specimen.
- 4.Ipsilateral lymph nodes may be involved if they develop simultaneously with the primary breast tumor.

Most common symptom of breast lymphoma is a painless breast mass, most frequently in the upper outer quadrants. Skin retraction, erythema, peau d' orange appearance, and nipple discharge are uncommon in

lymphomas.^[6,7] Distinct mammographic or sonographic features have not been described in the literature for breast lymphoma to differentiate it from breast carcinoma or benign breast entities. Histopathology remains the gold standard for the diagnosis of lymphoma of breast. 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 (DLBCL). Indolent histologies, follicular non-Hodgkin's lymphoma or extranodal marginal zone (MALT) lymphoma occur less commonly. More than 80% of PBL are B-cell lymphomas, mostly CD20 +. The most frequent histopathologic types are: diffuse large B-cell lymphoma (DLBCL) which accounts for up to 50% of all PBL, follicular lymphoma (FL) – 15%, MALT lymphoma – 12.2%, Burkitt's lymphoma (BL) and Burkitt-like lymphoma 10.3%. Other histological types of PBL include marginal zone lymphoma (MZL), small lymphocytic lymphoma (SLL), and anaplastic large cell lymphoma (ALCL)^[9].

Treatment of primary lymphoma of breast can be surgical, chemotherapy or a combined therapy. Off the multiple modalities available a combined approach of surgical followed by adjuvant chemotherapy has shown better results. The role of central nervous system (CNS) prophylaxis in DLBCL of the breast is controversial. Nevertheless, given this high incidence of CNS recurrence, central nervous system (CNS) prophylaxis should be considered.^[10,11,12]

Prognosis usually varies depending on the treatment received along with the time of presentation. Earlier the presentation and diagnosis better the prognosis.

IV. Conclusion

Primary lymphoma of breast is a rare entity and the incidence is on rise. It closely mimicks inflammatory breast cancer, hence Lymphoma should always be considered as a differential. Histopathology along with immunohistochemistry remains the gold standard in establishing the diagnosis. Prognosis is good especially if the diagnosis is made early followed by the appropriate treatment.

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Dr Muvva Sri Harsha. "Primary Lymphoma of Breast in A Male – A Rare Case." *OSR Journal of Dental and Medical Sciences (IOSR-JDMS)*, vol. 18, no. 3, 2019, pp 21-23.