



Primary Lymphoma of the Breast

Amol Dilip Amonkar^{1*}, Miguel Furtado², Vineeta Maryann De Souza¹ and Mervyn Correia¹

¹Department of General Surgery, Goa Medical College, India

²Department of Radiology, Goa Medical College, India

Abstract

Primary Breast Lymphoma (PBL) is defined as a localized involvement of one or both the breasts with or without ipsilateral axillary nodal involvement. It is usually a rare manifestation of extra nodal non-Hodgkin's lymphoma. It is a rare clinicopathological entity with only a handful of cases reported till date. We report a case of a 65-year old female with PBL with relevant discussion on the management from a surgeon's perspective.

Keywords: Primary lymphoma; Breast; Chemotherapy; Non-Hodgkins

Introduction

Primary lymphoma of breast is defined as a malignancy occurring in the breast in absence of previously diagnosed lymphoma [1]. It represents 0.04% to 0.5% of malignant breast tumors [1]. It is an uncommon manifestation of non-Hodgkins lymphoma (<1%) and occurs in 1.7% to 2.2% of patients suffering from extranodal lymphoma [2,3]. The disease occurs almost exclusively in women with a median age of 60 to 65 years [2]. The most common type of primary breast lymphoma is Diffuse Large B-Cell Lymphoma (DLBCL) which accounts for 50% of cases followed by follicular lymphoma (15%) and MALT lymphoma (12.3%) [3]. We reports a case of PBL with relevant discussion on management from a surgeon's perspective.

Case Presentation

A 65-year old female presented with an asymptomatic lump in her right breast which was of insidious onset and grew slowly over a period of 3-years. On local examination, there was a lump in the right breast in the upper outer quadrant measuring 3 cm × 4 cm. It was hard in consistency and had ill defined borders. Clinical examination revealed no regional or generalized lymphadenopathy and no other significant findings. Left breast was normal. Mammogram of the right breast was reported-BIRADS 4C. FNAC was reported as lymphoproliferative disease, advised biopsy (Figure 1). Trucut biopsy revealed PBL of B-cell type, confirmed by immunohistochemistry. CECT Thorax and abdomen showed no other significant findings other than the right breast lump. Medical oncology opinion was sort and advised wide local excision followed by chemotherapy and radiotherapy. Final diagnosis of PBL was made and the patient underwent wide local excision with clearance margin of 2 cm. Post-operative period was uneventful. The patient is set to undergo radiotherapy followed by chemotherapy (Figure 2).

Discussion

PBL is defined as localized involvement of one or both the breasts with or without ipsilateral axillary nodal involvement. It is a rare manifestation of extra nodal non-Hodgkin's lymphoma [1]. Unlike primary breast carcinoma, surgery is not the key treatment. Treatment is confined to a combination of radiotherapy and chemotherapy (R-CHOPP) [2]. Although lymphomas are common hematological malignancies, breast involvement with lymphoma is infrequent. Diagnostic criteria for PBL, Wiseman & Liao [3]: (a) Both mammary tissue and lymphomatous infiltrate present in close association in an adequate pathological specimen, (b) No evidence of widespread lymphoma by standard staging techniques or preceding extra mammary lymphoma except ipsilateral axillary nodal involvement if diagnosed simultaneously. PBL occurs usually 5th to 6th decade [4] and presents with a single or sometimes multiple painless palpable lumps which may be bilateral (11%) [2]. Nipple or skin retraction, nipple discharge or the usual B-symptoms- fever, weight loss, night sweats are infrequent [3]. It may sometimes present as diffuse breast enlargement and edema and can mimic an inflammatory process [3]. Right breast predominance noted [4]. The imaging findings of breast lymphoma are non-specific and may resemble any other breast malignancy or may have a more benign appearance [4]. The most common mammographic abnormality is a solitary non-

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*Correspondence:

Amol Dilip Amonkar, Department of General Surgery, Goa Medical College, Bambolim, Goa, India, Tel: 9886535829;

E-mail: amonkaramol@gmail.com

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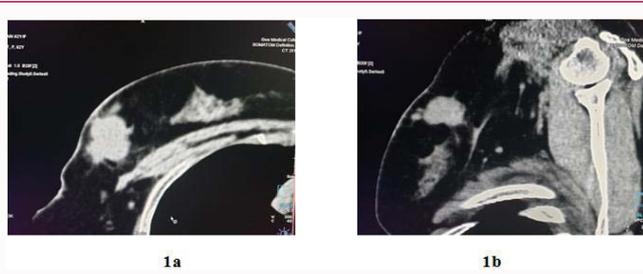


Figure 1: (a) CT picture showing an axial view of breast lymphoma with ill defined borders. (b) CT picture showing a sagittal view breast lymphoma with ill defined borders.

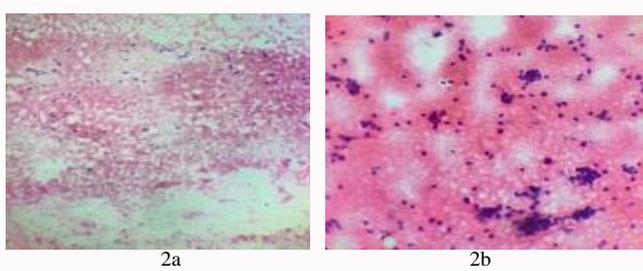


Figure 2: (a) Microscopy showing lymphoid cells with scanty cells and inconspicuous nucleoli. (b) Immunohistochemistry confirming primary breast lymphoma.

calcified breast mass with circumscribed or indistinct margins [1]. On ultrasound a hypoechoic solid oval round mass with circumscribed or indistinct margins is the most common appearance [2,3]. There are no large case reviews on MRI of PBL. PET-CT scan is valuable in staging and follow up of lymphoma patients [5]. There are no current established guidelines for treatment of PBL. The treatment may involve a combination of surgery, chemotherapy and radiotherapy [2]. Radical mastectomy has been described in older literature as

a part of treatment but subsequent studies have showed that it offers no benefit and may actually delay the start of chemotherapy [4]. Chemoimmunotherapy with consolidation radiotherapy is now considered the mainstay of treatment. CHOP or CHOP like anthracycline based chemotherapy combined with rituximab is now considered the standard for most DLBCL breast lymphoma [3]. This may be followed with radiotherapy to ipsilateral breast and regional lymph nodes. In the study by Avites et al. the 10-year overall survival in patients treated with either radiotherapy or chemotherapy alone versus a combination of both was 50% vs. 76% respectively. PBL is an aggressive tumor with a high relapse rate mainly involving extranodal sites especially the central nervous system [5].

Conclusion

PBL is an uncommon clinical entity. Literature shows a handful of cases. Due to its rarity, there are no standard treatment guidelines. Management is guided by literature and personal experiences of senior surgical and medical oncologists. We report this case on account of its rarity and lack of established guidelines which may help others in the management of such cases.

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