CASE REPORT

Primary Breast T Cell Lymphoma Involving Nipple-Areolar Complex in a Young Patient

Mithun Ram¹ · Pooja Ramakant¹ · Harshad Parmar² · Biju George³ · M. J. Paul¹

Received: 28 August 2014 / Accepted: 7 January 2016 / Published online: 25 January 2016
© Association of Surgeons of India 2016

Abstract We present a rare case of recurrent primary breast lymphoma involving the nipple-areolar complex and review literature on primary breast lymphoma, its clinical presentation and management. It is diagnosed by histopathology. It needs multimodality management protocols.

Keywords Breast lymphoma · T cell lymphoma · Ulcero-proliferative lesion of breast · Rare breast cancer pathology

Introduction

Breast lymphoma is a rare form of breast malignancy comprising up to 0.5 % of all breast malignancies [1–4]. It needs multimodality treatment in form of surgery, chemotherapy, and radiation therapy.

Case Report

A 35-year-old premenopausal lady presented with recurrent left breast lump since 1 year. She underwent left

breast lumpectomy elsewhere following which she noticed recurrent lump in the region of the nipple. She had also noticed ulceration and bleeding over the lump for past 1 month. Local examination of the left breast showed a 10 × 7 cm ulcero-proliferative, firm lesion involving and distorting the nipple-areolar complex with ulcerated surface bleeding (Fig. 1). There was an ipsilateral firm mobile axillary lymph node. Opposite breast, axilla, supraclavicular regions, and other lymph nodes area were normal.

Ultrasound showed a large exophytic, hypoechoic lesion 9.7 × 4.6 cm with increased vascularity, with few round hypoechoic lymph nodes with loss of fatty hilum in the left axilla. Wedge biopsy was reported as atypical T cell infiltrates suggestive of a T cell lymphoma of the breast. Chest radiograph, ultrasound abdomen and bone marrow biopsy were negative for disease involvement.

In view of a large ulceroproliferative lesion distorting the whole nipple-areolar complex with suspicious lymph nodal metastasis, she underwent a left modified radical mastectomy. Histopathology was confirmed as breast tissue infiltrated by a tumor arranged in sheets and composed of small to medium sized lymphoid cells with pleomorphic, hyperchromatic, mitotically active nuclei with clumped chromatin, few with prominent nucleoli and moderate amounts of eosinophilic and vacuolated cytoplasm. Lymph nodes showed reactive hyperplasia. Immunohistochemistry was positive for CD3 and CD5 suggesting a peripheral T cell lymphoma (Fig. 2). Hematology consult was sought, and she was given 6 cycles of adjuvant chemotherapy with Rituximab, Cyclophosphamide, Hydroxydaunorubicin, Vincristine, Prednisolone (R-CHOP) regimen along with intrathecal methotrexate for central nervous system (CNS) prophylaxis followed by locoregional radiotherapy.

✉ Pooja Ramakant
poojaramakant@rediffmail.com

¹ Endocrine and Breast Surgery Department, Christian Medical College, Vellore, India

² Pathology Department, Christian Medical College, Vellore, India

³ Hematology Department, Christian Medical College, Vellore, India

Fig. 1 Clinical image of the lesion in breast

Discussion

Primary lymphoma of the breast (PBL) is a rare condition. About 2 % of extra-nodal lymphomas and less than 1 % of all Non-Hodgkin's lymphoma present with breast as the primary organ of involvement [1–3].

PBL is thought to originate from two main sources—either from the migratory lymphocytes present within the breast tissue, which are considered to be part of mucosa-associated lymphoid tissue (MALT) or from lymphocytes within the intramammary node [5, 6].

Though a few case reports have been published of PBL in the male breast, it predominates in females with a bi-modal age distribution at 40 and 60 years with average age at presentation being 55 years. Our patient presented in much younger age group compared to the literature. The most common presentation is a painless unilateral mass with bilaterality seen in 10 % and contralateral metachronous disease seen in 15 % of patients. Involvement of the nipple-areolar complex, as in our patient, is rare. A rare form of presentation in young child bearing women may be bilateral breast masses resembling inflammatory carcinoma with rapid dissemination, especially to the Central Nervous System [7].

Diagnostic criteria for PBL was laid down by Wiseman and Liao [8]

- Adequate histopathological tissue for diagnosis
- Close relation between the lymphomatous infiltrate and breast tissue
- No evidence of widespread or prior extramammary disease
- Ipsilateral lymph nodal involvement maybe considered acceptable, if it arises simultaneous with the primary

Non-Hodgkin's lymphoma (NHL) is more common than Hodgkin's lymphoma (HL) in the breast. The most common histological subtype of NHL is diffuse large B cell type lymphoma. T cell lymphomas as seen in our patient are rare and follow an aggressive clinical course.

Staging of the disease is done according to the WHO Ann Arbor staging. International prognostic index is used to prognosticate the 5-year survival rate.

The modalities of management are systemic chemotherapy, locoregional radiotherapy, and surgery. These tumors are highly chemosensitive hence, chemotherapy (Anthracycline based) with at least 4 cycles of Cyclophosphamide, Hydroxydaunorubicin, Vincristine, Prednisolone with Rituximab is recommended. As these tumors are also highly radiosensitive, delivering an average of 40 Gy to the breast and lymph nodes is highly recommended for prevention of local recurrence. Surgery, on the other hand, assumes a more diagnostic role with chemotherapy and radiotherapy forming the primary modalities of management. Multimodal therapy with chemotherapy, radiotherapy, and surgery is currently advocated. In low grade variants, local therapy (Surgery, Radiotherapy) is sufficient whereas intermediate and high grade variants need systemic chemotherapy.

The common sites of recurrence are the CNS, lymph nodes and contralateral breasts. Though routine CNS prophylaxis is

Fig. 2 Histopathology showing lymphoid cells with mitotically active nuclei and eosinophilic vacuolated cytoplasm and Immunohistochemistry CD3 and CD5 positivity

not indicated, some authors recommend that a high IPI score may be an indication for CNS prophylaxis.

Conclusion

PBL is an uncommon differential for a breast lump, usually cannot be differentiated from invasive ductal carcinoma by imaging and needs histopathology with IHC for diagnosis. It is primarily managed by chemoradiation and CNS prophylaxis, with the role of surgery being mainly diagnostic.

Acknowledgments The authors would like to thank Mr. Santhanam for all technical assistance.

Compliance with Ethical Standards

Support/Funding There was no funding source for this case report.

Conflict of Interest The authors declare that they have no competing interests.

References

1. Validire P, Capovilla M, Asselain B, Kirova Y, Goudefroye R, Plancher C et al (2009) Primary breast non-Hodgkin's lymphoma: a large single center study of initial characteristics, natural history, and prognostic factors. *Am J Hematol* 84:133–139
2. Talwalkar SS, Miranda RN, Valbuena JR, Routbort MJ, Martin AW, Medeiros LJ (2008) Lymphomas involving the breast: a study of 106 cases comparing localized and disseminated neoplasms. *Am J Surg Pathol* 32:1299–1309
3. Aviv A, Tadmor T, Polliack A (2013) Primary diffuse large B-cell lymphoma of the breast: looking at pathogenesis, clinical issues and therapeutic options. *Ann Oncol* 24:2236–2244
4. Brogi E, Harris NL (1999) Lymphomas of the breast: pathology and clinical behavior. *Semin Oncol* 26:357–364
5. Lamovec J, Jancar J (1987) Primary malignant lymphoma of the breast. Lymphoma of the mucosa-associated lymphoid tissue. *Cancer* 60:3033–3041
6. Venizelos ID, Tatsiou ZA, Vakalopoulou S, Mandala E, Garipidou V (2005) Primary non-Hodgkin's lymphoma arising in an intramammary lymph node. *Leuk Lymphoma* 46:451–455
7. Anne N, Pallapothu R (2011) Lymphoma of the breast: a mimic of inflammatory breast cancer. *World J Surg Oncol* 11:125
8. Wiseman C, Liao KT (1972) Primary lymphoma of the breast. *Cancer* 29:1705–1712