CASE REPORT

Bilateral breast swelling in a 23-year-old woman with Burkitt lymphoma

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Primary breast lymphoma (PBL) of the breast is a rare type of localized non-Hodgkin lymphoma, which is usually of the B-cell. The majority of breast lymphoma present as a unilateral painless breast masses in an older woman, average age at diagnosis 55-60. A less common but distinctive presentation is a young woman of childbearing age who presents during or immediately after pregnancy. We present a 23-year-old postpartum woman with bilateral breast swelling. After surgical drainage and debridement and pathologic examination, the diagnosis of breast Burkitt lymphoma (BL) was confirmed. Chemotherapy was immediately started for her and the patient showed a good response with complete remission.

Key words: Breast swelling, Burkitt lymphoma, postpartum

INTRODUCTION

Burkitt lymphoma (BL) is a highly aggressive non-Hodgkin lymphoma (NHL) of the B-cell.[1] Three distinct clinical forms of BL are recognized: Endemic, sporadic, and immunodeficiency-associated. Although they have differences in their clinical presentation and geographic distribution, they have similar morphology and immune phenotypic features, and all possess translocation and deregulation of the c-Myc gene which is most commonly resulted from t (8;14), although variant translocations such as t (8;22) and t (2;8) have been described.[2] Non-Hodgkin's lymphoma may originate in, or spread to, any extranodal organ.

Breast lymphoma is a rare disease, either as a primary site or as secondary involvement, representing 0.04-0.5% of malignant breast tumors.[3] It is almost always of non-Hodgkin's type. Secondary involvement of the breast in patients with diffuse disease is more common.[4]

Most patients with primary breast lymphoma (PBL) develop a distant disease to other regions. Within the breast, the most common primary lymphomas are B-cell (more rarely T cell) non-Hodgkin's lymphoma.[3] The specific criteria to 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 from 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.[5]

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A 23-year-old woman suffered from redness, pruritus and tightness of her breasts in the last trimester of her pregnancy. Symptoms were considered to be related to hormonal change in pregnancy, but her problems aggravated after delivery. She also complained of fever and painful rapid enlargement of her breasts [Figure 1]. While feeding her child a large amount of serosanguinous fluid drained from her breasts. This time she was admitted in a general hospital under the diagnosis of breast abscess.

Physical examination showed tenderness, swelling, redness and ulcer in both breasts and right side axillary lymphadenopathy. No other abnormalities were found on examination and significant laboratory findings are shown in Table 1.

A course of antibiotic treatment was started for her. Incision and drainage were done by a surgeon, but

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Table 1: Results of hematologic and serum chemistry tests

Variable	Reference range, adults	On admission
Hematocrit (%)	36.0-46.0 for women	33
Hemoglobin (g/dl)	12.0-16.0 for women	12/1
White-cell count (per mm^3)	4500-11,000	15,400
Platelet count (per mm^3)	150,000-350,000	308,000
Prothrombin time (s)	11.1-13.6	12
Prothrombin time (international normalized ratios)		1.3
Partial-thromboplastin time (s)	22.1-34.0	33
Erythrocyte sedimentation rate (mm/h)	1-25	182
Glucose (mg/dl)	70-110	122
Creatinine (mg/dl)	0.6-1.5	0.8
Alkaline phosphatase (U/l)	30-100	334
Aspartate aminotransferase (U/liter)	9-32	27
Alanine aminotransferase (U/l)	7-30	15
Lactate dehydrogenase (U/l)	110-210	4415
Uric acid (mg/dl)	2.5-7	10.7
Na (meq/l)	135-145	142
K (meq/l)	3-5.5	3.8

the patient showed no clinical improvement. Breast biopsy was done and the result was: Sheet of medium size neoplastic cells with large nuclei, multiple distinct nucleoli, amphophilic cytoplasm and frequent mitosis suggestive for malignant lymphoma probably Burkitt type [Figure 2]. The patient was referred to the surgical oncologic service in Emam Khomeyni Hospital in Sari, Mazandaran, Iran in 2012.

Bone marrow aspiration and biopsy (BMA/B) was done and immunophenotyping revealed a large mononuclear cell population with vacuolated dark-blue cytoplasm about 16% of total cell, immunophenotyping expressed CD19, CD20, HLA-DR (human leukocyte antigens), but negative about CD10, consistent with B-cell lymphoma [Figure 3]. Brain magnetic resonance imaging was normal; chest computed tomography (CT) showed bilateral breast's tissue swelling. Abdominopelvic CT showed the small amount of fluid in abdominal cavity and bilateral kidney enlargement with heterogeneous enhancement.

Chemotherapy with CODOX-M/IVAC (Cyclophosphamide, Oncovin, Doxorubicin, Methotrexate, Ifosfamide, Vepesid and Ara-C) regimen started for her, and the patient showed complete remission on BMA/B, cerebrospinal fluid analysis and clinical course.

DISCUSSION

The term "PBL" is used to define malignant lymphoma primarily occurring in the breast in the absence of previously detected lymphoma localizations. Breast lymphoma is a rare

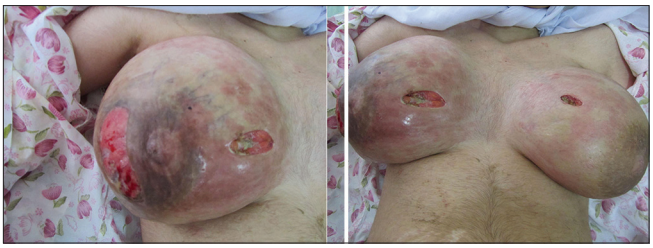

Figure 1: Burkitt lymphoma involving both breasts

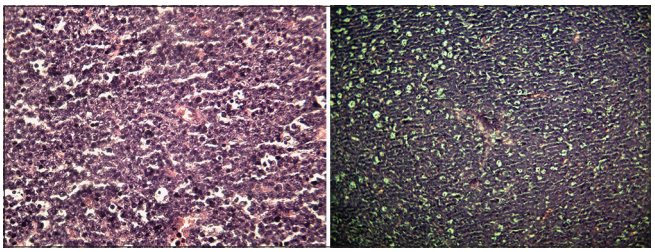

Figure 2: Burkitt lymphoma showing the "starry sky appearance" (H and E stain)

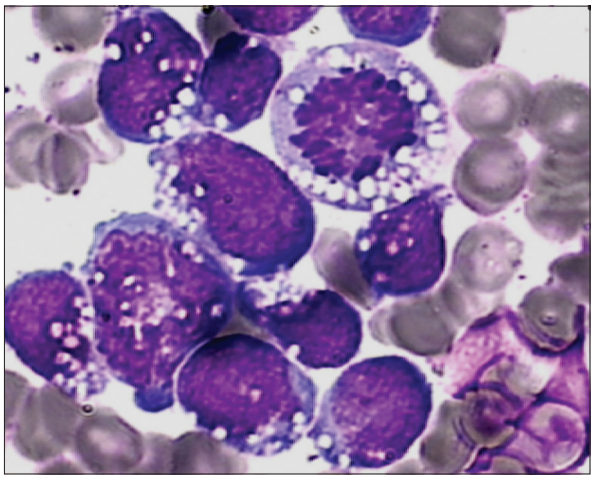

Figure 3: Bone marrow aspiration large mononuclear cell population with vacuolated dark-blue cytoplasm

disease, either as a primary site or as secondary involvement, representing 0.04-0.5% of malignant breast tumor.[3] PBL of the breast is a rare type of localized NHL, which is usually of the B-cell. It consists only 1% of primary breast tumor, and 2% of extranodal NHL.[6] The most frequent histopathologic types are: Diffuse large B-cell lymphoma which accounts for up to 50% of all PBL, follicular lymphoma 15%, mucosa-associated lymphoid tissue (MALT) lymphoma 12.2%, BL and Burkitt-like lymphoma 10.3%. Other histological types of PBL include marginal zone lymphoma, small lymphocytic lymphoma, and anaplastic large cell lymphoma.[7] Secondary involvement beyond the breast in patients with diffuse disease is more common.[4,8] The majority of breast lymphoma present as a unilateral painless breast masses in an older woman (average age at diagnosis 55-60) and is usually B-cell non-Hodgkins lymphoma.[9-11] 10% of cases are bilateral, which make it essential to assess contralateral breast. For unknown reasons, the right breast is involved more often than the left. Ipsilateral

axillary lymphadenopathy is present in 30-40% of cases.[9,12] A less common but distinctive presentation is a young woman in childbearing age who presents during or immediately after pregnancy and is a Burkitt-type lymphoma.[12,13] Disease is often bilateral and may clinically mimic inflammatory breast cancer. Breast lymphoma has been reported in men, although they are rare. Systemic "B" symptoms (i.e., fever, weight loss, night sweats) are uncommon.[14] On physical examination, PBL frequently appears as a benign or less suspicious lesion.[10,15] PBL usually presents as a single palpable mass that is mobile and contender. Multifocal tumors or diffuse breast involvement also was reported.[16] Clinical signs of advanced breast malignancy are rare, such as inflammatory changes, nipple involvement, skin retraction, or tumor fixation.[12] Most patients with primary lymphoma of the breast develop the disease to other sites. Early diagnosis is crucial for outcome.[17] PBL of the breast remains a diagnosis of exclusion a high index of suspicion and an understanding of the clinical behavior of PBL are necessary for proper patient management,[3] and the diagnosis cannot be made without a complete evaluation.[5] If a patient present with a rapidly growing breast tumor, lymphoma should be considered before any surgical intervention is performed. Early decision is vital considering the aggressive nature of the lesion and the prognosis.[3] A distinct mammographic or sonographic pattern has not been reported on the literature because primary lymphoma shows no specific characteristics, which differentiate it from other benign and malignant breast disorders.[4] Mammography usually demonstrates a well-circumscribed, uncalcified mass with sharp or minimally irregular margins.[18] Survival rates reported in PBL varied widely. Some authors have reported 5-year survival rates up to 89% and 50% for stages I and II, respectively.[19,20] Others have reported PBL to have a poor prognosis irrespective of treatment with median survival time as low as 12 months.[21,22] Local control is excellent with radiotherapy or combined modality treatment but systemic relapses, including in the Central nervous system (CNS), still occur frequently.[23] In this case, we chose CODOX-M/IVAC regimen and the patient exhibited complete remission. Five month later, while on maintenance therapy, the patient returned with fever and pancytopenia this time in BMA/B showed more than 50% blast, and the patient was treated as acute leukemia with DVP (Daunorubicine, Vincristine, Prednisone) Regimen. This time the patient did not respond to chemotherapy and 1 month later she died from treatment-related complication and sepsis in ICU.

CONCLUSION

BL of the breast is a rare neoplasm during the pregnancy and postpartum period but highly invasive and associated with poor prognosis if left untreated. PBL is a diagnosis of exclusion. Usually, it takes time to confirm the diagnose BL of breast because patient's symptoms considered to be related to pregnancy, breast engorgement and hormonal change during the lactation period. Due to highly invasive nature of BL, most patients are diagnosed in high stages or even postmortem. So in any pregnant or lactating woman, who complains of abnormal bilateral breast enlargement, the physician should think about BL or other breast cancers. Diagnostic procedure should be done, and the treatment should start as soon as possible.

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