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Case Report

A Case Report of Pancytopenia that Unmasked Primary Breast Lymphoma: A Rare Extra Nodal Lymphoma

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Abstract

Primary breast lymphoma (PBL) is an exceedingly rare entity. Review data suggest that this tumor is scarcely representing the 0.04-0.5% of all reported breast malignancies, and it does constitute a very low percentage of non-Hodgkin's as well as extranodal disease, less than 1% and 2%, respectively [1]. It affects mainly women around the 7th decade of life. The majority of cases are located unilateral, with an 11% affecting both glands [2]. The most common histological type is diffuse large B cell lymphoma (DLBCL), while follicular, MALT and Burkitt-like lymphoma are following [3]. The clinical presentation may mimic that of carcinoma, though frequently appears as a benign lesion and presents as a single palpable mass that is mobile and nontender. Pathologists' evaluation of the biopsy specimen along with immune-histochemical staining poses the final diagnosis. Data regarding treatment regimens are conflicting, since there is no particular consensus [4]. This case presentation illustrates a female patient aged 70 years-old, who visited the emergency department complaining of fatigue. Clinical, laboratory and imagine studies were performed and the patient was eventually diagnosed with primary breast lymphoma; with histological characteristics compatible with marginal zone lymphoma. The disease had already spread to bone marrow. The patient was immediately referred to hematology department and chemotherapy was initiated.

Keywords: Breast lymphoma; Marginal zone B-NHL; Pancytopenia; Lmphadenopathy; Splenomegaly

Introduction

Primary breast lymphoma is a malignant lymphoma, where the clinical site of presentation is the breast tissue, in the absence of previously detected lymphoma or evidence of widespread disease. However, ipsilateral lymph node involvement may be present and develops simultaneously with the primary lesion [5]. It consists only the 0.4-0.5 of breast malignancies, 0.38-0.7% of non-Hodgkin lymphomas (NHL), and 1.7-2.2% of extra nodal NHL [1]. Clinical signs such as inflammatory changes, nipple involvement, skin retraction and tumor fixation are exceptionally rare, since these are characteristics of different types of advanced breast malignancies [6]. Hormonal fluctuations may play a critical role in tumor growth, since it has been principally observed during pregnancy or postpartum [6]. Where do the lymphocytes, that give rise to lymphoma, originate from is inconclusive. Mucosal-associated lymphoid tissue (MALT), lymphatic tissue present adjacent to ducts and lobules within the breast or intramammary lymph nodes may be the 'birthplace' of PBL [7]. The therapeutic plan varies since there is no standard consensus treatment. As adjuvant or primary therapy have been used various schemes of chemotherapy and radiation therapy. Besides, surgical therapy has ranged from excisional biopsy to radical mastectomy. Although these treatments approaches can be applied alone or in combination, it has been demonstrated that extra-nodal lymphomas have better outcomes when treated with chemotherapy. On the other hand, mastectomy offers no benefit in the treatment of PBL. Survival rate fluctuates widely. Five-year survival rate ranges from 49% to 78%, depending on the stage of the disease [8]. Herein, we present a female patient, aged 70 years-old, who was initially admitted to the internal medicine department due to profound fatigue for the last two months and she was finally diagnosed with PBL. Since today, she has received 4 cycles of chemotherapy with the agents that are included in the CHOP regimen; cyclophosphamide, hydroxydaunorubicin, oncovin, prednisone, in addition to rituximab.

Case Presentation

A 70-year-old woman presented to the emergency department complaining of persistent fatigue during the last period. Past medical history included surgical excision of uterine fibroids, 25 years ago. Her hospitalization at that time was complicated by deep vein thrombosis and pulmonary embolism. Medication list involved rivaroxaban. Her vital signs were the following; blood pressure 130mmHg over 80mmHg, heart rate 80bpm, oxygen saturation 97% on room air and temperature of 36.7C. The patient was alert and oriented to time and place. Muscle strength and tone were normal. The cranial nerves were intact. The gait was steady and Romberg sign was absent. On cardiac auscultation, the heart sounds were audible, rhythmic and no murmurs or gallops were noted. There was no jugular venous distention and no lower extremity edema. Lung auscultation was normal, while abdominal examination revealed splenomegaly. Bowel sounds were present and there was no tenderness or rebound. A mass was palpated in her left breast at the left upper quadrant. When she was asked if she has noticed that before, she said that she discovered the mass 6 months ago, but thought that this was due to a trauma. No inflammatory changes or retraction of the skin were noted. Palpation of the lymph nodes revealed a palpable axillary node ipsilateral to the breast mass, while inspection of the skin revealed purpura at the lower extremities. 12-lead electrocardiogram showed sinus rhythm. The chest X-ray was normal. The abnormal laboratory findings were; WBCs 4.750, Hct 29, Hb 9.3 mg/dl, PLTs 18.000, C-reactive protein (CRP) 35 mg/L, Lactate dehydrogenase (LDH) 387 U/L. She was admitted to the internal medicine department and computed tomography of the chest and abdomen was ordered, which revealed lymph nodes of the chest and abdomen, splenomegaly (18,7cm) and a solid mass on the left breast. At that time, the following thoughts were made; is it an hematological malignancy, is it a breast cancer or these two conditions are connected to each other. Hematological and surgical consultation was asked. Bone marrow biopsy and core needle biopsy of the breast mass were conducted. The histology of the bone marrow and breast mass revealed non-Hodgkin lymphocytes of marginal zone. Lymph node biopsy of the axillary node displayed neoplastic lymphocytes of marginal zone too. After a thorough discussion with the hematologists it was deduced that this was an extra-nodal lymphoma, primarily sited on the breast tissue. R-CHOP chemotherapy was started according to the current bibliography although no standard scientific consent is present.

Discussion

Even though breast cancer is one of the most common causes of malignancies worldwide, lymphoma originating in breast tissue is indeed infrequent. Non-Hodgkin lymphoma located at the breast gland is either primary extra-nodal disease or secondary localization in the setting of a systemic disease. From all breast malignancies and extra-nodal non-Hodgkin's lymphomas, a percentage much less than 0.5% and 1.7% respectively, corresponds to PBL [1]. In 1972, the clinicians Wiseman and Liao, suggested specific criteria in order to differentiate and categorize the primary from non-primary lymphomas of the breast. The classification is as follows; a. findings of adequate pathological lesions at the histological specimen, b. the presence of mammary tissue and lymphomatous infiltrate, c. no preexisting lymphoma, d. ipsilateral axillary involvement may also be present [9]. Women are most commonly affected, while the age distribution is bimodal; one peak in the middle of the fourth decade and another in the seventh decade of life. It preferentially affects the right breast. Nonetheless, bilateral synchronous breast lymphoma appears in 10% of patients and in up to 15% of cases do develop contralateral metachronous disease [2]. Physical examination reveals a palpable mass that mimics breast carcinoma. Although, nipple discharge and skin retraction are not common in PBL cases, skin fixation and cutaneous inflammatory changes have been reported. Thus, it is impossible based on physical exam findings to make the diagnosis of breast lymphoma. The most common finding in mammography is a solitary noncalcified mass with circumscribed or indistinct margins. Diffuse increased parenchymal density with or without skin thickening may be seen on mammogram of patients with high grade lymphoma. However, there are no pathognomonic radiological findings as well for the differential diagnosis between breast lymphoma and carcinoma. Ultrasound and magnetic resonance (MRI) identify any solid breast mass, but the ability to differentiate between primary and secondary lesion is lacking. Histology is the gold standard for PBL diagnosis [10]. Malignant lymphomas of the breast are subdivided into diffuse large B-cell lymphoma, Burkitt lymphoma, extranodal marginal-zone-B cell lymphoma of MALT type, and follicular type. The most common is DLBCL. Staging and histological type determine the prognosis [11]. Young age, tumor size > 5cm, synchronous bilateral and or axillary nodal involvement are poor prognostic factors, while low grade and MALT lymphomas have a better prognosis than DLBCL [12]. Due to the rarity of PBL there is no general agreement regarding treatment; chemotherapy according to histological subtype and few of the cases radiation adjuvant to chemotherapy are the main treatment modalities. Central nervous system (CNS) involvement has been reported in some cases of aggressive disease, thus many authors suggest CNS prophylaxis in such cases [13].

Conclusion

To summarize, PBL is a distinct and exceptionally rare clinical entity that is documented only by histology. Clinical and radiological findings are not specific for the disease and in the majority of cases PBL mimics breast cancer. Nonetheless, the clinician should include lymphoma in the differential when a mass is lacking spiculations, micro-calcifications or architectural

distortion. Although no clear consensus has been established, the current treatment consists of chemotherapy combined or not with radiation therapy, with surgical approach constituting an ‘’old remedy’’, since offers no benefit in regards to survival or recurrence risk.

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