Case Report

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Primary Breast Lymphoma with Inflammatory Appearance: A Rare Case Report

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Abstract

Primary breast lymphoma (PBL) is an uncommon and curable disease. It usually presents as a clinically palpable mass and is rarely detected in screening. Although the imaging characteristics are not specific, they may sometimes mimic benign masses. However, the clinical and imaging findings in PBL can mimic those of breast carcinoma, and sometimes, its presentation is suggestive of inflammatory breast cancer. Diagnosis depends on adequate tissue sampling for histology examination and immunophenotyping. Unlike primary breast carcinoma, surgery is not the key treatment for PBL. Treatment is mainly confined to combination of chemotherapy and radiation therapy.

Keywords: Inflammatory breast; Breast lymphoma; Diffuse large B cell lymphoma

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Introduction

Primary breast lymphoma (PBL) is a rare manifestation of extranodal non-Hodgkin's lymphoma. It was first described in 1972 by Wiseman and Liao, [1] in a group of 31 patients diagnosed between 1951 and 1970, defining it as the infiltration of breast tissue by lymphoma with or without regional lymph node in patients without a history of prior nodal or extranodal lymphoma and systemic disease at the time of diagnosis. These criteria were reviewed in 1990 by Hugh et al. [2].

We report a case suspected of an inflammatory breast cancer that was diagnosed as primary breast diffuse large B cell lymphoma (PBDLBCL), and discuss diagnosis and management as well as a review of the literature.

The application of Game is diverse and can be applied in: (1) Medicine Practice (2) Students of Medical Science (3) Research and Development of Medical Science.

Case report

This is a 78-year-old woman with a history of hypertension who consults for a small, painless right axillary mass gradually increasing in volume. The clinical examination found a patient in good general condition, PS at 0, infra-centimetric lymph nodes along the right axillary extension with redness and inflammation of the ipsilateral breast, realizing the appearance of orange peel. The lymph node biopsy found a proliferation with malignant appearance with a ganglionic architecture erased by a medium-sized lymphocyte population, made of rounded elements with high nucleocytoplasmic ratio; the nucleus contains a dense chromatin. The mitoses are numerous. This lymphoid proliferation tends in places to cross the capsule and diffuse into the perinodular adipose tissue. Immunohistochemistry found markers profile as follow: CD20+, CD5-, CD10-, Anti-Bcl2-, anti-Bcl6+, anti-MUM1+ with a Ki67 which marks 90% of the tumor cells, concluding to a diffuse large B-cell lymphoma not centro-germinal.

The cervico-thoraco-abdomino-pelvic CT performed as part of the extension assessment found a voluminous mass of the right breast measuring 89mm x 73mm with polylobed contours and clear limits, its center is hypodense and not containing visible calcifications.

The patient received six courses of R miniCHOP alone (Rituximab, Cyclophosphamide, Doxorubicin, Vincristine, and Prednisone). Pet-scan evaluation at the end of treatment shows a complete metabolic remission after 12 months follow-up.

Discussion

The incidence of non-Hodgkin lymphoma (NHL) has increased in the last decades, particularly for extra-nodal sites [3]. Breast involvement by lymphoma is very rare, and it can occur as a primary breast tumor or as an extra-nodal manifestation in systemic disease [4]. PBL represents 1-2% of extra-nodal lymphomas [5]. In addition, breast lymphoma accounts for approximately 0.04-0.5% of malignant breast tumors [6,7]. This rarity may be explained to the fact that the breast contains very little lymphoid tissue [8]. With the advancement in diagnostic modalities, the incidence of PBL is increasing and about 95%-100% of reported cases are diagnosed in women, while it is very rare in men [9]. Unilateral involvement of the upper right quadrant of the breast is the most common reported presentation of PBL [9] but the explanation for this remains unclear [10]. Only 1% to 14% of all reported PBL cases have bilateral disease [10].

Usually, PBL is presented as a palpable mass in the breast that may or may not be accompanied by painless axillary lymph nodes, rapidly expanding which makes it clinically difficult to differentiate from a breast carcinoma. However, sometimes, PBL presentation is suggestive of inflammatory breast cancer, which is the case of our patient.

The imaging findings of PBL are non specific and may resemble to any other breast malignancy or sometimes may have a more benign appearance. The most common mammographic abnormality is a solitary non-calcified breast mass with circumscribed or indistinct margins. Calcifications, architectural distortion, nipple retraction, or spiculations are usually not seen in association with this malignancy. Global asymmetry may also be a mammographic presentation of PBL, seen in one-third of the patients in the study by Sabate et al, and usually associated with high-grade lymphomas [6]. In one of the largest series by Liberman et al. of 32 cases of NHL in 29 women (66% classified as PBL), the most common mammographic finding was a solitary noncalcified mass (69%). Multiple masses were seen in 9% cases, diffuse increased opacity with skin thickening in 9%, and no mammographic abnormality was seen in 13% cases. Multiple masses are more common with secondary breast lymphoma (SBL), compared to primary [4].

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. Indolent histologies such as follicular non- Hodgkin's lymphoma or extranodal marginal zone (MALT) lymphoma occur less commonly. However, breast involvement with Hodgkin's disease or T-cell tumor is very rare.

Treatment of primary breast lymphomas follows treatment recommendations for lymphomas of the same stage and histology in other locations. There are no specific guidelines for PBL management. Performing mastectomy is not the primary line of management of PBL. Several studies found that mastectomy offered no benefit in the treatment of primary breast lymphoma [10], and role of surgery should be limited to a biopsy to establish the correct histological diagnosis, leaving the curative treatment to chemotherapy and radiotherapy. The choice of chemotherapeutic regimen should be based upon histological subtype, disease extent and the individual patient. The treatment of PBDLBCL might include combination of chemotherapy, and radiotherapy. CHOP or CHOP-like anthracycline-based chemotherapy combined with rituximab is now considered the standard treatment for most DLBCL breast lymphomas. This may be followed with radiation to the ipsilateral breast and regional nodes. In the study by Avilés et al. the 10 year overall survival in patients treated with either radiation or chemotherapy alone was 50% vs 76% in those who received combination of radiation and chemotherapy.

The role of central nervous system (CNS) prophylaxis in DLBCL of the breast is controversial. There have been no prospective trials of CNS prophylaxis in this population. Case series have reported a high incidence of CNS recurrence, estimated at 12 to 27%. Given this high incidence, central nervous system (CNS) prophylaxis should be considered [10]. In Rayan et al.'s patients, one unexpected finding was that the risk of CNS relapse was relatively low, occurring in only 5% of patients. This contrasts with the reports of other smaller studies finding considerably lower than the risk seen in primary testicular DLBCL. It may be that primary breast DLBCL does not have the same tropism for CNS as testicular DLBCL, and this difference explains the generally superior survival for patients with primary breast DLBCL compared with that of patients with primary testicular DLBCL. However, it is possible that limiting the eligibility to patients with localized disease has led to underestimation of the rate of CNS involvement. This result should be considered with caution and the authors suggested using CNS prophylaxis to avoid central nervous system involvement.

Conclusion

Clinicians who are managing breast cancers must be aware of this rare entity to detect its clinical presentation and management which is totally different from breast carcinoma cases. The imaging features are not specific, but the radiologists must be aware of the imaging presentation of such rare entity to recommend appropriate management and establish radiological-pathological concordance.

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