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Clinical Hematology

About A Case: Primary Breast Lymphoma (Diffuse Large B cell Lymphoma)

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Abstract

Case Report

Primary breast lymphoma is a rare neoplasia, with little clinical and imaging specificity. These are essentially type B lymphomas, of which the most common is diffuse lymphoma with large B cells. It is an entity that presents a prognostic impact from the outset by their localization but also by the risk of neurological relapse. The treatment of primary breast lymphoma depends on lymphomatous localizations and of course the histological type. There remain some questions to be raised regarding the place of radiotherapy in the treatment of certain localized histological forms, as well as the interest of systematic neuromeningeal prophylaxis in this breast localization. We report a case of primary breast lymphoma in a 50-year-old patient with no particular history. **Keywords:** Primary lymphoma, Breast cancer, DLBCL.

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1. INTRODUCTION

Primary breast lymphomas are rare, about less than 1% of all non-Hodgkin lymphomas and 2% of extra-node lymphoma. These are mainly type B lymphomas, very rarely type T or NK. The most common are diffuse large B cell lymphomas, the least common are small B cell lymphomas, especially follicular type or marginal lymphoma "mucosaassociated lymphoid tissue" known as MALT.

Primary breast lymphomas are a rare entity that has a prognostic impact from the outset by their location but also by the risk of neurological relapse. We report a case of primary breast lymphoma in a patient with no particular history.

2. OBSERVATION

This is Patient B.M, 58 years old, housewife, married and mother of 3 children. Menopausal without hormone replacement therapy, and with no medical personal or family history, she had consulted in July 2018 for nonpainful right breast nodule without other associated signs.

The senological examination had found a breast mass of 2 cm at the upper-internal quadrant of the right breast, not painful, without inflammatory signs in sight. Examination of the lymph nodes did not show any lymphadenopathy and the left breast showed no abnormalities at the clinical examination.

Considering the ACR 4 result of the mammogram, a micro biopsy was performed. As the result was inconclusive, it was completed by a lumpectomy. The anatomopathological analysis showed a lymphomatous tumor proliferation made of non-cohesive cells with a basophilic cytoplasm, an increased size nucleus and a dense chromatin along with very frequent mitoses.

The immunohistochemical study showed tumour cell positivity for CD 20, BCL 6, and negativity for CD 10, TdT, BCL 2, CD 5, and MUM 1. The proliferation index assessed by Ki67 was 97%. These tests confirmed the diagnosis of the diffus large B cell lymphoma with a germinal centre phenotype.

The extension assessment did not reveal any other tumor location. The PET-scan showed a very intense hypermetabolic lesion (SUV 11.8) in the superior-internal quadrant of the right breast without any other suspicious focus (Fig. 1 and Fig. 2).

The osteomedullary biopsy showed no marrow infiltration. The lumbar puncture did not reveal any meningeal involvement. The biological work-up showed no abnormalities: blood count was correct, LDH was 172. protein electrophoresis was flawless.

Considering the staging established according to the age, the LDH rate, and the Ann Arbor classification system, The Prognostic Index (IPI) and age-adjusted IPI (aaIPI) were both calculated for our patient. The result was 0 and therefore of low risk.

The therapeutic decision was adjuvant immunotherapy to chemotherapy. Thus, the patient received Six cycles of combination chemotherapy with cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP) treatment combined with six doses of rituximab given every 21 days with 3 intrathecal injections of methotrexate for central nervous system (CNS) prophylaxis.

The evolution was marked by a complete metabolic remission obtained and maintained clinically, biologically and radiologically throughout the monitoring period of our patient for four years so far, noting no relapse.



Fig. 1: PET scan axial slice, Lesion Hyper metabolic (SUV 11.8) right breast



Fig. 2 (a, b): PET scan, Hyper-metabolic lesion of the upper-internal quadrant of the right breast

3. DISCUSSION

Breast lymphomatous neoplasia is rarely primary. To speak of primary breast lymphoma, it is necessary to meet the criteria of Wiseman and Liao:

- Obtaining sufficient tissue material to perform a complete histologic study
- Presence of mammary glandular tissue in the middle of the lymphomatous infiltrate or in direct contact with it
- Absence of concomitant lymph node lymphoma except for a possible homolateral axillary lymph node extension
- No known lymphoma in the medical history

This rare occurrence of breast lymphoma is often unilateral. Bilateral involvement is seen in 10% of cases. The typical clinical presentation would be a firm, painless mammary nodule, with the possibility of finding axillary adenopathy on clinical examination. In imaging, breast lymphoma has no typical or specific presentation. As with any breast nodule, a mammogram must be performed and a biopsy will be indicated. The diagnosis will be made by histopathological examination supplemented by an immunohistochemical study to define the type of lymphoma. The most common histological type is diffuse large B-cell lymphoma. Low-grade MALT lymphoma is the second most common.

The management of primary breast lymphoma requires an initial performance status PS assessment, a biologic workup by determining the LDH lactate dehydrogenase level, and an extension assessment, performed by the PET scan, to determine the stage and assess the post-treatment therapeutic response. Bone marrow biopsy is only required if the PET-CT does not show bone or marrow involvement. Exploratory lumbar puncture is indicated for breast localization.

The treatment of primary breast lymphoma depends on lymphomatous localizations and of course the histological type. For DLBCL, the most frequently observed entity, the use of chemotherapy combined with rituximab is imperative unless there are contraindications. Mastectomy-type surgery has no legitimacy for diffuse large B-cell lymphoma, to the same degree as radiotherapy, whose indication remains controversial and requires further research and studies to determine its effect on overall survival and the risk of relapse.

Neuromeningeal relapse in primary breast lymphoma, especially of the DLBCL type, is a rare but particularly serious event. Neuromeningeal prophylaxis is most often performed with intrathecal injection methotrexate, based on the breast localization but also the CNS IPI score.

4. CONCLUSION

Primary breast lymphoma is a rare entity, with poor clinical and imaging specificity. The diagnosis is based on histopathological study completed by immunohistochemistry. The treatment and the evolution of the disease are related to the histological type of lymphoma.

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