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Primary Breast Lymphoma

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Introduction

The term "primary breast lymphoma" (PBL) is used to define a malignant lymphoma primarily occurring in the breast in the absence of previously detected lymphoma localizations [1].

PBL is a rare disease, accounting for only 0.4-0.5% of all breast malignancies, 0.38-0.7% of all non-Hodgkin lymphomas (NHL), and 1.7-2.2% of extranodal NHL. The median age of patients with diagnosed PBL ranges from 60 to 65 years [1-12]. The disease occurs almost exclusively in women. Bilateral breast involvement accounts for 11% of all breast lymphomas [13] or 5% according to Ryan *et al.* [11]. This rare situation is especially observed during pregnancy or postpartum, suggesting that tumour growth is influenced by hormonal stimulation.

Breast lymphoid cells probably originate in mucosa-associated lymphoid tissue (MALT) [14]. PBL may also originate from lymphatic tissue present within the breast adjacent to ducts and lobules, or from intramammary lymph nodes [15, 16].

More than 80% of PBL are B-cell lymphomas, mostly CD20+. The most frequent histopathologic types are: diffuse large B-cell lymphoma (DLBCL) which accounts for up to 50% of all PBL, follicular lymphoma (FL) – 15%, MALT lymphoma – 12.2%, Burkitt's lymphoma (BL) and Burkitt-like lymphoma – 10.3% [17]. Other histological types of PBL include marginal zone lymphoma (MZL), small lymphocytic lymphoma (SLL), and anaplastic large cell lymphoma (ALCL).

Diffuse large B-cell lymphoma (DLBCL) is the most common histological diagnosis. These lymphomas have been shown to be of a non-germinal centre B-cell phenotype with a high proliferation index and are thought to be associated with a poor outcome [18]. There is a close association between ALCL and silicone breast implants [19, 20].

Burkitt's lymphoma is observed particularly in pregnant or lactating women and HIV-seropositive patients. The clinical presentation of PBL and the radiological features are usually no different from those of carcinoma of the breast. PBL is usually classified according to the Ann Arbor staging system. Other diagnostic criteria for PBL were described by Wiseman and Liao in 1972 [21]. According to the last classification, the clinical site of presentation is the breast. A history of previous lymphoma or evidence of widespread disease are absent at diagnosis. There is present close association of lymphoma to breast tissue in the pathologic specimen. Ipsilateral lymph nodes may be involved. This definition of PBL comprises only tumours being in stage I (lymphoma limited to the breast) and stage II (lymphoma limited to the breast and axillary lymph nodes) and not to tumours originating from non-breast sites.

Case Presentation

A 65 years old patient was admitted to our hospital in April 2016 with a mass 6 cm of the right breast, upper outer quadrant with a clinical diagnosis of breast carcinoma.The USG revealed a well circumscribed , no spiculated margins, hypoechoic mass . Detailed physical examination and ultrasonography (USG) revealed no axillary lymphadenopathy. Suspected for a primary breast carcinoma was performed radical modified mastectomy.

Histopathological and immunohistochemical findings in the tumour mass and lymph nodes confirmed the diagnosis of a **diffuse large B-cell type primary breast lymphoma**; CKAE1/AE3 negativ, CD 20+, CD 79a+, CD 3-, Bcl 2+, Ki 67 in 85% of neoplasmatic cells. There were no abnormalities in computerized tomography (CT) of the thorax and abdomen.

Photo 1, 2, 3, 4.

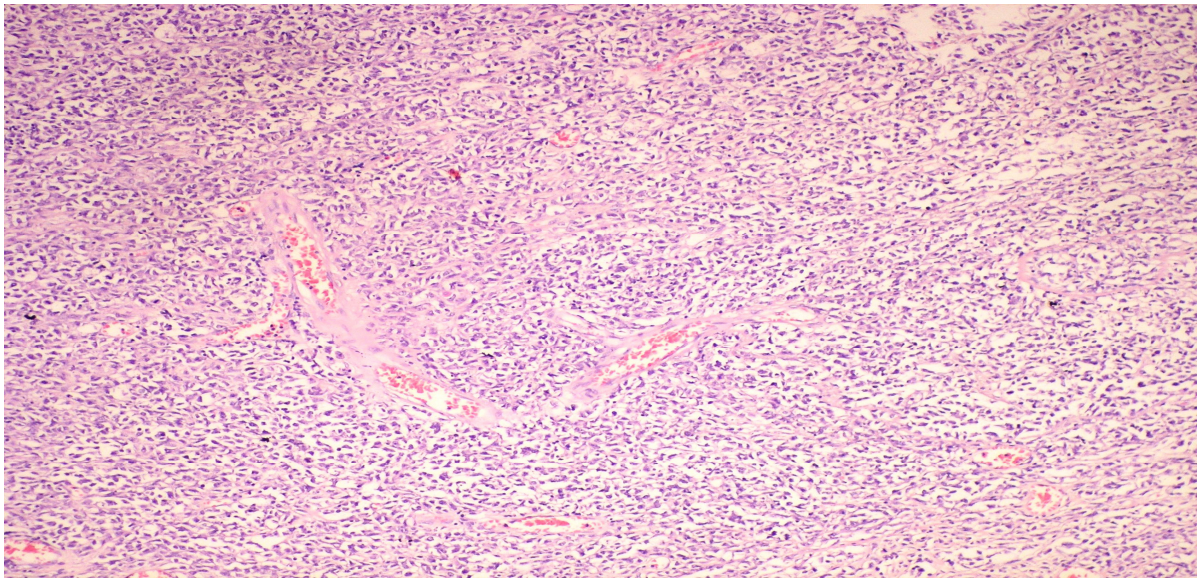

Photo 1. (HEx20)

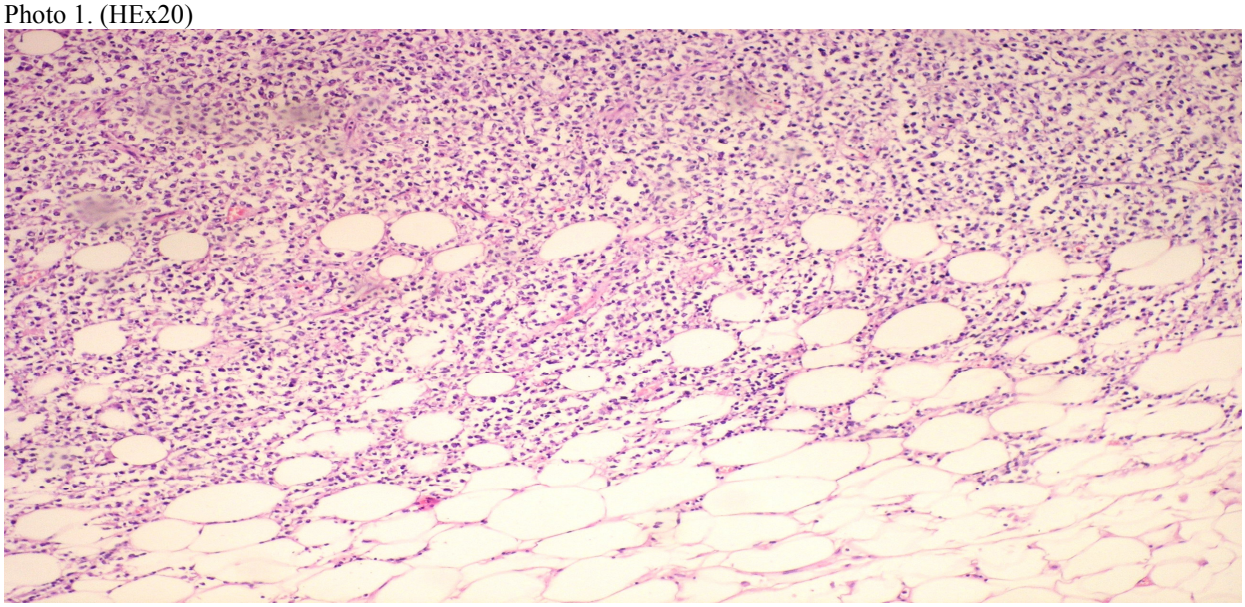

Photo 2. (Hex20)

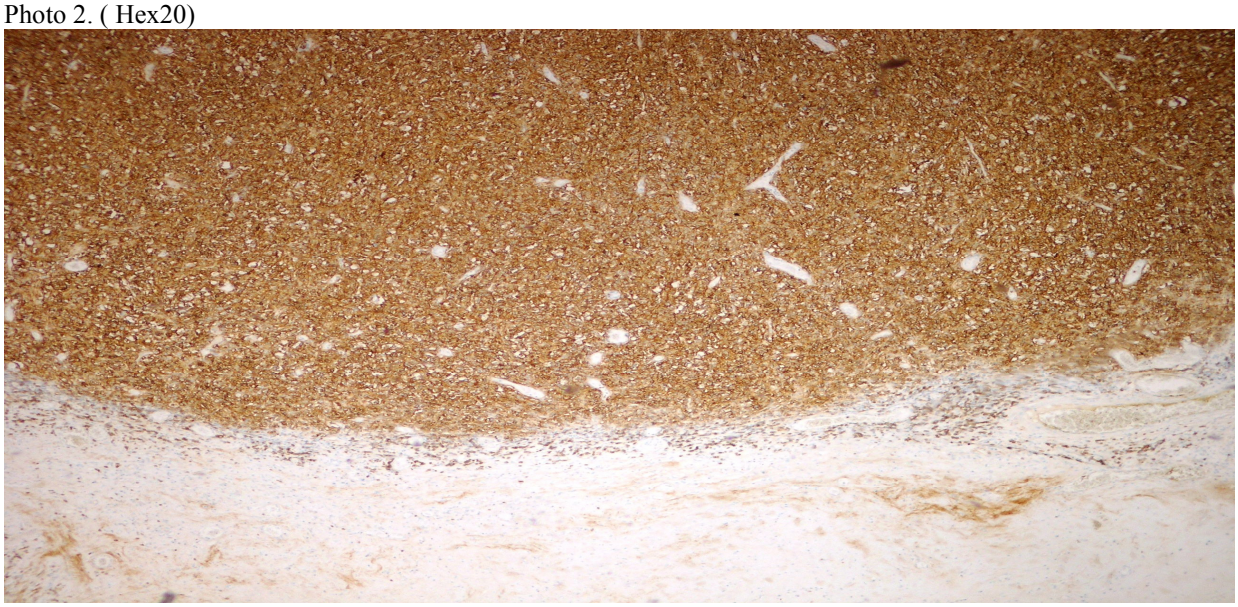

Photo 3. (CD20 x20)

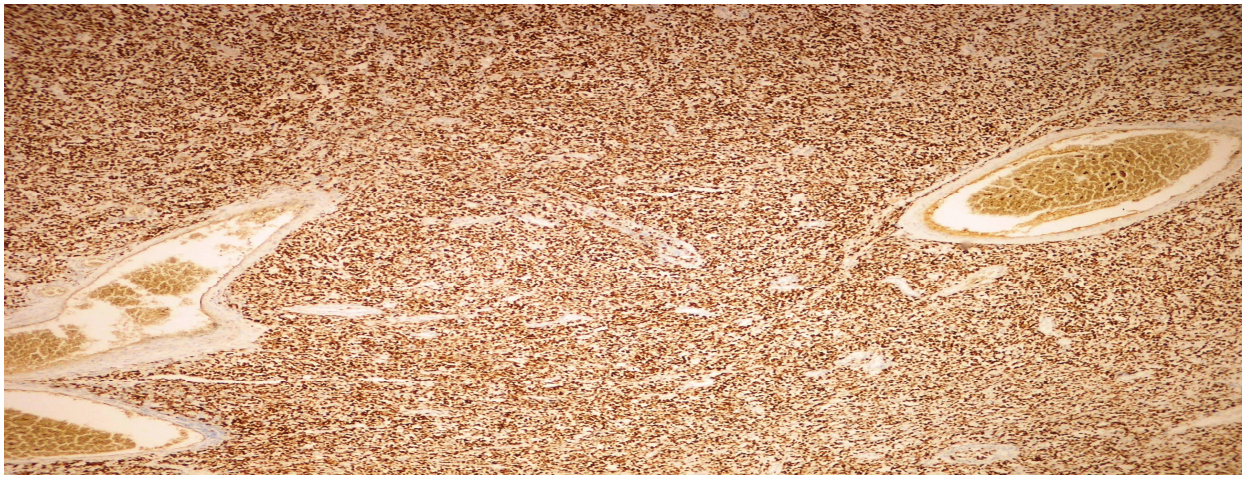
Photo 4. (Ki67 x20)

Discussion

A painless mass is the most common presenting sign in PBL [1] .Mammographic findings are nonspecific. The USG pattern of the mass is usually hypoechoic . No masses have spiculated margins or calcifications [22]. Fine needle aspiration, core biopsy and excisional biopsy are effective techniques used in the evaluation of breast nodules and axillary lymph nodes. However, histological, immunohistochemical and, sometimes, genetic studies are necessary for establishing the diagnosis.

The behaviour of primary lymphoma of the breast is thought to be similar to that of lymphomas of the same histological types and stages arising at other sites.

Mastectomy is not recommended because it offers no benefit as regards survival or recurrence risk.

Conclusion

Primary non-Hodgkin lymphomas of the breast, though rare, should be considered in the differential diagnosis of breast malignancies especially when you are in front of FNAB,
Tru-cut or Frozen biopsy.
Once this diagnosis is suspected in FNAB, it should be confirmed by multiple core biopsies, especially when the tumoral mass is big followed by Immunohistochemistry exams.
The differential diagnosis should be done with Invasive lobular carcinoma and other Lymphomas subtypes.
Once that the diagnosis is established with preoperative exams conservative treatment with chemotherapy should be done instead of radical surgery.

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