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Primary Lymphoma of the Breast

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1. Abstract

Primary Breast Lymphoma (PBL) is a rare entity that accounts for only 0.4 to 0.5 % of malignant breast neoplasms. It is characterized by the presence of a breast lump, with or without associated regional adenopathy and without systemic involvement. Available information regarding PBL are difficult to analyze and to compare because of the relatively small number of patients in most series and the substantial differences in criterions of classification that has been used, such as lymphoma subtypes, histopathologic terminology, treatment modalities and staging systems. There has been no known published documentation of Primary Breast Lymphoma in the Philippines. With its rarity, there is still no consensus on the best approach. This paper aims to present a literature review as well as to present a clinical case of a 41 year old female diagnosed with Primary Breast Lymphoma.

2. Introduction

Primary Breast Lymphomas (PBLs) have a reported incidence ranging from 0.04% to 0.5% of all breast malignancies [1]. It is characterized by the presence of a breast lump, with or without associated regional adenopathy and without systemic involvement [2]. PBLs account for less than 1% of all patients with non-Hodgkin's lymphomas and approximately for 1.7% of all extranodal non-Hodgkin's lymphoma [3]. The criteria for the diagnosis of primary breast lymphoma were suggested by Wiseman and Liao in 1972 include [4]: (a) adequate pathologic evaluation, (b) close association between lymphomatous infiltrate and mammary tissue and (c) exclusion of either systemic lymphoma or extramammary lymphoma, except simultaneous ipsilateral axillary node involvement. Available information regarding PBL are difficult to analyze and to compare because of the relatively small number of patients in most series and the substantial differences in criterions of classification that has been used, such as lymphoma subtypes, histopathologic terminology, treatment modalities and staging systems [3]. There has been no known published documentation of Primary Breast Lymphoma in the Philippines. Primary Lymphoma of the Breast is a rare type of Breast malignancy and that there has been few reports of such incidence in the Philippines. Aside from contributing to medical literature, we can share our experience with this case to the medical community. Being able to discuss the surgical course and management of this case at our local setting will aid surgeons that will encounter this rare type of malignancy. The objective of this case is to present a case of a 41-year old female diagnosed with Primary Lymphoma of the Breast and to discuss about its surgical course and management.

3. Case Presentation

A 41-year old female with a 13-year history of a left breast mass. The lesion initially began as a 0.5 cm mass on the left breast noted to be movable, non-tender and notably round shaped. On the interim, condition was tolerated. Progression of the growing breast mass prompted consult at the Outpatient Department. The roundly shaped mass was already noted to be 10 cm at largest diameter, mobile and non-tender. A core needle biopsy was done which revealed Chronic Mastitis, Poorly Differentiated Carcinoma, Lymphoma of the Breast. She was then advised for neoadjuvant chemotherapy and surgery. The patient has no known comorbidities nor any allergies. She has an unremarkable past medical history. She has familial history of breast carcinoma on her maternal side.

On examination, the patient was sthenic with normal systemic examination findings. On the left breast, she had a 5 cm x 6 cm in largest diameter, non-erythematous, non-tender, and irregular shaped mass (Figure 1).

There were no palpable axillary lymph nodes. Contralateral breast and the bilateral axilla were unremarkable.

Eight Cycles of neoadjuvant chemotherapy of Doxorubicin, Cyclophosphamide an Doxetaxel were given. Lumpectomy of the Left Breast mass was done under general anesthesia. A Tennis Racket Incision done (Figure 2). Patient was then discharged improved on the 4th post operative day and was advised surveillance follow-up as per outpatient basis.

On gross inspection, there consists of soft tissue mass (8 x 5.5 x 2 cm) with an attached ellipse of skin (6 x1.2 cm), and two separate smaller tissue fragments (4.5 x 2 x 1cm, and 2 x 0.6 x 0.6 cm). On sectioning, two ill-defined pale tan solid areas within the fatty breast tissue (5 x 4.5 x 1.4 cm and 2 x 2 x 1.5 cm) are noted.

The microscopic examination of the mass shows sections of diffusely proliferating population of monomorphic round cells. The individual tumor cells are small to intermediate in size, with moderate size variation. The nuclei are enlarged, hyperchromatic, with regular contours, surrounded by a thin, basophilic cytoplasm. The tumor cells form solid cohesive sheets with minimal stroma. Nerve axons are seen within the central portions of the solid sheets of the tumor. Indicative of perineural invasion. Zones of necrosis within the tumor is seen. Multinucleated giant cells are seen engulfing necrotic debris. (Figure 3 & 4). The final biopsy of the mass revealed residual Primary Breast Lymphoma, Status Post Chemotherapy.

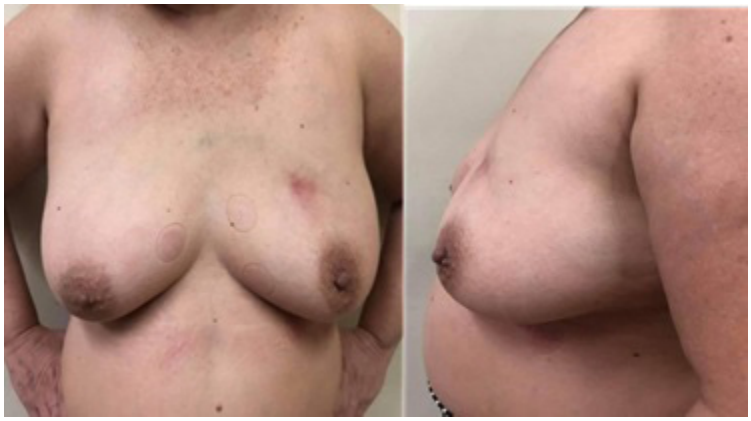

Figure 1: Left Breast Mass

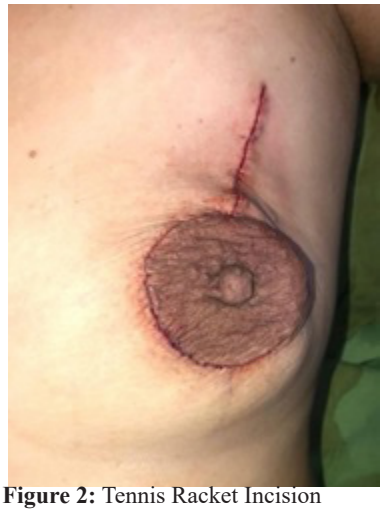

Figure 2: Tennis Racket Incision

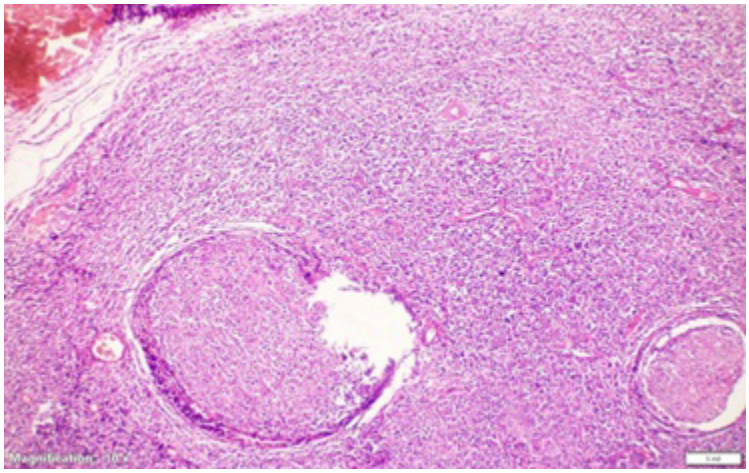

Figure 3: Section of the Patient's Breast Mass on H and E Stain at Low Power Field Magnification Note the diffuse proliferation of monomorphic round cells with perineural invasion.

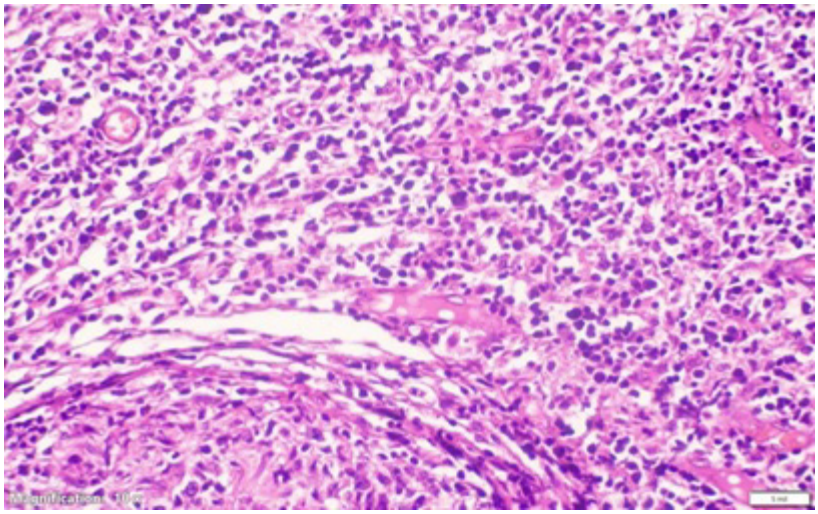

Figure 4: Section of the Patient's Breast Mass on H and E Stain at High Power Field Magnification. On Higher Magnification, the cells possess enlarged, hyperchromatic nuclei with moderate size variation and surrounded by scant basophilic cytoplasm.

4. Discussion

Non-Hodgkin lymphomas of the breast are uncommon cancers which occur either as primary extranodal diseases or as secondary localizations of a systemic disease [1]. Overall, the characteristics of this disease, such as natural history, prognostic factors and impact of treatment have not been yet well established [5]. Most studies reported a preponderance of right-sided presentation in breast lymphoma, but others reported the opposite [6, 7]. Bilaterality has been reported to occur in 1% to 14% of cases [6]. However, the patient presented with left sided breast mass only. The reported median age for breast lymphoma ranges from 51 to 60 years, but with a wide range [6, 7]. This patient is only 41 years old. Diffuse large B-cell lymphoma (DLBCL) is the most common histological subtype, representing 5% to 79% of all PBL cases [6, 7].

The immunohistochemical staining of the patient of CD20 showed to be strongly reactive, identifying B lymphocytes and different types of B cell Lymphoma and her CD45 also showed to be strongly reactive, identifying tumors of lymphoid origin. The clinical presentation and the imaging characteristics are no different from breast carcinoma. The presence of a palpable lump is the most common manifestation (61% of cases), usually painless and often located in the superoexternal quadrant. Other signs include: pain (12%), local inflammatory signs (11%) and palpable adenopathies (25%) [8]. In about 12% of cases, it appears as an incidental finding on mammogram as the patient is asymptomatic [8]. The imaging findings of breast lymphoma are nonspecific and may resemble any other breast malignancy or sometimes may have a more benign appearance. The most common mammographic abnormality is a solitary noncalcified breast mass with circumscribed or indistinct margins [9-11]. Global asymmetry may also be a mammographic presentation of PBL, seen in one-third of the patients [12]. On US a hypoechoic solid oval or round mass with circumscribed or indistinct margins is the most common appearance.9 PBL appears to be a rare disease with a good overall prognosis and low incidence of local relapse, following chemotherapy alone or in combination with other treatments [14]. Regarding management, surgery, radiotherapy, chemotherapy and immunotherapy have been used alone or in combination; however, there is still no consensus on the best approach. Chemotherapy, alone or combined, is the standard treatment [13]. To date, there has been no established clinical protocol or guidelines for both staging and management for Primary Breast Lymphoma [15]. No recommended surveillance measures have also been established to date.

5. Conclusion

Primary Breast Lymphoma (PBL) is a rare entity that accounts for only 0.4 to 0.5 % of malignant breast neoplasms. Available information regarding PBL are difficult to analyze and to compare because of the relatively small number of patients in most series and the substantial differences in criterions of classification that has been used, such as lymphoma subtypes, histopathologic terminology, treatment modalities and staging systems. With its rarity, there has been no established clinical protocol or guidelines for both staging and management for Primary Breast Lymphoma to date. Although there has been no established clinical protocol for the staging, medical and surgical management for Primary Breast Lymphoma, early diagnosis accompanied with the proper medical and surgical intervention are paramount to the diagnosis, course and treatment of this patient given its rarity.

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