Primary Mammary (Non-Hodgkin) Lymphoma of Breast: A Case Report

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Abstract

Primary breast lymphoma is a rare tumor that presents commonly as a large mass with no specific mammographic appearance and can be confused with poorly differentiated carcinoma on frozen section. With appropriate treatment, this breast malignancy has a relatively good prognosis. We report a case of primary mammary non-Hodgkin lymphoma (NHL) in a 30-year-old young female presented with the complaint of painless lump in the right breast. Fine needle aspiration cytology (FNAC) suggested malignancy. Investigations ruled out metastasis, hence mastectomy was done. Immunohistochemistry of specimen revealed NHL of breast. Investigations to rule out any other site of lymphoma proved it to be primary NHL of breast with no metastasis. Anticancer drugs are main treatment rather than surgery so it is very important to accurately diagnose primary lymphoma of breast.

Keywords: Non-Hodgkin lymphoma; Fine needle aspiration cytology; B-cell lymphoma; Contrast enhanced computed tomography

Introduction

Primary lymphoma is rare disorder of breast and constitutes less than 0.6% of all breast malignancies and 2.2% of extra nodal lymphomas [1,2]. Most breast lymphomas are the non-Hodgkin’s type, which represent approximately 70–90% [3]. In patients diagnosed with NHL, primary involvement of the breast is seen in 0.4–0.7% of the cases. Almost all primary breast lymphoma has a B-cell phenotype, while primary breast lymphoma with T-cell phenotype is extremely rare. In all, 46–71% of primary breast lymphoma is diffuse large B-cell lymphomas (DLBCL) [4,5]. Primary breast lymphoma is reported to exhibit a poor prognosis and the therapeutic management is controversial and is not fully established. We report a case that was diagnosed as malignant primary lymphoma of the breast and discuss diagnosis and management.

Case Report

30 years old female presented to the Department of Radiotherapy, J.A. Group of Hospitals, Gwalior, India, with the complaint of painless lump in the right breast from two month, swelling over right scalp from one month, headache and dizziness from one month. She denied associated pain, fever, chills, or skin changes. Both axilla and left breast were normal. Her medical history was unremarkable, and review of symptoms was negative for night sweats, weight loss, or fever. On examination patient was conscious, oriented and afebrile. The physical examination was notable for a large fungating mass, firm in consistency seen in the upper outer quadrant of right breast (Figure 1A). Left breast was normal. Swelling of soft tissue seen over right side of the scalp. No axillary lymphadenopathy is observed. The rest of the physical examination findings were normal.

Blood profile shows decreased haemoglobin and leucocytosis. FNAC of breast and scalp lesion showed it was a high grade malignant Non Hodgkin’s B cell lymphoma. Immunohistochemistry showed high grade Non Hodgkin’s B cell lymphoma.

Radiological findings

On Sonography nodular type is seen as well marginated hypoechoic lobulated lesion (Figure 1B), which is similar to nodular lymphoma elsewhere in body where as diffuse type well defined hypoechoic area replaces normal breast parenchyma and Color Doppler imaging showed low resistance blood flow (Figure 1C). Ultrasound of scalp lesion showed hypoechoic lesion with bone erosion (Figure 1D,E). Contrast Enhanced Computed Tomography (CECT) of brain showed metastasis in right frontoparietal region with culvarial defect (Figure 1F,G). X ray of skull shows solitary osteolytic lesion in right parietal bone (Figure 1H). Chest X-ray is normal. Pathological fracture is seen in right femur (Figure 1I). No lymphadenopathy is observed in ultrasound of abdomen.

After confirming the diagnosis the therapy plan included CHOP ( Injection Vincristin 1.5mg, injection cyclophosphamide 500mg, injection doxorubicin 50mg, Tab wysolone 100mg / 5 days) and standard dose given after completing the six cycle of chemotherapy. However at the time of diagnosis of PNHBL the CNS and bony metastasis was not directly involved but it was extra dural skull metastasis. Extra dural skull metastasis is highly vascular so the penetration of drug is very easy. That’s why it was not the proper time for surgery so that patient is subjected to chemotherapy. On physical examination there is a 90 -95% volume reduction of tumor mass and after that patient is subjected to total mastectomy with lymph node resection.

Discussion

NHL involving the breast either as a primary site or as a site of recurrence from lymphoma previously diagnosed elsewhere is rare. Several series have reported varying incidences of primary and secondary cases. Primary NHL of the breast is a rare disease, representing only 0.04%-0.50% of malignant breast neoplasms, [6] 1.7% of all extranodal NHL and 0.7% of all NHL [7].

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Histologically, primary breast lymphoma is predominately of B-cell origin and most commonly large cell type [8]. Numerous authors have found that lymphoma occurs more frequently in the right breast and that the rate of secondary lymphoma metastatic to the breast only slightly exceeds primary breast involvement [8,9]. Primary NHL presents most commonly as a palpable mass [10].

Our patient had all the clinical cutaneous features of Primary breast Lymphoma and PNHBL was finally diagnosed. NHL (nodal and extranodal), have variable histopathologic features dependent on the morphology of the tumor cells. In most cases of PNHBL, diffuse large B-cell lymphoma is the predominant histopathologic type [11-13] as in our patient. The following strict criteria must be met for a neoplasm to be characterized as PNHBL: (1) an adequate pathologic specimen, (2) close association of mammary tissue and lymphomatous infiltrate, (3) no evidence of disseminated lymphoma at the time of diagnosis, and (4) involvement of ipsilateral axillary nodes only if it occurs concomitantly with the primary lesion [14]. Our case met all these criteria for the diagnosis of PNHBL.

In addition to physical examination, X-ray of the chest, skull and pelvis is a reliable method for detecting visceral and nodal dissemination and should always be performed.

The treatment of PNHBL is similar to that used for other lymphomas and depends on the histological type. Most Clinicians agree that multimodality treatment is necessary [10,15,16]. However, recent studies have shown that aggressive B-cell lymphomas should always be treated with chemotherapy alone or in combination with radiotherapy [10,15,17,18]. The most effective combination reported in the literature is radiotherapy and 3 to 10 cycles of treatment with CHOP [10,13,15].
Only studies with relatively small cohorts of patients have been reported in the literature. For aggressive tumors, the literature recommends CHOP type chemotherapy and mastectomy with lymph node resection, if needed, for management of PNHBL. We were able to achieve an excellent response in our patient without surgical intervention. Survival rate of primary breast lymphoma is better as compare to both lobular cases and systemic lymphoma with secondary involvement of breast. Anticancer drugs are main treatment rather than surgery so it is very important to accurately diagnose primary lymphoma of breast.

References