PRIMARY HODGKIN’S LYMPHOMA OF THE BREAST INITIALLY TREATED BY SURGICAL EXCISION AND AXILLARY DISSECTION

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SUMMARY – Primary breast lymphoma accounts for 0.04%-0.5% of all breast malignancies. Primary non-Hodgkin's lymphomas of the breast are extremely rare and represent approximately 0.38%-0.7% of all cases. Epstein-Barr virus (EBV) has been postulated to play an important role in the pathogenesis of Hodgkin's lymphoma. We present a case of EBV-positive primary Hodgkin's lymphoma of the breast. Although there is no sufficient data on this challenging diagnosis, it is known that it could be misdiagnosed with inflammatory breast carcinoma. A crucial part of the proper histopathologic diagnosis is immunohistochemistry. Different modalities of treatment include operative procedure, chemotherapy and radiotherapy. Recent studies suggest chemotherapy and radiotherapy to be initial treatment for patients with primary breast lymphomas.

Key words: Primary breast lymphoma; Hodgkin's lymphoma; Epstein-Barr virus; Immunohistochemistry

Introduction

Breast lymphoma is a rare condition, both as primary and metastatic disease. The primary form has an incidence ranging from 0.04% to 0.5% of all breast neoplasms, whereas the metastatic form has an incidence of approximately 0.07%. Primary breast lymphoma accounts for 0.04%-0.5% of all breast malignancies1,2. Primary non-Hodgkin's lymphomas (NHL) of the breast are extremely rare and represent 0.38%-0.7% of all cases3. Epstein-Barr virus (EBV) has been postulated to play an important role in the pathogenesis of classical Hodgkin's lymphoma. We present a case of EBV-positive primary Hodgkin's lymphoma.

Case Report

A 66-year-old patient, para III, presented with 2 fast-growing solid, painless lumps in the upper right quadrant towards the axilla. Clinical examination revealed regularly shaped, mobile, painless lumps, without any significant changes on the skin. Ultrasonographic examination showed a homogeneous tumor within the axillary fossa that spread towards the right breast, measuring 8x8 cm in diameter. All laboratory tests and tumor markers were within the normal range. Magnetic resonance imaging (MRI) of the abdomen and thorax did not reveal any primary or metastatic disease. The patient underwent total excision of the breast tumor and axillary dissection. Operative procedure and postoperative course were uneventful. A semi-solid tumor of 114 g within fatty tissue was extirpated and measured 10x7x4 cm (Fig. 1). Histopathologic examination showed disruption of lymphatic architecture with fibrosis within 3 out of 8 lymph nodes,
surrounded by eosinophilic infiltration, interfollicular atypical blasts, lymphomatous infiltration and histiocytes. Large multinuclear tumor cells with prominent nuclei and Reed-Sternberg cells were identified.

Blastoid cells were CD30 positive showing immunohistochemistry coexpression with CD15, LCA (CD45) and were CD20 negative, with CD3 positive T-lymphocytes. Five of 8 lymph nodes were without metastatic tumor tissue. The histopathology finding was suggestive of classical EBV-positive Hodgkin's lymphoma of mixed type. The patient was referred for further hemato-oncologic treatment.

Discussion

According to the literature, there are no sufficient data on malignant lymphoma of the breast. They may present both as a primary or secondary tumor, and both are rare. There are no morphological criteria to differentiate between the two. Although primary breast lymphoma may appear at any age, the majority of patients are postmenopausal women.

Liu et al. have published data on 93 patients with primary breast lymphoma; according to Ann Arbor classification, 57% of patients were stage I, 23% were stage II, 4% stage III and 16% stage IV. With 3-year survival, overall survival rate of 70% and high relapse rate of 48%, it is considered to be a reason for concern. The patients having received chemotherapy and radiotherapy had better survival rate and lower relapse rate; therefore, the authors suggest that chemotherapy and radiotherapy be the initial treatment for patients with primary breast lymphoma. Zarnescu et al. have published a case report about unusual presentation of primary Hodgkin's lymphoma of the breast. The tumor clinically presented as inflammatory breast cancer and core biopsy showed granulomatous mastitis, requiring surgical biopsy. This case showed that despite the uncommon clinical presentation, step-by-step diagnosis led to immunohistochemistry and proper diagnosis. Some recent studies have shown that primary non-Hodgkin's lymphoma of the breast involving the axilla usually presents with the clinical appearance of inflammatory breast carcinoma without infiltration of the breast dermal lymphatics. Tumor growth is usually accelerated, which leads to enlarged tumors like the one described by Rahmat et al. EBV has been postulated to play an important role in the pathogenesis of Hodgkin's lymphoma. It is important to mention that immunodeficiency status such as HIV infection may predispose to EBV-associated Hodgkin's lymphoma.

The aim of this case report is to highlight an uncommon oncologic disorder such as primary Hodgkin's lymphoma of the breast and particularly to emphasize its clinical, radiological and histopathological presentations, and to discuss all the possible modalities of treatment (surgery, chemotherapy, radiotherapy).

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Sažetak

PRIMARNI HODGKINOV LIMFOM DOJKE INICIJALNO LIJEČEN KIRURŠKOM EKSCIZIJOM I AKSILARNOM DISEKCIJOM

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I incidencija primarnih limfoma dojke kreće se od 0,04% do 0,5%. Primarni non-Hodgkinov limfom dojke iznimno je rijedak s učestalostu od oko 0,38%-0,7%. Dokazano je kako u patogenezi Hodgkinova limfoma sudjeluje Epstein–Barrov virus (EBV). Prikazujemo slučaj bolesnice s EBV-pozitivnim primarnim Hodgkinovim limfomom dojke. Iako do danas nema dovoljno literaturnih podataka o ovoj iznimno intrigantnoj bolesti, poznato je da je u najvećem broju slučajeva naj-ozbiljniji diferencijalno dijagnostički problem upalni karcinom dojke. Temeljna odrednica histološke dijagnostike je imunohistokemija. Postoje različiti modaliteti liječenja koji uključuju operacijski zahvat te kemoterapiju i radioterapiju. Novije studije preporučuju kombinaciju kemoiradijacijskog liječenja kao inicijalnog tretmana primarnog limfoma dojke.

Ključne riječi: Primarni limfom dojke; Hodgkinov limfom; Epstein–Barrov virus; Imunohistokemija