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Primary Breast Burkitt: A Rare Entity

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Abstract **Case Report**

Primary breast lymphoma (PBL) is a rare localization of non-Hodgkin's lymphoma (NHL), accounting for less than 0.5% of breast cancers and only 2% of extraganglionic lymphomas. Its clinical and radiological manifestations, often non-specific (painless breast mass, skin changes), make diagnosis complex, and can lead to delayed management. We report the case of a 22-year-old patient, a particularly rare age for this pathology. Through a review of the literature, we explore the epidemiological, clinical, histological and therapeutic aspects of mammary Burkitt's lymphoma, emphasizing the importance of early diagnosis to improve management and prognosis. Thanks to rapid and intensive management, the patient was in complete remission after 20 months of follow-up.

Keywords: Primary Lymphoma, Breast, Burkitt.

INTRODUCTION

Breast Burkitt's lymphoma is a rare and particularly aggressive variant of non-Hodgkin's lymphoma that specifically affects B cells. Although the breast is an atypical site for this type of lymphoma, it can occur in women, particularly during periods of hormonal and immunologic change. This unusual localization makes it a complex clinical entity that requires prompt and appropriate management [1]. Burkitt's lymphoma is characterized by extremely rapid proliferation of tumor cells associated with a key genetic abnormality: a chromosomal translocation involving the MYC gene. This translocation leads to permanent, uncontrolled activation of MYC, a key regulator of cell growth, which explains the aggressiveness and rapid evolution of this pathology [2].

Diagnosis of Burkitt's lymphoma is based on histopathologic and immunohistochemical analyses, which generally reveal a germinal center B-cell phenotype and high expression of MYC, the key marker of this pathology [2]. Imaging techniques, such as 18F-FDG PET/CT, play an essential role in assessing the extent of disease and guiding therapeutic decisions by providing precise visualization of affected areas [3]. Together, these diagnostic tools allow for optimal, personalized treatment.

CASE REPORT

A 22-year-old woman, nulliparous and with no previous medical history, consulted a general practitioner for a right breast mass that had appeared only 4 weeks ago and had increased in volume dramatically over the past 1 week. The patient describes the lump as painless, but disturbing due to its large size and aesthetic impact. She also reports mild localized skin redness, with no associated nipple discharge or fever. No other systemic symptoms (night sweats, weight loss or marked fatigue) are mentioned.

On clinical examination, a right breast mass measuring 18 cm long was palpated. It was hard, fixed to the superficial and deep planes, and had limited mobility. The skin was slightly erythematous but without ulceration or major inflammatory signs. No axillary or cervical adenopathy was detected, and the general examination (abdomen, liver, spleen, peripheral lymph nodes) was unremarkable.

The diagnostic approach was progressive. Mammography revealed confluent tissue lesions of the right breast classified as ACR4 (suspicion of malignancy), measuring up to 65 mm x 60 mm. A breast ultrasound showed an hypoechogenic, heterogeneous mass with irregular contours and increased vascularity on Doppler, reinforcing the suspicion of malignancy.

An excisional biopsy of the mass was performed. Histological analysis revealed a diffuse proliferation of medium-sized lymphoid cells, with a characteristic starry sky appearance. Immunohistochemistry confirmed CD20+ (B cells), CD10+, BCL2+ and MUM1+ expression, with a Ki67 proliferation index of 97%, highly suggestive of Burkitt's lymphoma. FISH translocation testing identified the presence of t(8;14), implicating the MYC gene, confirming the diagnosis.

An extension assessment was performed, including 18F-FDG PET/CT, which showed several hypermetabolic right breast masses and pathological right axillary adenopathies, without systemic dissemination. A lumbar puncture ruled out meningeal involvement, and a myelogram revealed no bone marrow infiltration. Biological tests showed elevated LDH levels, reflecting the high tumour burden, but preserved renal and hepatic function.

Considering the large tumor size and the high grade of the tumor, urgent and intensive management was initiated. Pre-phase COP (cyclophosphamide, vincristine, prednisone) was administered for 7 days to reduce tumor burden and prevent tumor lysis syndrome.

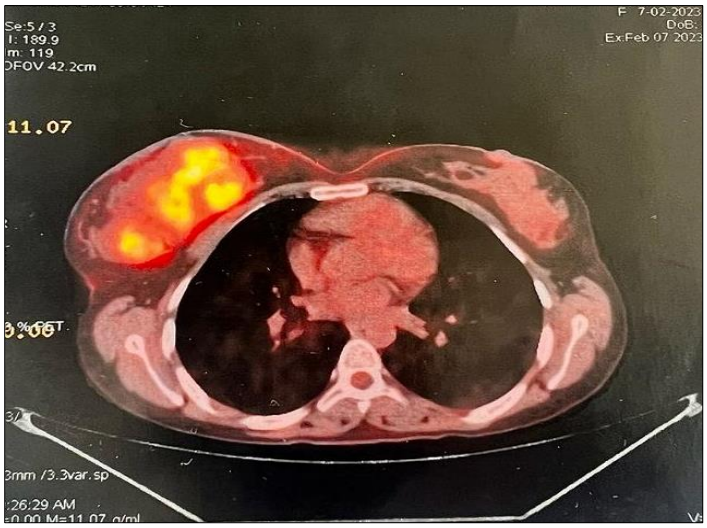

Figure 1: PET scan image showing multiple hypermetabolic masses in the right breast

The main treatment consisted of 6 courses of chemotherapy according to the R-DA-EPOCH protocol (rituximab, etoposide, prednisone, vincristine, cyclophosphamide, doxorubicin). Central nervous system prophylaxis was performed with 4 intrathecal doses of methotrexate.

After 4 courses, a re-evaluation PET/CT scan showed a complete metabolic response with a discrete residual lesion. Surgical biopsy of this lesion revealed no residual tumour infiltration, confirming the efficacy of the treatment. The last two courses of R-DA-EPOCH were administered to consolidate the therapeutic response.

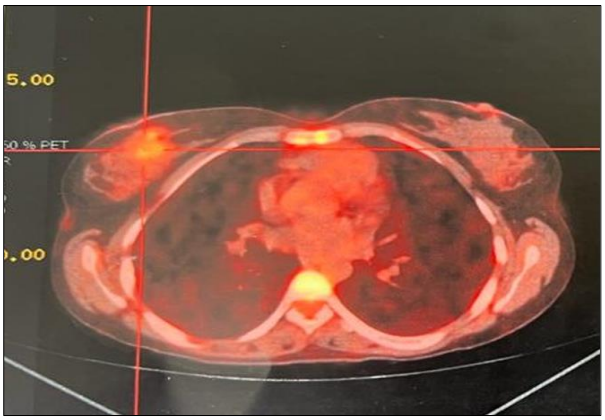

Figure 2: PET scan showing residual hypermetabolism after 4 cycles of R-DA-EPOCH

To this day, 20 months after the end of treatment, the patient is in complete remission, with no signs of relapse. Her general condition is excellent, with no late chemotherapy-related complications.

DISCUSSION

We report the case of a 22-year-old woman presenting with Burkitt's lymphoma of the breast, a rare and aggressive entity. This case illustrates the diagnostic and therapeutic challenges of this pathology, while highlighting the importance of prompt management. The point of this discussion is to contextualize this case within the existing literature and to highlight the clinical, diagnostic and therapeutic particularities of Burkitt's lymphoma of the breast.

In our patient, the presence of a painless but rapidly growing breast mass strongly suggests Burkitt's lymphoma, despite the rarity of this localization. This type of involvement is more common in younger women, particularly during pregnancy or breast-feeding, suggesting a possible involvement of hormonal fluctuations and immunological changes in its pathogenesis [4]. In our case, the absence of pregnancy or breast-feeding accentuates the unusual nature of this presentation.

The diagnosis of Burkitt's lymphoma is based on histology and immunohistochemistry. In our case, analysis revealed a diffuse proliferation of medium-sized lymphoid cells, with a characteristic “starry sky” appearance. Expression of CD20+, CD10+ and BCL2+ markers, together with a Ki67 proliferation index of 97%, corresponded to criteria established in the literature. Molecular confirmation by FISH revealed the t(8;14) translocation involving the MYC gene, thus validating the diagnosis. Present in over 90% of cases, this genetic alteration leads to persistent activation of MYC, a key regulator of cell proliferation, which explains the aggressiveness of the disease [5].

The management of Burkitt's lymphoma is based on early and intensive chemotherapy. The R-DA-EPOCH protocol, combining rituximab, etoposide, prednisone, vincristine, cyclophosphamide and doxorubicin, was chosen for its proven efficacy in the treatment of aggressive Burkitt's lymphoma [6]. Prophylactic intrathecal administration of methotrexate was necessary to prevent the high risk of meningeal involvement associated with this disease [7].

Our case matches the descriptions in the literature in terms of the clinical, biological and therapeutic features of Burkitt's lymphoma. However, breast localization remains rare, with only a few cases reported [8].

With intensive treatment, 5-year overall survival for localized Burkitt's lymphoma is around 70-80%, illustrating the importance of well-adapted therapeutic management [9].

This case highlights the rare and aggressive nature of mammary Burkitt lymphoma, an atypical and poorly documented localization. It underlines the importance of early diagnosis based on a multidisciplinary approach integrating imaging, histology and molecular biology, as well as intensive management including adapted chemotherapy protocols and central nervous system prophylaxis. The complete response observed in this young patient illustrates the efficacy of rapid, personalized treatment, while opening up prospects for future research into prognostic factors and innovative therapeutic strategies in this rare pathology.

CONCLUSION

In conclusion, this case illustrates the importance of early diagnosis and multidisciplinary management to improve the prognosis of breast Burkitt lymphoma, a rare and aggressive pathology. The complete remission achieved with the R-DA-EPOCH protocol confirms the efficacy of intensive treatment in this pathology. However, further research is needed to better understand the specificities of this unusual localization and optimize patient management.

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