Primary Breast Burkitt Lymphoma: Case Report

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Linfoma de Burkitt Primário de Mama: Relato de Caso

Linfoma de Burkitt Primario de Mama: Relato de Caso

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ABSTRACT

Introduction: Primary breast lymphoma (PML) represents about 0.5% of breast cancers, being considered a rare type of tumor. Some of the types of PML, in turn, have an intense connection with the pregnancy and postpartum period due to hormonal stimulation. The aim of this study is to report a case of PML with a rare presentation of Burkitt's lymphoma, considering effective therapeutic proposals for follow-up. **Case report:** A 23-year-old female patient with a peri-areolar tumor in the right breast with orange peel aspect and rapid growth for one month, phlogistic complaints at the lesion site, good general condition and no other associated symptoms. The reported condition started seven months after fetal expulsion with a dead fetus. The patient underwent lesion biopsy and was referred to an outpatient follow-up with unsuccessful outcome, requiring return to the hospital due to worsening of clinical conditions and extension of the tumor. There was a diagnosis of Burkitt's lymphoma, with multidisciplinary treatment. She underwent the CODOX-M chemotherapy protocol, and died after 22 days of hospital follow-up. **Conclusion:** This report demonstrates a rare situation in a young patient, emphasizing the importance of effective investigation of breast changes so that correct early diagnosis and appropriate treatment can be made for all age groups.

Key words: Burkitt lymphoma/drug therapy; Burkitt lymphoma/radiotherapy; breast neoplasms/drug therapy; breast neoplasms/radiotherapy; case reports.

RESUMO

Introdução: O linfoma primário de mama (LPM) representa cerca de 0,5% das neoplasias mamárias, sendo considerado um tipo raro de tumor. Alguns dos tipos de LPM, por sua vez, possuem ligação intensa com o período gravídico e pós-parto em virtude do estímulo hormonal. O objetivo deste estudo é relatar um caso de LPM com apresentação rara de linfoma de Burkitt, considerando propostas terapêuticas eficazes para o seguimento. **Relato do caso:** Paciente do sexo feminino, 23 anos, portadora de tumoração periareolar em mama direita com aspecto de casca de laranja e crescimento rápido há um mês, queixas flogísticas no local da lesão, bom estado geral e sem outros sintomas associados. O quadro relatado iniciou-se sete meses após a expulsão fetal com feto morto. A paciente foi submetida à biópsia da lesão e encaminhada para seguimento ambulatorial sem sucesso, necessitando de retorno ao ambiente hospitalar por piora das condições clínicas e extensão da tumoração. Houve diagnóstico de linfoma de Burkitt, com tratamento multidisciplinar, sendo submetida a protocolo CODOX-M de quimioterapia, com óbito após 22 dias de acompanhamento hospitalar. **Conclusão:** Este relato demonstra uma situação rara em uma paciente jovem, ressaltando a importância de investigar as alterações mamárias, de maneira eficaz, para um diagnóstico precoce correto e um tratamento adequado, em todas as faixas etárias.

Palavras-chave: linfoma de Burkitt/tratamento farmacológico; linfoma de Burkitt/radioterapia; neoplasias da mama/tratamento farmacológico; neoplasias da mama/radioterapia; relatos de casos.

RESUMEN

Introducción: El linfoma primario de mama (LMP) representa aproximadamente el 0,5% de los cánceres de mama, siendo considerado un tipo de tumor poco común. Algunos de los tipos de LPM, a su vez, tienen una conexión intensa con el embarazo y el posparto debido a la estimulación hormonal. El objetivo de este estudio es reportar un caso de LMP con rara presentación de linfoma de Burkitt, considerando propuestas terapéuticas efectivas para el seguimiento. **Relato del caso:** Paciente, 23 años, con un tumor periareolar en la mama derecha con aspecto de piel de naranja y rápido crecimiento durante un mes, quejas flogísticas en el sitio de la lesión, buen estado general y ningún otro. síntomas asociados. La condición reportada comenzó siete meses después de la expulsión fetal con un feto muerto. El paciente fue sometido a biopsia de la lesión y fue derivado sin éxito a seguimiento ambulatorio, requiriendo el retorno al entorno hospitalario por empeoramiento de la clínica y extensión del tumor. Hubo un diagnóstico de linfoma de Burkitt, con tratamiento multidisciplinario. Se sometió al protocolo de quimioterapia CODOX-M y murió a los 22 días de seguimiento hospitalario. **Conclusión:** Este informe demuestra una situación poco común en una paciente joven, enfatizando la importancia de investigar de manera efectiva los cambios en los senos para un diagnóstico temprano correcto y un tratamiento adecuado en todos los grupos de edad.

Palabras clave: linfoma de Burkitt/tratamiento farmacológico; linfoma de Burkitt/radioterapia; neoplasias de la mama/tratamiento farmacológico; neoplasias de la mama/radioterapia; informes de casos.

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INTRODUCTION

Breast cancer is a high morbimortality neoplasm in females, it is the most common in women in Brazil and worldwide – after non-melanoma skin cancer – and accounts for 25% of new cancer cases each year. It can also affect men, though rarely, with 1% of the total cases[1]. Among the histological types of this neoplasm, lymphomas comprehend just a few cases[1,2]. Lymphoma as primary presentation of the disease is yet more rare, barely reaching 0.5% of all breast cancers[2-5].

For the pathology in question, the involvement of ipsilateral axillary lymph nodes is not a pattern, may or not be present[5]. In most of the cases, the histological subtype of primary breast lymphomas (PBL) is non-Hodgkin[2,4,5]. Burkitt lymphoma is another subtype of highly aggressive tumor accounting for 10.3% of all the types of PBL[6-8].

PBL originates from periductal and perilobular lymphoid tissues of the breast and may arise from mucosal-associated lymphoid tissue[1,6,9], likely hormone-triggered specially during pregnancy or puerperium[2,3,6,8].

Because of its rarity there is no standard-of-care reported, however, targeted chemotherapy for the histological type and radiotherapy are methods with good outcome. Surgeries are controversial according to the literature, as opposed to diagnostic biopsies which are widely adopted[1,5,7].

The aim of this study is to describe a case of PBL with rare presentation of Burkitt lymphoma. In addition, report the clinical follow-up of the patient, with therapeutic management to expand the knowledge about breast cancer and its less common and aggressive forms, mainly during high hormone demand as in pregnancy-puerperium.

The Institutional Review Board of "*Hospital São José do Avaí*" in Itaperuna, Rio de Janeiro approved the study on April 15, 2021 (CAAE 47226121.8.0000.5288).

CASE REPORT

Black female patient, 23-years old admitted to the Oncology and Gynecology Clinic of "*Hospital São José do Avaí*" – reference of the Northeast Region of Rio de Janeiro – in the municipality of Itaperuna in December 2019 because of orange-peel like large volume tumor, mainly involving the periareolar region of the right breast with one month of evolution and progressive increase (Figure 1). The patient's history indicated fetal death seven months ago, submitted to biopsy of breast lesion with penrose drain in a complication-free procedure with subsequent medical discharge and surgical wound properly healing (Figure 2). She was on antibiotic therapy followed-up at outpatient with gynecological exams and waiting for the anatomopathological biopsy result.

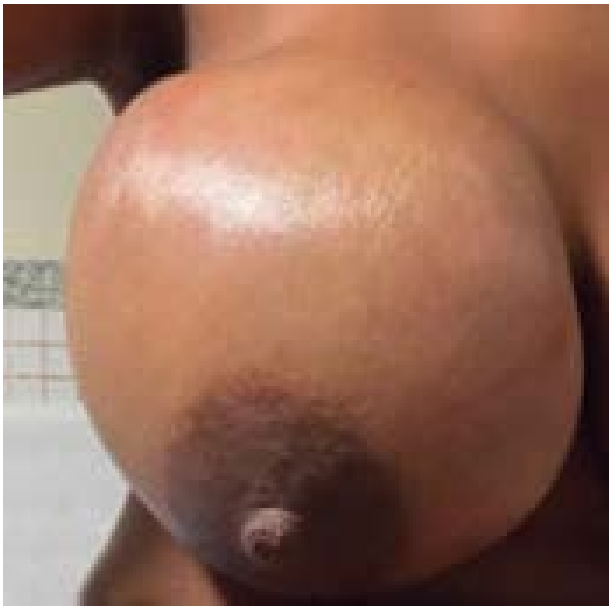

Figure 1. Right breast tumor

Figure 2. Post-biopsy breast of suspected lesion

In January 2020, returned to the hospital with diagnosis of undetermined breast neoplasm and advanced cervix neoplasm, with worsening of clinical condition and altered lab tests, leukocytosis of 19,200 with presence of rods and metamyelocytes, further to anemia and compromised renal function. In addition, the patient presented acute kidney lesion (AKL) after pelvic-tumor related urethral compression, which triggered hydronephrosis and pyelocalyceal dilation, being prescribed left nephrostomy by the general surgery clinic. She kept hemodynamically stable and presented altered blood count.

Seven days post-admission, diagnostic video laparoscopy was performed, revealing free-liquid in the cavity and specimen collection for analysis. The hematology clinic diagnosed Burkin's lymphoma and requested lab tests to determine staging, liquor evaluation and bone marrow biopsy.

After 15 days of the current admission, chemotherapy was initiated with protocol CODOX-M, with doxorubicin, vincristine, cyclophosphamide, methotrexate, leucovorin and granulocytes-colony stimulating factor (G-CSF). In addition, it was discussed intrathecal chemotherapy with cytarabine, methotrexate and dexamethasone. This protocol was initiated even in the absence of tests results earlier requested due to the worsening of the patient's clinical status.

After the chemotherapy began, the patient was sleepy and tachypneic, with altered blood count but improved leukocytosis and extremely high values of potassium (6.5) and low values of calcium (0.95). She evolved with critical metabolic acidosis and palpable mass in the right hemiabdomen. At the fourth day of the protocol, breast tumor improved with control of earlier complications, pancytopenia and tumor lysis syndrome. In the fifth day, severe case of neutropenia, thrombocytopenia and uremia, expressing uremic/septic encephalopathy, convulsive crisis, hypokalemia, urinary sepsis and gingival bleeding. For these reasons, chemotherapy was discontinued until dialysis, clinical improvement and revaluation of nephrostomy at hemodynamic clinic.

The patient evolved to hemodynamic instability needing orotracheal intubation due to hypoxemia and administration of intravenous amines, but despite the hemodynamic condition there was progressive improvement of the breast tumor, abdomen with right wall edema, chest petechias and poor extremities perfusion. After chest radiography, there was suspicion of adult respiratory distress (ARDS).

After 22 days of the current admission and three days after the discontinuation of the chemotherapy, her condition was severe with multiple organ disorders, pulmonary septic shock, hemodynamic instability with elevated doses of amines, coagulopathy, acute kidney failure (AKF), mixed acidosis refractory to mechanic ventilation and bicarbonate (HCO_3), oral and sclera bleeding and death after three cardiorespiratory arrests by hypoxemia and mixed acidosis.

DISCUSSION

Breast malignant lymphoma is a rare diagnosis, may manifest as a primary or secondary form as part of the metastatic process[3]. The present case is remarkable because the patient presented breast tumor after intense hormone stimulation due to pregnancy-puerperium period and unaltered clinical evaluation in general[8], which brings up the hypothesis of a primary pathology. The age is uncommon considering the typical presentation of PBL cases, mostly in older patients[1], however, it was diagnosed in a young woman compatible with Burkitt's lymphoma[4].

The diagnostic criteria for PBL are: 1) breast is the initial site where the neoplasm appears; 2) absence of early history of lymphoma; 3) absence of widespread neoplasm disease at the diagnosis; 4) histopathology analysis revealed association of the lymphoma with the breast tissue; 5) possibility of compromise of axillary lymph nodes simultaneous with breast lymph nodes[1,4,10,11].

Therefore, the suspected clinical diagnosis was breast neoplasm due to the similarities with most of the breast tumors[1]. The specific clinical particularities of Burkitt's lymphoma are painful mass usually at the upper lateral quadrant with local inflammation and fast evolution[6].

For diagnostic confirmation, the investigation consisted in breast biopsy and complementary gynecological evaluation to detect possible secondary involvement of other organs affected[2,8], which helps the histopathological diagnosis and determination of the prognosis of the patient. The colpocytology found advanced cervical neoplasm, likely secondary to the initial process, further to the systemic involvement revealed by the lab tests.

The initial complementary investigation included mammogram images, nuclear magnetic resonance, ultrasound and positron emission computed tomography (PET-TC)[6]. However, pathognomy findings were not detected and the alterations included vascular lumps and modifications and ganglia compromise[6,11]. Because of unspecificity, the findings must hold clinical-radiological correlation[11].

Abdominal ultrasound revealed that the cervix metastatic neoplasm provoked a urethral compression and triggered hydronephrosis and later, AKF. Even if not directly related, the systemic repercussion of PBL were elucidated through a holistic approach.

After the diagnosis of the pathology and beginning the discussions about the course of the therapeutic management, the literature presents diverse approaches to PBL treatment[11]. Chemotherapy combined with radiotherapy is the most known approach with positive results. Mastectomy is not beneficial to disease survival and/or recurrence[4]. Chemotherapy appears to be the only conduct consistent with the patient's axillary status.

In the case of aggressive forms as Burkitt's lymphoma, chemoprophylaxis of the central nervous system (CNS) is

mandatory even at initial stages with the best results and significant reduction of CNS recurrence[2].

To evaluate the prognosis of Burkitt's lymphomas, the histological type was associated with lymph node compromise and with risk factors of each patient[7].

For the patient investigated, the proposal was to initiate systemic chemotherapy associated with intrathecal chemotherapy to improve survivorship, inhibit tumor growth, reduce the risk of CNS involvement to evaluate the best response and continue with the protocol. And because of the aggressiveness of the histological type, after intense hormonal discharge, it was considered a dismal prognosis with more aggressive therapeutic.

The discussion of the case brought up the necessity of substantial scientific evidence in relation to the propedeutics of PBL expressed as Burkitt's lymphoma due to the aggressiveness and rarity of the neoplasm.

CONCLUSION

PBLs are fast-growing, progressive rare neoplasms and dismal prognosis. Positive diagnosis consists in targeted-biopsy and immunohistochemistry. There is no standard-of-care due to its rarity, however, the current literature recommends specific person-centered chemotherapy for the histological type possibly associated with radiotherapy of the affected breast, in addition to CNS prophylaxis of more aggressive forms of PBL even in initial stages because it can improve the result and reduce the risk of relapse.

Surgical management is controversial because it is argued that mastectomy may not be beneficial due to survivorship and PBL recurrence.

Burkitt's lymphoma is a tumor subtype of PBL with highly aggressive compromise and dismal prognosis. The follow-up of these patients demands more invasive management and according to the systemic repercussions caused by the neoplasm.

This case portrayed a rare condition in a young patient, emphasizing the importance of investigating breast changes effectively for correct early diagnosis and proper treatment in all age ranges.

The study was of great relevance for scholars and physicians of this institution because it contributed to expand the knowledge of breast cancer and its less common and aggressive forms, encourage the practice of breast self-exam and screening, most of all during high hormone demand as in the pregnancy-puerperal cycle and making women aware about early diagnosis and better outcome. It was beneficial too for teaching and research because of the material produced to help other investigators to expand the theme, suggesting acknowledged and effective therapies to improve the prognosis.

CONTRIBUTIONS

Laís Gomes Ferreira, Isabela Nagime Barros Gomes, Bruno de Almeida Castro Soares and Camila Fleckner Navarro Rodrigues Caldas contributed to the study design, acquisition, analysis and interpretation of the data, wording and critical review with intellectual contribution. Luciana Ximenes Bonani Alvim Brito and Rogério Martins de Castro contributed to the critical review with intellectual contribution. All the authors approved the final version to be published.

DECLARATION OF CONFLICT OF INTERESTS

There is no conflict of interests to declare.

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