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Primitive Myeloid Sarcoma of the Breast: A Case Report

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Abstract

Myeloid sarcoma is a disease that involves first the breast with rapid systemic dissemination. There is no scientific standard consensus on surgical and medical treatment of myeloid sarcoma of the breast and there is no role for radical surgery because chemotherapy and radiotherapy are required for longer survival. Surgery is needful in investigatory work-up to define immunohistochemistry diagnosis. We reported one case of 39-year-old Caucasian female with myeloid sarcoma without systemic involvement that was admitted to our Operative Unit for breast neoplasm. The patient is alive 19 months after surgery she is in good health, without relapse and she never developed neither clinical nor hematologic signs of myelo-proliferative disease. We can deduce that our therapeutic strategy was correct but we can't consider the disease completely cured because of its aggressiveness.

Keywords: Myeloid sarcoma; Breast; Quadrantectomy; Bone marrow transplantation

Introduction

Primitive Myeloid Sarcoma (MS) of the breast is a very rare type of leukemia (67 cases reported in literature) with primitive mammary growth [1,2].

Case Report

A 39-year-old Caucasian female was admitted to our Operative Unit on May 2014 with a not painful lump in the upper-outer quadrant of the left breast, noticed two weeks previously. Her family history was not significant for breast cancer and her past medical history was unremarkable. A physical examination revealed hard mass in the upper-outer quadrant of left breast. Mammography revealed an area of asymmetric increase of tissue density with hazy margins in the upper-outer quadrant of the left breast, without microcalcifications and neoplastic changes of the skin. A breast ultrasound showed lesion with altered echotexture and with heterogeneous adjacent areas, measuring 6 cm in the left breast; axillary lymph nodes appeared to be normal. Due to these findings, a vacuum assisted breast micro-biopsy with ultrasound guidance was performed. Microscopic examination revealed frustules of breast tissue with fibrosis and with infiltrating tumor composed of small-medium size blast cells. The tumor cells were positive for CD34, myeloperoxidase (MPO) and Leukocyte Common Antigen (LCA). Based on these immuno-positivity an oriented diagnosis of MS was performed. The complete blood count showed White Blood Cells (WBC) count of 8.18 x 103/µl (77.7% neutrophils, 17.2% lymphocytes, 4.3% monocytes, 0.6% eosinophils, 0.2% basophils), Haemoglobin Levels (HB) of 11.6 g/dl and platelets 297 x 103/µl. Other peripheral blood examination (kidney and liver function and coagulation tests) results in the correct range.

On 13th May 2014, after collegial and multidisciplinary meeting with onco-haematologists, the patient underwent left upper outer quadrantectomy with axillary lymph node dissection of some larger lymph nodes. Macroscopic histological examination of all tissue removed by surgery revealed whitish-grey neoplasm, measuring 6.5 × 4 × 2.5 cm, with polylobate margins; immunohistochemical analysis was positive for MPO, CD34, CD68/KP1 and negative for CD68/PGM1, c-kit, CK and CAM 5.2. Definitive histological diagnosis was MS of the breast (Figure 1). Subsequently, bone marrow aspirate and biopsy were performed and they didn't evidence localization of disease. Afterwards, HLA typing brother was performed and the results are the same. Patient also underwent Fluorodeoxyglucose-Positron Emission Tomography (FDG-PET) that highlighted a tenuous concentration of tracer in the left breast. On 11th June 2014 patient received inducted chemotherapy according to FLAI-5 regimen and then, although PDG-PET was negative for pathologic findings (Figure 2), based on high risk of disease progression, she received consolidation chemotherapy

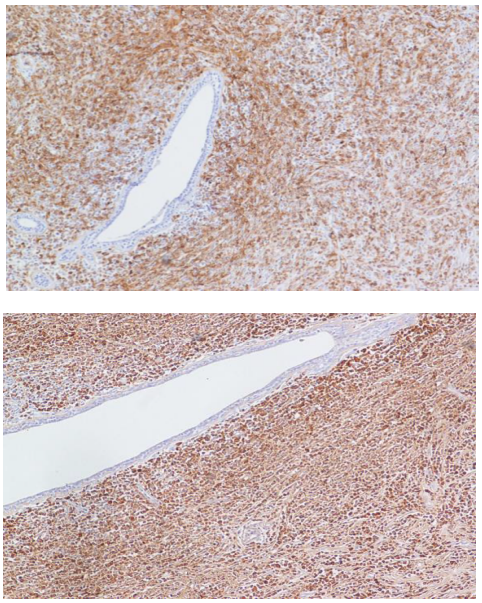

Figure 1: Histological analysis of breast tissue removed by surgery in our patient (case report analyzed): typical pattern of myeloid sarcoma with small-medium size blast cells mixed with eosinophil myelocytes.

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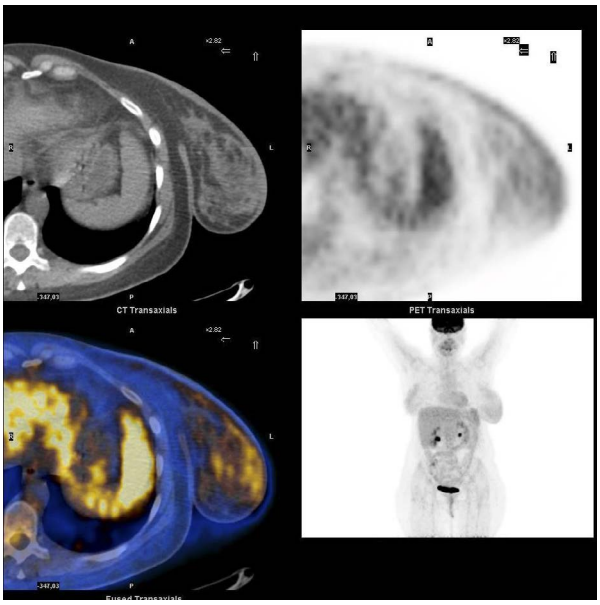

Figure 2: Post-operative FDG-PET: no evidence of disease recurrence.

according to IDA-AraC scheme. These treatments were well tolerated by the patient and after chemotherapy laboratory exams were in the normal ranges. Four months after surgery the patient received bone marrow transplantation from HLA-identical family member. Because of the absence of International Guidelines, the patient will continue follow-up according to collegial plan: onco-haematological and breast exams. The patient is alive 19 months after surgery, she is in good health, without relapse and she never developed neither clinical nor hematologic signs of myelo-proliferative disease.

Discussion

MS is a solid tumor composed of immature white blood cells called myeloblasts [2-4]. Granulocytic sarcoma can be classified into three clinically distinct occurrences: in AML, in myelodisplastic syndromes (MDS) progressing to leukemia, or when erytroleukemic blast crisis occurs from chronic myeloid leukemia, and as an extramedullary lesion before the occurrence of hematologic diseases such as acute leukemia [4-6]. MS of the breast can occur without a know pre-existing or concomitant diagnosis of acute leukemia or myeloproliferative (MPS) or myelodysplastic syndromes; this is know as primary chloroma [1,6,7,8]. 6% of primitive MS occurs in the breast without medullary involvement.

In our case report the patient presented only breast mass without medullary involvement. Chloroma is classified in three histotypes according to cells maturation: blastic type, immature type and differentiated type [1,2,6].

Our case report belongs to immature type. The age of presentation in isolated MS ranged from 20 to 72 years with a mean age of 42,4 years [1,3,9,10]: the patient analyzed entered again this range. Malignant cells of MS possesses some recurrent clinical characteristics and cytogenetic aberrations such as t(8;21), inv(16), M4 and M5 subtypes according to the French-American-British (FAB) classification of SML, MLL-AF9 rearrangement, trisomy 8, monosomy 7, leukocytosis, N-CAM expression and T-lineage markers (CD2, CD4, CD7) [5,8,10]. In our patient, mutational surveys, after bone marrow aspirate and biopsy, revealed no significant mutations involving FMS-like tyrosine kinase 3 (FLT3), no mutations of the nucleophosmin gene (NPM1) and of DNA cytosine-5 methyltrasferase 3A (DNMT3A). MS involving the breast usually manifests itself as unilateral or bilateral palpable mass, without pain or other associated symptoms such as nipple inversion or discharge [1,2,9,11]: this was the case treated.

Radiographic findings are not pathognomonic or specific in diagnosis of granulocytic sarcoma [12]; this varied imaging appearance makes it difficult to distinguish MS from breast cancer or malignant lymphoma [11].

This tumor on histopathology shows various infiltrative patterns like diffuse, Indian file, tergetoid and starry sky thus lobular and ductal structures are typically preserved and there aren't areas of destroyed tissue and tumor necrosis [1]. The case report analyzed showed typical histological pattern (Figure 1). 95% of patients with primitive MS will develop AML, MPS or MDS in 6-12 months [3]. Our patient didn't develop concurrence myeloid malignance and the definitive diagnosis of a chloroma was performed by immunohistochemistry, according to literature.

A panel of immunohistochemical markers comprising MPO, lysozyme, CD45, CD15, CD68/PGM1, CD13, CD34, CD43, CD117 and TdT is positive in the majority of cases and these are extremely useful to ascertain the myeloid differentiation [3,9,8,12]. In our patient the positivity for MPO and CD34 was confirmed. Differential diagnosis of MS of the breast includes high-grade non-Hodgkin lymphoma, lymphoblastic lymphoma, carcinoma of the breast, melanoma, extramedullary haematopoiesis, inflammation, neuroendocrine tumor and small round cell tumor in paediatric patients [1,4,6].

The current treatment regimen of MS is the conventional AML-type chemotherapeutic protocol; much is yet to be determined about the role of radiotherapy, along with chemotherapy in comparison to chemotherapy alone, on better outcome [6,7,10]. Surgery and radiotherapy alone aren't curative: as described in literature MS should always be considered manifestation of systemic disease. World literature agrees for bone marrow transplant (autologous or allogenic) as complementary treatment to chemotherapy in young patients with good conditions.

Our patient received bone marrow transplantation (four months after surgery) and she tolerated very well this treatment.

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