Primary Myeloid Sarcoma of the Breast: A Case Report and Review of Literature

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Abstract

Myeloid sarcoma (MS) of the breast is uncommon. We report this case of a 29-year-old female presented with a breast lump that was diagnosed as hamartoma by radiology. The lump was removed surgically. Pathological examination revealed a malignant tumor composed of immature cells with eosinophilic cytoplasm and single or multiple nucleoli, diffusely infiltrating the mammary parenchyma and sparing a few ducts and lobules. Immunostaining revealed positivity for leukocytic common antigen, myeloperoxidase, and CD68, and focally positive for CD34 and CD117. The final diagnosis was released as primary MS of the breast. The patient was treated with radiotherapy and chemotherapy. Three consecutive bone marrow biopsies were negative for neoplastic infiltration.

Keywords: Breast, myeloid sarcoma, primary

Case Report

Clinical findings

A 29-year-old female presented to King Abdulaziz University Hospital, Jeddah, Saudi Arabia, with left breast lump for 3 months before the clinical examination. Breast ultrasound of both breasts revealed a left supraclavicular well-defined mass which had a consistency of parenchymal ducts and fat. Mammography of left breast suggested the possibility of hamartoma. Magnetic resonance imaging findings suggested the possibility of breast hamartoma. The patient gave a consent prior to surgery.

Pathological findings

Frozen section examination was requested, and the specimen was received fresh. It consisted of fibrofatty tissue measuring 15 cm × 12 cm × 4 cm. Cut sectioning revealed a 13 cm × 10 cm × 4 cm, circumscribed, homogenous and well-defined mass. Immunostaining revealed positivity for leukocytic common antigen, myeloperoxidase, and CD68, and focally positive for CD34 and CD117. The final diagnosis was released as primary MS of the breast. The patient was treated with radiotherapy and chemotherapy. Three consecutive bone marrow biopsies were negative for neoplastic infiltration.
yellow-white mass with slimy cut surface [Figure 1a]. Frozen sections examined and revealed sheets and cords of atypical pleomorphic neoplastic cells. Surgeons were advised to wait for permanent sections examination. Several permanent sections were submitted for processing. Hematoxylin and eosin sections examined revealed a neoplastic growth formed of large immature cells exhibiting rounded nuclei with a thin rim of eosinophilic cytoplasm. Cells infiltrate mammary parenchyma and spare few ducts and lobular structures. Numerous atypical mononuclear granulocytes with eosinophilic cytoplasmic granules were seen. Periductal hyalinization was seen. Representative sections are shown in Figure 1b-d. Immunostaining was ordered.

**Immunohistochemistry**
A panel of immunohistochemistry was done including epithelial membrane antigen, cytokeratin-Pan, leukocytic common antigen (LCA), S-100, CD20, CD79, CD3, CD68, bcl-2, bcl-6, MPO, CD34, and CD117. Malignant cells were positive for LCA, MPO, CD68, and focally positive for CD34 and CD117. Representative sections are shown in Figures 2 and 3. The rest of antibodies were negative. The final diagnosis was released as extramedullary MS.

**Follow-up**
The patient was treated with radiotherapy and chemotherapy. Follow-up for 6 months’ postoperative was observed. Three consecutive bone marrow biopsies were examined and were negative for malignancy. Complete blood picture revealed a normal white blood cell count with no blast cells identified.

**Discussion**
In the current case, we report a tumor with large immature cells showing rounded nuclei and a thin rim of eosinophilic cytoplasm infiltrating the breast parenchyma. Tumor cell showed positive immunostaining for MPO, CD68, and LCA, indicating the myeloid nature of tumor cells. The following question was whether this tumor is a secondary involvement of the breast from myeloid leukemic or a primary lesion of the breast. Follow-up of bone marrow biopsy and peripheral blood examination failed to identify blast cells. This tumor was considered as a primary MS of the breast.

MS usually presents as a systemic manifestation of acute myelogenous leukemia; however, it could rarely come as an isolated mass without a history of leukemia. These patients frequently develop AML later on.[8,9] The most common body organs affected by MS are the skeletal system, soft tissues, and lymph nodes. Very few cases have been reported as an isolated mass with no significant history or subsequent development of acute myelogenous leukemia while on clinical follow-up.[10] Overall, MS has been classified into four categories as follows: (a) primary MS, (b) MS as a complication of AML, (c) MS as isolated recurrence of AML particularly during bone marrow remission and not followed by medullary relapse, and (d) MS with concurrent bone marrow relapse of AML.[11] Primary MS yields an unfavorable prognosis if not treated, and 88% of the cases tend to progress to AML within 1 year.[5,12] MS was shown to precede AML at which the bone marrow aspiration and biopsy reveal no hematological disease. This type of MS is called isolated, primary, or nonleukemic MS.[13]

Primary MS occurs in the absence of antecedent myeloproliferative disorder.[14] Primary MS is a rare entity with an incidence of two cases per million adults.[15] Sixty-seven cases of primary breast MS were reported until 2018.[16] In the breast, MS can be presented as a unilateral or bilateral mass, usually not accompanied by other symptoms such as a discharge from the nipple or nipple retraction.[10] Due to its rare incidence, MS of the breast is frequently misdiagnosed as breast lobular carcinoma, sarcoma, or lymphoma.[17-21] When the possibility of primary MS of the breast is not considered,
Figure 3: Immunostaining for CD79a, vimentin, CD34, and CD117. Tumor cells show diffuse membranous staining for CD79a (a), cytoplasmic staining for vimentin (b), membranous CD34 (c), and membranous CD117 (d) (×100).

many cases are often mistaken for non-Hodgkin’s lymphomas or other tumors. The big pitfall in the diagnosis of MS is due to its microscopic appearance that could vary from well-differentiated tumors showing all the differentiated stages of myeloid cells to tumors with no signs of differentiation. The latter could be easily misdiagnosed as other neoplasm.

Conclusions

MS affecting the breast is a rare disease, and its primary occurrence without accompanying myeloid leukemia is extremely uncommon. We report this case of a primary breast MS without any evidence of a myeloid neoplasm within a 6-month follow-up. Histopathological examination is the key element in the diagnosis of MS. MS should be highly considered in the differential diagnosis of breast tumors especially those with poorly differentiated morphology. MS should be in the pathological differential diagnosis of breast masses.

Declarations of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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