Myeloid sarcoma of the breast: a rare case
Published on 07.01.2021

DOI: 10.35100/eurorad/case.17106
ISSN: 1563-4086
Section: Breast imaging
Area of Interest: Breast Pelvis
Imaging Technique: MR
Case Type: Clinical Cases
Authors: Dr Sonali Sharma (MD), Dr. Jeevanjot Matharoo (MD), Dr Vidhi Bakshi (MD), Dr Jyoti Arora (CCST MD&DMDRE MBBS), Dr. Nupur Akash Patlel (DMRD), Dr Ruchika Goel (MD)
Patient: 27 years, female

Clinical History:
A 27-year-old lactating female on treatment for Acute Myeloid Leukemia (FLT-3 mutation-positive) presented with complaints of a firm, immobile lump in the outer half of the right breast and a fixed lump in the right axillary region.

Imaging Findings:
We could not acquire a mammogram because of the very poor ambulatory status of our patient.

Breast ultrasound showed a well-circumscribed large mass with heterogeneous low echogenicity and internal vascularity. Few other similar lesions were noted in the upper outer quadrant of the right breast. Enlarged discrete and conglomerated right axillary nodes with loss of fatty hila were seen.

On Contrast-Enhanced Magnetic Resonance Imaging (CEMRI) the right breast was enlarged, engorged, and had skin thickening. There were multiple well-circumscribed and irregular intensely enhancing lesions of variable sizes in the outer half of the right breast revealing restricted diffusion, low Apparent Diffusion Coefficient Values (ADC), and Type 3 curves. Multiple rounded, discrete, and conglomerate nodes were seen in the right axilla. Prominent right internal mammary nodes were also seen.

Histopathological and Immunohistochemical evaluation revealed features favouring the diagnosis of Myeloid Sarcoma of the breast with positivity for CD33, CD117, MPO (myeloperoxidase), negativity for CKCK (creatine kinase), GATA3, ER (estrogen receptor), CD68, and focal patchy positivity for CD 34.

Discussion:
Myeloid sarcoma (MS) is an extramedullary tumour which was first described in literature by Burns in 1811. King in 1853 was the first to call it chloroma (due to its green colour attributed to the enzyme myeloperoxidase (MPO). Dock in 1893 established its association with leukaemia.[1] Myeloid sarcoma is also known as granulocytic sarcoma, monocytic sarcoma, extramedullary myeloid cell tumour, myeloblastoma, and chloroma [2].

Myeloid sarcoma can occur in subcutaneous tissue, orbit, lymph nodes, small intestine, pelvic organs, and the brain [3,4], testis or the breast [5,6]. In 2–14% of acute myeloid leukaemia (AML) patients, myeloid sarcoma can be found, though it is most commonly seen associated with chronic myeloid leukaemia or myelodysplastic syndromes [6,7,8]. Breast involvement without a leukemic or myeloproliferative disorder is extremely rare [9,10,11]. However rarely myeloid sarcoma can present with isolated extramedullary mass without any previous or coexisting leukaemia [12,13]. The isolated cases may subsequently develop AML [12].
Overall, MS has been classified into four types: (i) primary MS, (ii) MS as an extramedullary manifestation of AML, (iii) MS as isolated recurrence of AML and (iv) MS with coexisting bone marrow relapse of AML.

Histopathological examination with immunohistochemical testing remains the gold standard of diagnosis, with MPO stain often strongly positive in myeloid sarcoma.

Clinically, non-specific symptoms are seen in MS which include mostly painless or painful breast lumps involving one or both breasts. Breast skin involvement and axillary lymph node enlargement may or may not be seen. Sonological findings of MS are variable. While some researchers like Thachil et al. have reported homogenous areas of low attenuation, with well-or-ill-defined margins others like Guermazi et al. have reported that breast masses in myeloid sarcoma were irregularly shaped, heterogeneous, hypoechoic masses with visible posterior acoustic shadows. Some researchers have documented cases with hamartoma like appearance as well.

In our case, the mass lesions were visualized as hypoechoic lesions with circumscribed margins creating an overlap with the imaging features of the benign masses like fibroadenomas, benign phyllodes and granulomatous mastitis.

On CE-MRI nonspecific findings such as variably enhancing masses and non-mass enhancements have been reported.

Histopathology along with immunohistochemistry for CD68/KP1, MPO, CD99, CD68, CD117, CD33, CD30, CD56, lysozyme is the markers for tumours myeloid differentiation. Our patient was positive for CD33, CD117, very strongly positive for MPO and showed patchy positivity for CD34.

Treatment regime includes AML type chemotherapy regime with suggested radiotherapy in cases with massive tumours with spinal cord compression. MS has the same prognosis as underlying leukaemia.

**Differential Diagnosis List:** Myeloid sarcoma of the breast, Fibroadenoma, Hamartoma, Granulomatous mastitis, Phyllodes tumour, Haematoma

**Final Diagnosis:** Myeloid sarcoma of the breast

**References:**


Description: Ultrasound of the sonography right breast revealing low echogenicity masses in the upper outer quadrant. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Ultrasound sonography with doppler of the right breast revealing low echogenicity mass with internal and peripheral vascularity. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Ultrasound sonography with doppler of the right breast revealing internal vascularity in a low echogenicity mass. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Ultrasound of the sonography right axilla revealing enlarged nodes with thickened cortex and effaced hilum. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Contrast-enhanced magnetic resonance imaging of the breast revealing variable-sized enhancing masses in the outer half. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Contrast-enhanced magnetic resonance imaging of the breast subtracted image revealing intensely enhancing masses in the outer half. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Contrast-enhanced magnetic resonance imaging of the breast (coronal plane) revealing irregular enhancing mass in the outer half of right breast. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: T2-weighted image right breast revealing isointense masses in the outer half. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Description: Adc map revealing low ADC values. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Figure 3

Description: Hrct chest revealing low-density mass in the outer half of the right breast. Origin: Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.
Figure 4

**Description:** Histopathology and immunohistochemistry revealing effaced breast parenchyma with neoplastic cells and positivity for cd 33, MPO, cd117 and negativity for cd 68, gata 3, CK and er and patchy positivity for cd34. **Origin:** Department of Radiology and Nuclear medicine, MEDANTA, The Medicity, Gurugram, Haryana, India.