Myeloid sarcoma presenting as pathological fracture in humerus: a case report

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Abstract: Myeloid sarcoma arise de novo or are associated with acute or chronic myeloid leukemia and other myeloproliferative disorders. Sometime myeloid sarcoma is first presenting sign of underlying undiagnosed case of leukemia. It is unrecognized for longer periods, appropriate chemotherapy may be delayed. Here we reported a case of myeloid sarcoma presenting as pathological fracture in humerus in 35 years female with chronic myeloid leukemia.

Keywords: Myeloid sarcoma, pathological fracture, chronic myeloid leukemia

INTRODUCTION
Myeloid sarcoma (also known as chloroma, granulocytic sarcoma) is a tumor mass due to aggregation of myeloid blasts at extramedullary site. It arises de novo or is associated with acute or chronic myeloid leukemia and other myeloproliferative disorders [1, 2]. Myeloid sarcoma is due to migration of blast cells to extra-osseous locations via haversian canals [3]. Most common site is facial bone [4]. Owing to its rarity in extremities, it becomes diagnostic dilemma in pathological fracture at these sites and if it is unrecognized for longer periods, appropriate chemotherapy may be delayed.

CASE REPORT
A 35 yrs old female presented to emergency department of Pt BD Sharma PGIMS Rohtak, with chief complaints of swelling and pain over left arm after fall on shoulder and gradually progressed to present size. At presentation, the patient was a febrile. Physical examination revealed a diffuse swelling of about 11x10 cms over the anterolateral aspect of left proximal arm, severe tenderness and loss of motion. The skin over the affected area was tense, shiny, and erythematous and with dilated vein.

X-ray left shoulder was done which revealed pathological fracture of proximal humerus. Patient was a known case of chronic myeloid leukemia diagnosed 5 years back. She was on imatinib therapy since then. Peripheral blood smears examination revealed normal hematological study. Soft tissue biopsy was taken from the site of lesion and was sent to the department of pathology for histopathological examination.

Multiple grey white soft tissue pieces measuring together 2x1x0.8cm was received. Histopathological examination after decalcification in EDTA revealed monomorphic population of round cells with large number of apoptotic bodies. (Fig.1) these cells had vesicular nuclei, prominent nucleoli and scanty amount of cytoplasm. (Fig. 2) Immunohistochemistry was applied and these cells were found to be negative for CK, CD99, CD5, CD20, CD23, MPO, and TdT and positive for LCA and CD34. (Fig. 3) On the basis of histopathological findings and immunohistochemistry, diagnosis of myeloid sarcoma (with undifferentiated blast cells) was made.
Fig-1: H&E section showed monomorphic population of round cells.

Fig-2: H&E sections showed cells having vesicular nuclei, prominent nucleoli and scanty amount of cytoplasm.

Fig-3: CD34 was positive in malignant cells. (IHC, 200 xs)
DISCUSSION

Myeloid sarcoma is an uncommon, malignant solid extramedullary tumor of immature or mature myeloid blast cells occurring at an anatomical site other than bone marrow [1]. It occurs primarily in patients with leukemia (acute or chronic myeloid leukemia) or other myeloproliferative disorders. Sometime myeloid sarcoma is first presenting sign of underlying undiagnosed case of leukemia in peripheral blood or bone marrow. This tumor is also known as chloroma, derived from Greek word chloros (meaning green), due to characteristic green hue on gross examination of tumor. This color is secondary to myeloperoxidase enzyme reaction found principally in myeloid cells [1, 3].

The association of myeloid sarcoma in patients with CML is only 2-4%. Bone (57%) is most common sites of involvement in patients with CML followed by lymph nodes (29%) and skin or soft tissue (21%). Bone and periosteum involvement occurs by migration of leukemic cells from bone marrow through haversian canals. The tumor is locally destructive and lead to destruction of cortex and medulla with extension into adjacent soft tissue mass. Due to its rarity and non-specific presentation frequently pose a diagnostic dilemma for clinicians resulting in delayed treatment. Myeloid sarcoma is warning signs of acute mylogenous leukemia or onset of accelerated disease and blast crisis of CML [1, 5].

Diagnosis usually requires correlation of radiological imaging such as CT, MRI and tissue biopsy. Immunohistochemistry is found to be the most useful characterization of these extramedullary solid tumors [6].

REFERENCES