Research Paper

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Chronic Myelocytic Leukemia With Extramedullary Infiltrates in Skin and Lymph Nodes - a Rare Manifestation

Medical Science

KEYWORDS : CML, extramedullary infiltrates, blastic transformation

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ABSTRACT

Chronic myelocytic leukemia (CML) is a myeloproliferative neoplasm arising at the level of a pluripotent stem cell and consistently associated with BCR-ABL fusion gene and most commonly manifests in a chronic phase of the disease. CML with extramedullary disease is a relatively rare condition. The sites involved are the lymph node, CNS, sub-orbital mass and as cutaneous lesions. Infiltration of neoplastic leukocytes or their precursors into epidermis, the dermis or the sub cutis is called leukemia cutis. It is an uncommon manifestation of CML and when present, points towards blastic transformation of the disease. We describe a case of CML in blast crisis with cutaneous nodules and inguinal lymphadenopathy.

Introduction:
Chronic myelocytic leukemia(CML) is a myeloproliferative neoplasm arising at the level of a pluripotent stem cell and consistently associated with BCR-ABL fusion gene.

CML most commonly manifests in a chronic phase of the disease.

Infiltration of neoplastic leukocytes or their precursors into epidermis, the dermis or the subcutis is called leukemia cutis.

It is an uncommon manifestation of CML and when present points towards blastic transformation of the disease.

We describe a case of CML in blast crisis with cutaneous nodules and inguinal lymphadenopathy.

Case Report:
A 35 years old female presented with mass per abdomen and generalized weakness of 7months duration. Clinical examination showed massive spleenomegaly reaching beyond the umbilicus up to right iliac fossa and hepatomegaly. (Fig1: massive spleenomegaly and hepatomegaly).

There were multiple subcutaneous nodular swellings of 1-1.5cm diameter, over the left buttock region and anterior surface of the left leg.(Fig 1a: Subcutaneous nodules over the buttock region).

Complete hemogram, bone marrow aspiration and fine needle aspiration from the subcutaneous nodules and inguinal lymph nodes were done.

The hemogram revealed Hb-6.8gm/dl, platelet count - $92x10^9$/L, total leukocyte count of $160x10^9$/L and the differential count as neutrophils-23%, eosinophils-4%, basophils-8%, lymphocytes-2%, metamyelocytes-10%, myelocytes15% ,myeloblasts and promyelocytes together 35%(Fig 2).

Bone marrow examination showed a hypercellular marrow with 30% of the nucleated cells being blasts.

FNAC of the subcutaneous nodules over the buttock and left inguinal lymph node showed the presence of leukemic cells(Fig 3).

Based on the hematological and cytological features the diagnosis was given as chronic myelocytic leukemia in blastic transformation with extramedullary infiltrates in skin and lymph node.

Discussion:
CML most commonly manifests in a chronic phase of the disease.

The present case was diagnosed to be in the blastic transformation with extramedullary infiltrates in the skin(leukemia cutis) and lymph nodes.

Leukemia cutis is a relatively rare condition and manifests in a variety of leukemia subtypes, with highest incidence in acute monocytic , myelomonocytic and T-cell leukemia[(1)].

It is an uncommon manifestation of CML(2-8%) and when present points towards blastic transformation[(2)].

Although skin infiltrates have been reported in chronic phase of the disease, this either precedes or occur simultaneously with a blood picture of more aggressive phase, hence the incidence largely correspond to either the accelerated phase or the blast phase of the diseases [(3)].

As many as 90% of patients with skin infiltrates also have other extramedullary involvement and as many as 40% may have meningeal or central nervous system involvement[(4)] .

In some cases, histological examination of the skin and soft tissue lesions revealed extramedullary hematopoietic nodules with associated hemorrhage and necrosis only, termed as pseudochloromas. These should not be confused with leukemia cutis, occurring with blastic transformation [(6)].

The vast majority of patients have an insidious onset with common initial findings like fatigue, night sweats, splenomegaly and anemia.

Unusual manifestations include

1] Early – stage at which time there may be only a modest sustained leukocytosis that, however, is almost always accompanied by basophilia.
2] Leukocytosis composed mainly of segmented neutrophils mimicking chronic neutrophilic leukemia(CNL).
3] Marked thrombocytosis accompanied by minimal leukocytosis , resembling essential thrombocytosis.
4] Leukocytosis with monocytosis that prompts consideration of chronic myelomonocytic leukemia(CMML).
5] Occurrence in the blastic phase without a history of preceding chronic phase.
6] In addition CML, can manifest with unexpected clinical findings such as lytic bone lesions or exttramedullary infiltrates in the skin or lymph node, which is usually associated with

blastic transformation.

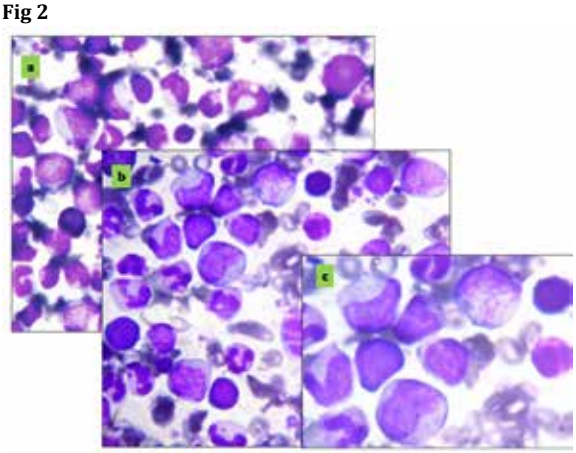
The present case was diagnosed in blastic transformation with extramedullary infiltrates in skin and lymph node.

However, the patient had a rapid downhill course, despite the therapy and supportive treatment was instituted.

Hence, in conclusion it is important to do a FNAC of the skin nodules and lymph nodes, whenever present, and look for the presence of leukemic cells, as is points towards blastic transformation of the disease or towards accelerated phase with impending blast crisis and therefore poor prognostic indicator.

Fig 1

Fig 2



Legends for images:
Fig 1: Hepatosplenomegaly and subcutaneous nodule over buttock region.

Fig 2a : x400, Leishman's stained blood smear showing leukocytosis with myeloblasts, promyelocytes, myelocytes, metamyelocytes and basophil, 2b: x1000, Leishman's stained blood smear, showing Myeloblasts, myelocyte and metamyelocyte, 2c: FNA of subcutaneous nodule(x400, Giemsa stain) with leukemic infiltrates.

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