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CASE REPORT

Extramedullary granulocytic sarcoma as an initial presenting feature of chronic myeloid leukemia

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ABSTRACT

Granulocytic sarcomas are rare extramedullary tumors consisting of granulocytic precursor cells. Rarely it may precede peripheral blood or bone marrow involvement, presenting as a diagnostic challenge. We report here a case of myeloid sarcoma in a 36-year-old man, who presented with cervical lymphadenopathy of one month duration. Fine needle aspiration cytology (FNAC) was performed and a diagnosis of myeloid sarcoma was offered. Subsequently, peripheral blood smear examination and complete blood counts done, which revealed features of chronic myeloid leukemia (CML) with accelerated phase. We report here a rare case to emphasize the diagnostic utility of FNAC in making the correct diagnosis of myeloid sarcoma.

Key words: FNAC, Wilms tumor, small round cell tumor (SRCT)

INTRODUCTION

Myeloid (granulocytic) sarcoma is a tumor mass consisting of myeloid blasts with or without maturation, occurring at an anatomical site other than the bone marrow.[1] It arises de novo, or are associated with other hematologic disorders such as acute myeloid leukemia, myelodysplastic syndrome (MDS), or myeloproliferative disorders (MPD).[2] If myeloid sarcoma occurs in a setting of MDS or MPD; it is equivalent to blast transformation.[3]

CASE REPORT

A 36-year-old male patient presented to Surgery outpatient department with easy fatigability, weakness and nodular mass in neck region (Fig. 1). Clinical diagnosis was kept as cervical lymphadenopathy. Patient was referred to department of Pathology for fine needle aspiration cytology. FNA was done on OPD basis which yielded blood mixed aspirate. Wet fixation of smears was done in ethyl alcohol. Air dried smears were also performed. Wet fixed smears were stained with Haematoxylin and Eosin (H & E) and Papanicolaou stain.

Smears revealed immature myeloid cells, few neutrophils and many large round cells with high nuclear cytoplasmic ratio and prominent nucleoli. Background showed some lymphocytes and lymphoglandular bodies (Fig. 2). In view of presence of myeloid cells, myeloperoxidase stain was done on air dried smears. They revealed MPO positive granules in blast cells and other myeloid cells (Fig. 3). The diagnosis of myeloid sarcoma was offered. Afterwards when

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Figure 1, Clinical photograph showing nodular mass on left side of neck.

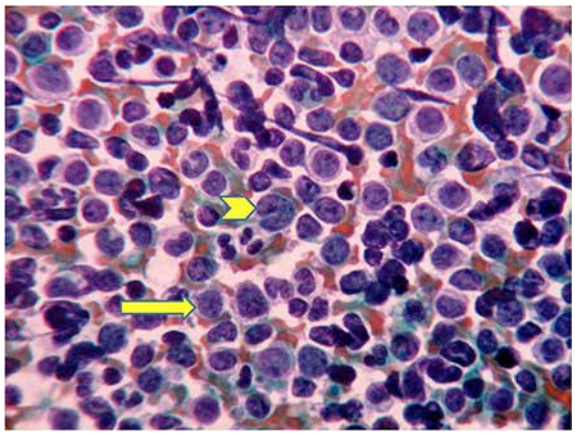

Figure 2, Photomicrograph of lymph node aspirate smear showing many blast cells (arrow), few myelocytes, metamyelocytes (arrow head), band forms and neutrophils (Papanicolaou stain, X 400).

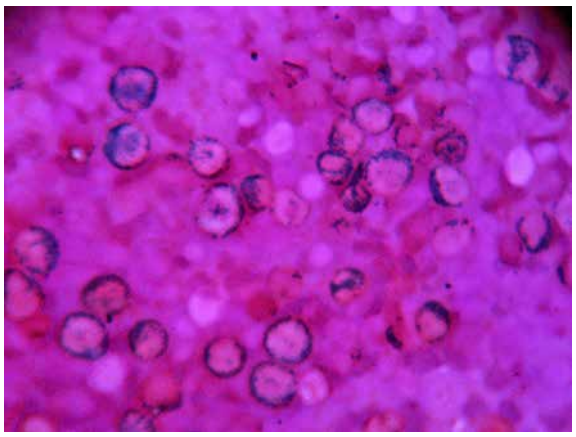

Figure 3, Photomicrograph of lymph node aspirate smear showing Myeloperoxidase positive granules in the cytoplasm of blast cells (Myeloperoxidase stain, X 1000).

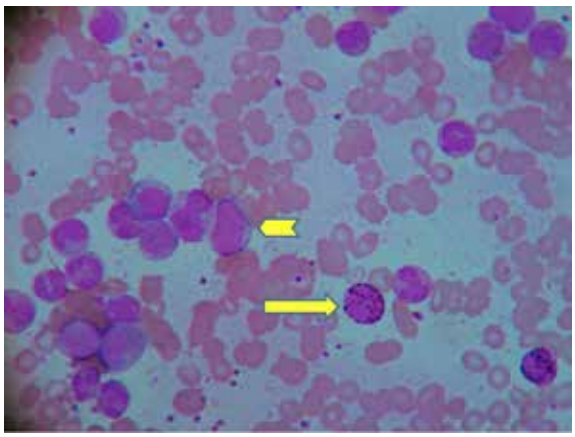

Figure 4, Photomicrograph of peripheral blood smear showing immature myeloid cells (arrow head), basophils (arrow), and plenty of platelets in the background (Leishman stain, X400).

patient came for report of FNAC after 3-4 days, we examined the patient. Clinical examination showed massive splenomegaly, but liver was not palpable. Chest X-ray was normal. There was no mediastinal lymphadenopathy. We performed peripheral blood smear examination and complete blood count. Haemoglobin was 9gm/dl. Erythrocyte sedimentation rate (ESR) was raised (40mm at the end of 1hour). RBC count 2.73 million per microliter and platelet count was 5.2 lakh per microliter. Total leucocyte count was one lakh per microliter and Peripheral blood smear showed many immature myeloid cells including basophils and plenty of platelets in the background (Fig. 4). Differential count showed Myeloblasts 11 %, Promyelocytes 2 %, Myelocytes 21 %, Metamyelocytes 18 %, Band forms 13 %, Neutrophils 22 %, Basophils 6 %, Eosinophils 2 % and Lymphocytes 5 %. Peripheral blood smear findings are of CML in accelerated phase. Considering FNAC and hematological findings final diagnosis was given as myeloid sarcoma of lymph node with CML in accelerated phase (CML-AP).

DISCUSSION

Granulocytic sarcoma is also known as chloroma, myeloid sarcoma or extramedullary myeloid cell tumor. Myeloid sarcoma refers to the presence of tumorous collections of immature cells at sites other than the bone marrow. Very rarely it may occur without a known pre-existing or concomitant diagnosis of acute leukemia, acute promyelocytic leukemia or MDS/MPS. This is known as primary chloroma. In almost all reported cases of primary chloroma, acute leukemia developed within a short time.[4] Kuan JW et al reported a case of primary GS where the tumor mass bcr-abl translocation was demonstrated by fluorescent in-situ hybridization in which no evidence of CML was present. This is an important finding as it highlights the possibility that CML may present as a sole extramedullary form.[5] The most common sites of occurrence are the subperiosteal bone structures of the skull, paranasal sinuses, sternum, ribs, vertebrae and pelvis; lymph nodes and skin are also common sites.[3] Ko SW reported a case of granulocytic sarcoma arising from uterine, cervical and parametrial region mimicking a haemorrhagic abscess in a 50 year old woman of CML with blast crisis.[6] Jenking and Sorour reported a case of large granulocytic sarcoma of the right groin. Bone marrow examination showed mild myeloid and eosinophilic hyperplasia but no evidence of increase in blast count.

However cytogenetic examination of the marrow showed t (9:22), indicating an unexpected diagnosis of CML.[2] A rare case of CML manifesting as multiple, non-pigmented, non-pruritic skin nodules in the chronic phase has been reported by Nagarajarao HS et al.[7] Rekha et al presented their experience with a patient manifesting with a non-healing ulcer of lower limb that was a chloroma associated with CML.[8]

Cytological preparations give better morphological details of blasts; the identification of Auer rods and MPO stain can help to provide the diagnosis of an undetected AML. The visualization of an Auer rod is possible only in cytological material, so it would be much easier to make diagnosis based on FNAC and imprint smears rather than in histological sections the work up of these masses can be aided by the use of cytochemistry (MPO, Sudan black, Nonspecific esterase) and immunophenotyping.[9]

Easy and rapid diagnosis of myeloid sarcoma is possible by FNA in fairly accessible lesions a demonstrated in our case.[7] The unusual presentation of CML in chronic phase or accelerated phase as seen in our case, should alert the clinician to consider FNA as a tool in the work-up of patients presenting with nodular lesions. FNA is cost effective. It provides rapid diagnosis and hastens further management of patients. In patients with MPS and MDS, myeloid sarcoma defines a blastic transformation often associated with a short survival.[9] Because of lack of diagnostic features particularly in patients without a clear pre-existing diagnosis of leukemia, blastic myeloid sarcoma may

be misdiagnosed as lymphoma or poorly differentiated tumor. In one published series on chloroma, the authors stated that 47% of the patients were initially misdiagnosed, as a malignant lymphoma.[10]

The present case was cytologically evaluated and interpreted as myeloid sarcoma. The correct diagnosis of myeloid sarcoma and differentiation from other mimics are crucial for proper patient management.

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