

Myeloid sarcoma de novo presenting as generalized lymphadenopathy

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ABSTRACT

Myeloid sarcoma manifesting as multiple lymphadenopathy is a rare entity. Bilateralism is even rarer. It can mimic lymphoma cytologically and histologically. We present a case of myeloid sarcoma mimicking as lymphoma in a 32-year-old male. The patient presented with bilateral neck masses and generalized lymphadenopathy for the past one month. A clinical diagnosis of lymphoma was made but fine needle aspiration cytology, histopathology of excised lymph node, immunophenotyping and cytochemistry on the FNA smears confirmed it to be a myeloid sarcoma and not lymphoma.

Key words: Cytochemistry, lymphadenopathy, myeloid sarcoma

INTRODUCTION

Myeloid sarcoma is also known as chloroma or granulocytic sarcoma. It is a rare localized tumor that is composed of myeloid progenitor cells. Burns first described the disease in 1811 and King's in 1853 introduced the term "chloroma" because of its green color due to myeloperoxidase enzyme.^[1] In 1966, Rappaport renamed the disease as granulocytic sarcoma.^[2] In recent years the term "myeloid sarcoma" has been preferred. The incidence of myeloid sarcoma is 2.5 to 9.1 % of the patients with acute myeloid leukemia and it is five times less frequent in patients with chronic myeloid leukemia. There is predilection for males with male and female ratio of 1.2:1. The median age of diagnosis is 56 years (Range-1 month to 89 years).^[3] Skin, lymph node, gastrointestinal tract, brain, bone, soft tissues and testis being more frequently affected. In less than 10% of cases, myeloid sarcoma presents at multiple anatomical sites.^[3] Myeloid sarcoma may develop de novo or concurrently with acute myeloid leukemia, myeloproliferative neoplasm or myelodysplastic syndrome. It may be the first manifestation

of AML, precede it by months or years, or represent the initial manifestation of relapse in a previously treated AML in remission.^[3]

Morphologically, there is partial or total effacement of tissue architecture by myeloblasts and in a significant proportion of cases; it displays a myelomonocytic or pure monoblastic morphology.^[3] A careful search can reveal erythroblasts. On cytochemical staining, granulocytic lineage (MPO +ve) can be differentiated from monoblastic forms (NSE +ve).

The major differential diagnosis is with malignant lymphoma. The diagnosis of myeloid sarcoma is validated by the results of cytochemical and/or immunophenotype analysis. These allow the distinction of myeloid sarcoma from lymphoblastic lymphoma, Burkitt lymphoma, diffuse large B cell lymphoma, small round cell tumor and blastic plasmacytoid dendritic cell neoplasm.^[3]

On immunohistochemistry, in paraffin sections, CD 68/KP1 is the most commonly expressed marker followed in decreasing frequency by MPO, CD117, CD99, Lysozyme, CD34, TdT, CD56, CD61/LAT/FVIII Ag, CD30, Glycophorin A and CD4.^[3] The combination of the above mentioned markers endorsed the recognition of tumors with immature myeloid phenotype, as well as cases with myelomonocytic, monoblastic, erythroid or megakaryocytic differentiation. Exceptionally, aberrant antigenic expressions were observed (cytokeratins, B or T cell markers).

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CASE REPORT

A 32-year-old male presented in our hospital with the complaints of fever, persistent vomiting, loss of appetite, difficulty in swallowing, abdominal pain since last one month. General condition of the patient was poor at the time of admission. On examination patient had bilateral palpable neck nodes, splenomegaly and mild ascites. A detailed CT scan revealed multiple enlarged lymph nodes in cervical and abdominal region and also bilateral internal iliac nodes. The patient had mild splenomegaly with mild ascites. On physical examination, the largest cervical node measured 3 × 2 cm in size. FNA biopsy of the node was done with a 23 gauge needle. Two smears were fixed in 95% ethanol and stained using Papanicolaou method and three smears were air dried and stained with modified Giemsa. Four unstained smears were reserved for ancillary testing. The smears showed a dispersed population of monocytoid like cells with abundant gray cytoplasm with vacuolation, few erythroid precursors along with neutrophils. Lymphoglandular bodies were not identified [Figure 1]. Diagnosis of granulocytic sarcoma was suggested because of variable population of erythroblasts like small cells. The concurrent lymph node biopsy showed complete effacement of the lymph node architecture by blast like cells with round to oval nuclei, prominent nucleoli, eosinophilic cytoplasm, few immature cells of the myeloid series, and few erythroid precursors with focal areas of necrosis [Figure 2]. The immature myeloid cell, erythroid precursors raised the suspicion of myeloid sarcoma but the blastic component resembled Non-Hodgkin lymphoma. Further ancillary testing like immunohistochemistry on the cell blocks and cytochemistry on the unstained FNA smears were done for confirmation. Bone marrow aspiration revealed no abnormality [Figure 3].

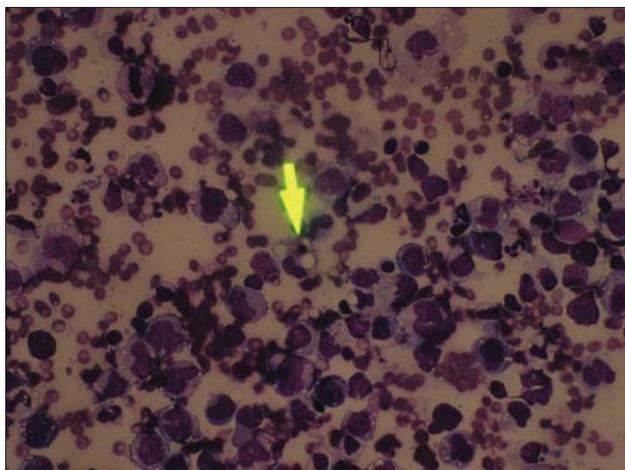


Figure 1: Smears show dis cohesive monocytoid cells with erythroblasts (arrow) (Giemsa stain ×400)

The immunohistochemical results showed the blasts to be CD 30 +ve [Figure 4], CD 43 +ve, CD 45 +ve [Figure 5] and EMA positive [Figure 6]. The blasts were TdT –ve, MPO –ve and PAN Cytokeratin –ve. The immunomarkers were non-conclusive but cytochemical stains showed strong NSE positivity [Figure 7] and MPO negativity [Figure 8] thus, giving way to the final diagnosis of monocytoid type of granulocytic sarcoma. After being diagnosed as myeloid sarcoma, the patient was referred to the Onco-hematology department for chemotherapy. However, because of personal reasons, the patient was transferred to another hospital before starting chemotherapy.

DISCUSSION

Myeloid sarcoma is a rare entity. It is more of a localized tumor than a systemic disease. There are few reports in the English medical literature of myeloid sarcoma manifesting as systemic, diffuse lymphadenopathy and bilateralism is even rarer.^[4-6] In this case also, generalized lymphadenopathy with bilateralism was observed. Very few cases of granulocytic sarcoma without evidence of leukemia have been reported. Cytochemistry plays a major role in establishing the diagnosis. In a study done by Meis JM *et al.*, 16 patients were diagnosed as granulocytic sarcoma without evidence of acute leukemia. Twelve of these 16 cases (75%) were initially labeled as large cell lymphoma. The naphthol ASD chloroacetate esterase stain was required to make the correct diagnosis in all cases.^[7] This case had an unremarkable marrow and a positive NSE and a negative MPO done on FNA smears established the diagnosis of monocytic myeloid sarcoma. The blasts were CD30 +ve, CD43 +ve, CD45 +ve, EMA +ve; few plasma cells were CD138 +ve while TdT, MPO and PAN Cytokeratin were negative in absence of NSE. The case was labeled as large cell lymphoma on immunohistochemistry. In a study done by Roth *et al.*,

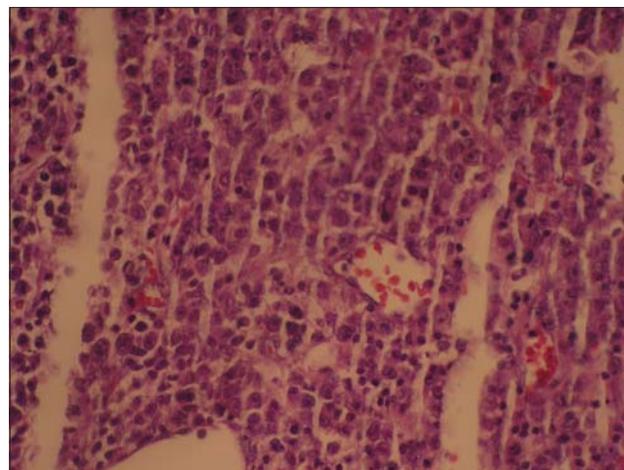


Figure 2: Section from lymph node show sheets and cords of blast cells in sinusoidal pattern, admixed small cells like erythroblasts (H and E stain ×400)

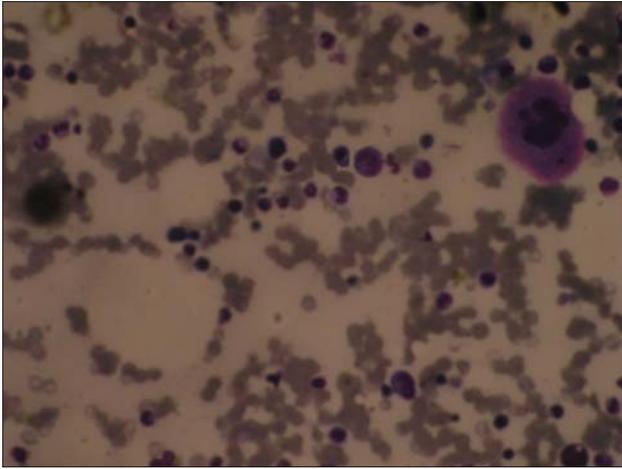


Figure 3: Bone marrow aspiration smear to rule out leukemia (Giemsa stain $\times 400$)

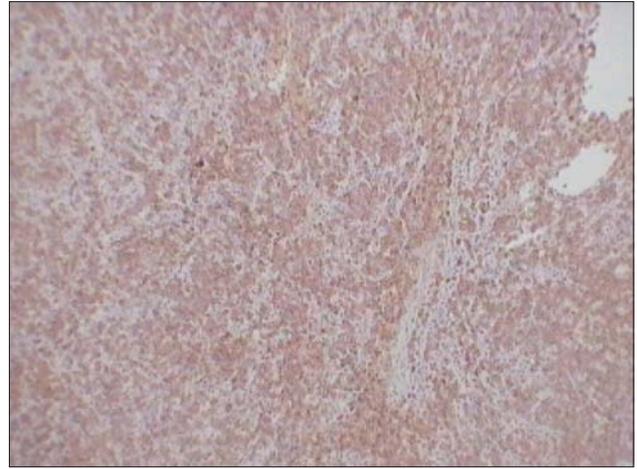


Figure 4: CD 30 +ve

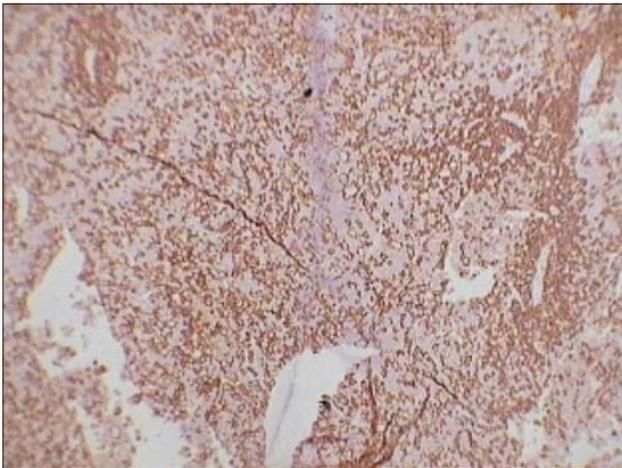


Figure 5: CD 45 +ve

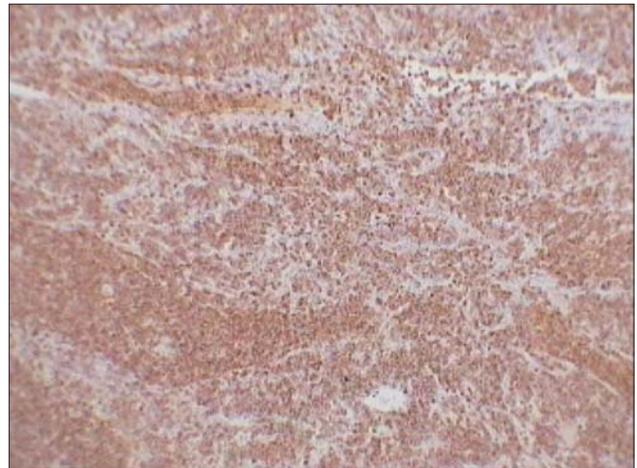


Figure 6: EMA +ve

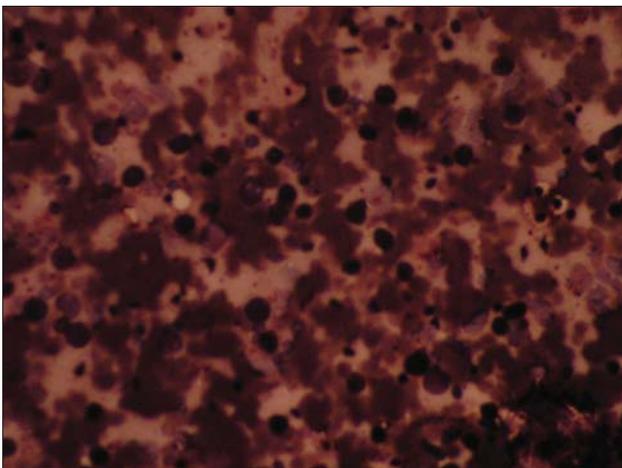


Figure 7: FNA smears showing NSE positivity (NSE stain $\times 400$)

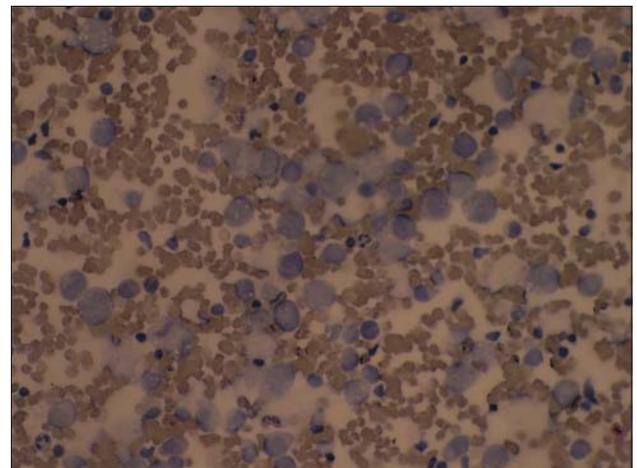


Figure 8: FNA smears showing MPO negativity (MPO stain $\times 400$)

29 cases of extramedullary myeloid cell tumors were studied. In this study, immunohistochemically the most sensitive antibodies were CD43 +ve in 96.6% of cases, MPO

in 91.3% cases, anti-lysozyme in 96.5%, CD45 +ve in 60% of cases.^[8] In myeloid sarcoma cases immunohistochemistry gives the final diagnosis, but in our case cytochemistry

on the FNA smears was more helpful in confirming the diagnosis.

In summary, this tumor is very often misdiagnosed as malignant lymphoma because of cytomorphologic and histologic similarities of the blasts to large cell lymphoma. A careful search for immature myeloid and erythroid cells on FNAC and biopsy is a useful clue to the diagnosis accompanied with cytochemistry and appropriate immunophenotyping. It is therefore, essential to obtain adequate material for ancillary testing in the form of additional passes in FNA cytology. It is important for the physicians to treat these patients on the lines of leukemia and not lymphoma as this entity may be aptly called as a "soft tissue leukemia".

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