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Myeloid Sarcoma Masquerading As Lymphoma- A Diagnostic Challenge

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Abstract: Myeloid sarcoma (MS) is a rare extra-medullary tumor and its presentation as generalized lymphadenopathy is still rarer. It can mimic lymphoma clinically radiologically, cytologically and even histologically, presenting as a diagnostic challenge. This is an unusual case of myeloid sarcoma in a 52 year old man who presented with generalized lymphadenopathy and then detected to have chronic myeloid leukemia. Clinical, radiological and cytological features favored a diagnosis of lymphoma. But histopathological evaluation of cervical lymph node, simultaneous bone marrow trephine biopsy and immunohistochemical work up helped us to exclude lymphoma and confirm the diagnosis of myeloid sarcoma. BCR/ABL translocation analysis confirmed the diagnosis of associated chronic myeloid leukemia. We report this case to emphasize the need of appropriate use of ancillary tests like immunohistochemistry and cytogenetic study. Even though generalized lymphadenopathy is a presentation of lymphoma, it can also be an early manifestation of MS. The correct timely diagnosis is crucial for proper patient management.
Keywords: Myeloid sarcoma, Chronic myeloid leukemia, Leukocytosis, Immunohistochemistry, Lymphadenopathy, Lymphoma

INTRODUCTION

Myeloid sarcoma also known as granulocytic sarcoma, chloroma or extra-medullary myeloid tumor is a rare tumor composed of myeloid blasts with or without maturation occurring at an anatomic site other than bone marrow [1]. Skin, lymph node, gastrointestinal tract, bone, soft tissue and testis may be affected. It may occur de novo or may precede or coincide with acute myeloid leukaemia (AML), myelodsplastic syndrome (MDS), myeloproliferative neoplasia (MPN) or MDS/ MPN. The major differential diagnosis is with malignant lymphoma.

CASE REPORT

A 52 year old man presented in our hematology department with one month history of swelling neck, recurrent fever and fatigability .On clinical examination, his vital signs were stable. He had right cervical, bilateral axillary and bilateral inguinal lymphadenopathy. Splenomegaly and mild hepatomegaly were here. CT scan thorax and abdomen showed multiple pretracheal, precarinal, mediastinal and left para aortic lymphadenopathy. Clinical and radiological differential diagnosis included tuberculosis and lymphoma. Fine needle aspiration cytology of cervical lymph node was performed and a diagnosis of non-Hodgkin lymphoma, diffuse large cell type was given. Peripheral blood examination showed anemia (Hb-7.9g %), leukocytosis (32.6X10 ^{3} /µL) and thrombocytopenia (78,000/cmm). A high erythrocyte sedimentation rate (ESR) was noted (120mm at the end of one hour).Peripheral smear revealed a leukoerythroblastic blood picture with thrombocytopenia. RBC_S showed many cells with tear drop morphology (Fig. 1). Immature cells with blast morphology constituted 4% of differential count, eosinophils 9% and basophils 3%.

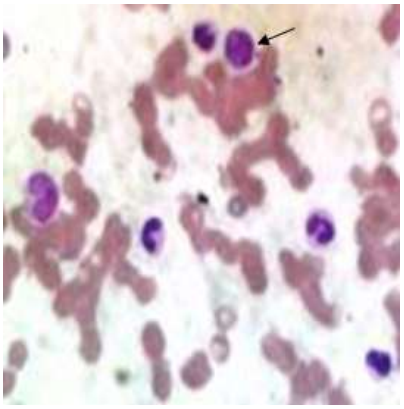

Fig. 1: Peripheral smear showing leukocytoses with cells at different stages of maturation Leishman stain x 1000. A myeloblast is shown (arrow)

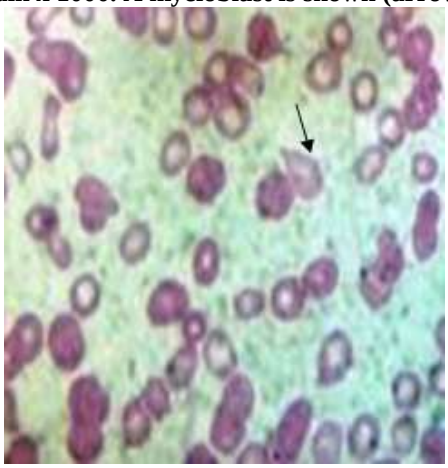

Fig. 2: RBCs with tear drop morphology (arrow). Leishman stain x 1000

Bone marrow showed mainly diluted marrow blood. Trephine biopsy showed a hypercellular marrow (Fig. 3a) with atypical cell infiltrate with focal areas of fibrosis. Cervical lymph node biopsy was performed simultaneously which showed effacement of nodal architecture by proliferating cells, majority having cytoplasmic granules (Fig. 3b & c). Predominant cells were medium sized with scattered population of large nucleolated cells. Immunohistochemical work up was done both in lymph node and bone marrow trephine biopsy sections. The atypical cell population in both showed CD117 positivity (Fig. 3d). Cells were negative for CD3, CD20, CD30 and CD15. Detection of BCR/ABL transcript using a real time quantitative RT-PCR assay confirmed the diagnosis of chronic myeloid leukemia. The patient was started on imatinib and clinical examination showed disappearance of all lymph nodes. His total WBC and platelet count came back to normal range.

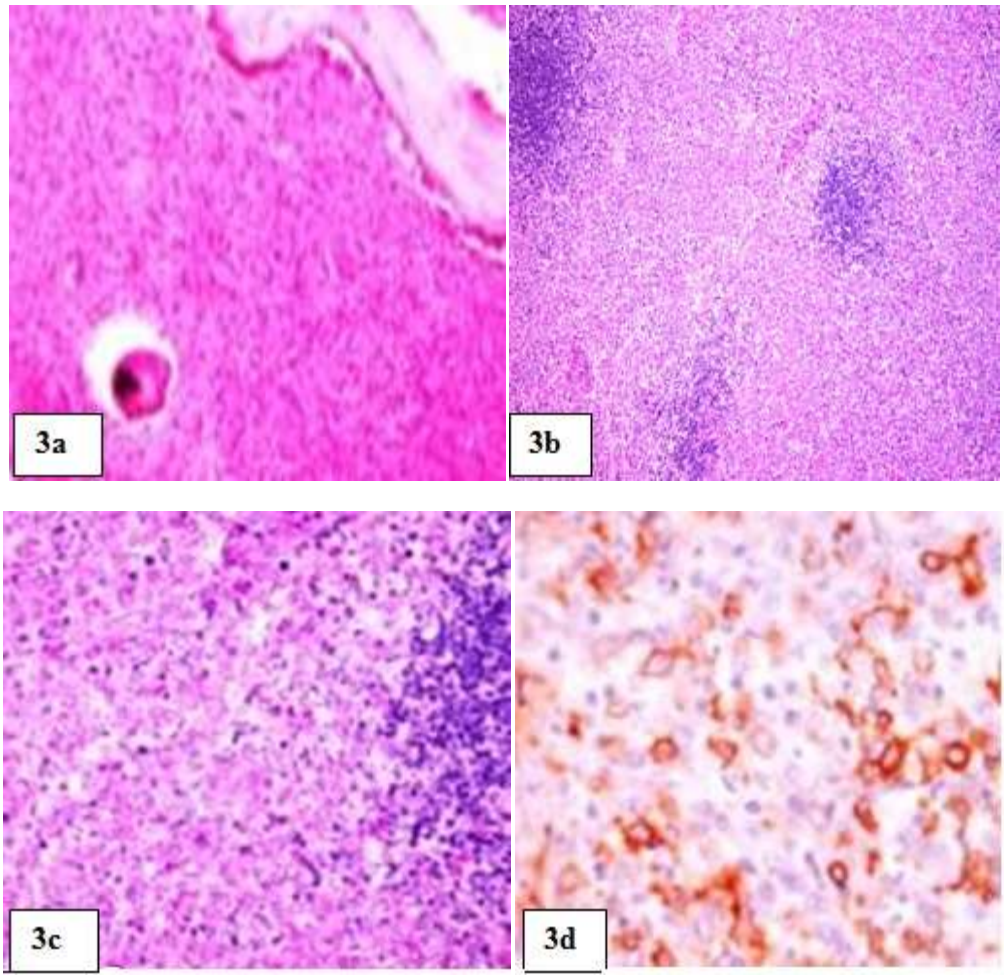

Fig. 3a: Bone marrow trephine biopsy showing hypercellular marrow; Fig. 3b: Lymph node showing effaced architecture by proliferating myeloid cells (Hematoxilin and eosin, 4x); Fig. 3c: Infilrating cells have immature nuclei and cytoplasmic granularity (Hematoxilin and eosin, 10x); Fig. 3d: CD117 positivity in lymph node infiltrate (40x).

DISCUSSION

Myeloid sarcoma is a localized tumor composed of myeloid blasts with or without maturation occurring at an anatomic site other than bone marrow. It was first described in the early 19th century. The incidence of myeloid sarcoma is 2.5-9.1% of the patients with acute myeloid leukemia and it is five times less frequent in patients with chronic myeloid leukemia [2] .The most common setting for MS is disease progression in acute myeloid leukaemia.The clinical and pathological diagnosis of myeloid sarcoma continues to be a problem. It can be challenging on hematoxylin and eosin sections alone. Immunohistochemical stains help to characterize the cell of origin and provide definitive diagnosis of myeloid sarcoma [3]. Immunohistochemically most useful antibodies include CD43, MPO, antilysozyme, CD117, CD34, CD68 together with CD99, CD3,CD20,CD 15 and CD30 [4].

Pileri *et al*. [5] in his study of 92 patients with myeloid sarcoma has emphasized the relevance of immunophenotyping in diagnosis of MS. In his retrospective study, ten MS cases were originally misdiagnosed as diffuse large B cell lymphoma because of the lack of proper immunohistochemical work up. In their study Mansi *et al*. illustrated the diverse presentations of MS and the extent to which they may mimic non Hodgkin-lymphoma [6].

Ghaleb *et al*. [4] has reported a case with generalised lymphadenopathy as the first presentation of MS(4). Like in our case, an initial diagnosis of malignant lymphoma was made. In patients with MPN and MDS, myeloid sarcoma defines a blastic transformation often associated with a short survival.

The differential diagnosis of myeloid sarcoma include non Hodgkin lymphoma (including precursor B or T cell, Burkitt, and diffuse large B cell lymphoma),small round cell tumors (including neuroblastoma, rhabdomyosarcoma ,Ewing's sarcoma, peripheral neuroectodermal tumor and medulloblastoma),undifferentiated carcinoma, malignant mastocytosis. In our case apart from lymphoma we also thought of the possibility of malignant mastocytosis. But the diagnosis was excluded by using special stain with metachromatic property (giemsa stain).

CONCLUSION

Myeloid sarcoma is a tumor very often misdiagnosed as malignant lymphoma especially when they present without any history of antecedent myeloproliferative disorder. A high index of suspicion should be there. So, careful evaluation of morphology for any evidence of myeloid differentiation, proper use of immunohistochemical markers and a high index of suspicion are required to avoid this important diagnostic error.

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