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MYELOID SARCOMA AS THE PRESENTING SYMPTOM OF CHRONIC MYELOID LEUKEMIA CHRONIC PHASE: A CASE REPORT

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ABSTRACT

Myeloid sarcoma is an extramedullary tumor of immature granulocytic cells. It is a rare condition; most often associated with acute myeloid leukemia but can be associated with myeloproliferative neoplasms or myelodysplastic disorders. The more frequent sites of involvement are skin, lymph nodes, gastrointestinal tract, bone, soft tissue and testis. We described a 30 years old female patient with myeloid sarcoma presented as mass in right elbow that ultimately was found to have chronic myeloid leukemia in chronic phase.

KEYWORDS: Myeloid Sarcoma**,** Chronic Myeloid Leukemia, Chronic Phase.

INTRODUCTION

Myeloid sarcoma also may be known as granulocytic sarcoma, myeloblastoma, extramedullary myeloid tumor, and chloroma. The term chloroma is derived from the Greek chloros, meaning green, as the tumor appears green when exposed to air.[1]

Tumor mass composed of myeloblast or immature myeloid cells occurring in an extra-medullary site due to migration of these blast cells to extra-osseous locations via haversian canals. It's precede or occur concurrently with acute or chronic myeloid leukemia, myeloproliferative disorders and myelodysplastic syndrome.[2-4]

Because of its rareness in extremities, it becomes diagnostic dilemma.If it is unrecognized for longer periods, appropriate chemotherapy may be delayed.Once diagnosed, myeloid sarcoma is treated according to the associated underlying disease state.[5,6]

CASE REPORT

A 30 years old previously healthy woman presented with a firm, fixed 3 cm soft tissue palpable mass in the right elbow. Imaging with MRI revealed single lesion in right elbow. Fine needle aspiration (FNA) was performed and histologic evaluation revealed granulocytic hyperplasia (Figure 1). Review of the peripheral blood revealed sever leukocytosis (>150,000) with an absolute neutrophilia, left shifted myeloid lineage, and 3% circulating blasts. The aspirate smear showed 2.3% blasts, sever hyper cellular aspirate especially of myeloid series with full spectrum of maturation, significant increment of basophil and eosinophil (Figure 2).

The bone marrow biopsy showed sever hyper cellular marrow, loss of fatty cells, marked increase in granulocytic precursors with a variable degree of left shift and moderate increment of eosinophil precursors. Megakaryocytes increased in number, occasionally forming small clusters, no obvious fibrosis and no granuloma are seen (Figure 3).

BCR-ABL probe detected fusion of ABL 1 proto-oncoge located at 9q34.1 within the breakpoint cluster region (BCR) at 22q11.Philadelphia chromosome, t (9; 22) (BCR-ABL) fusion gene is detectable in 98% of the cells. Molecular studies were positive for the t (9; 22) BCR/ABL1 major fusion transcript in 74% International Scale (IS) by reverse transcriptase–polymerase chain reaction (RT-PCR) quantification analysis. These findings confirmed the diagnosis of the chronic phase of chronic myeloid leukemia. This patient was treated with Imatinib 400 mg daily, 3 months later patient achieved complete hematological response and dramatic decrease in mass size.

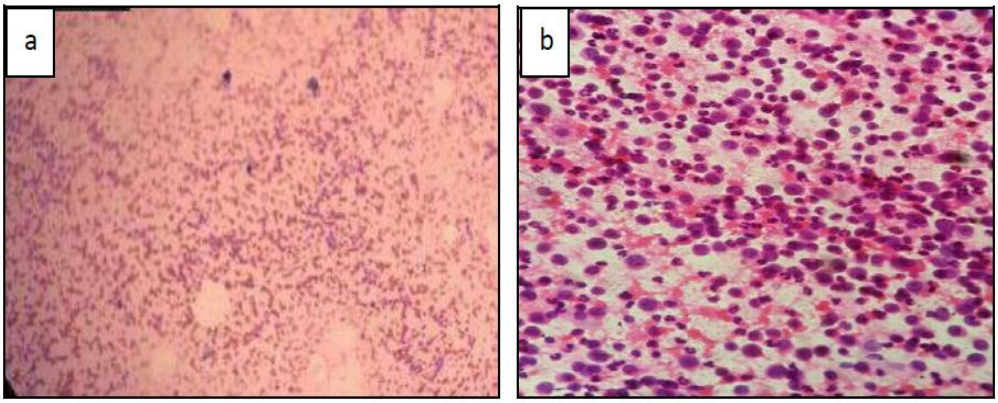

Figure. 1. Right elbow mas FNA (a) Low-power image (H &E, X100) and (b) High-power image (H &E, X400) showed granulocytic hyperplasia.

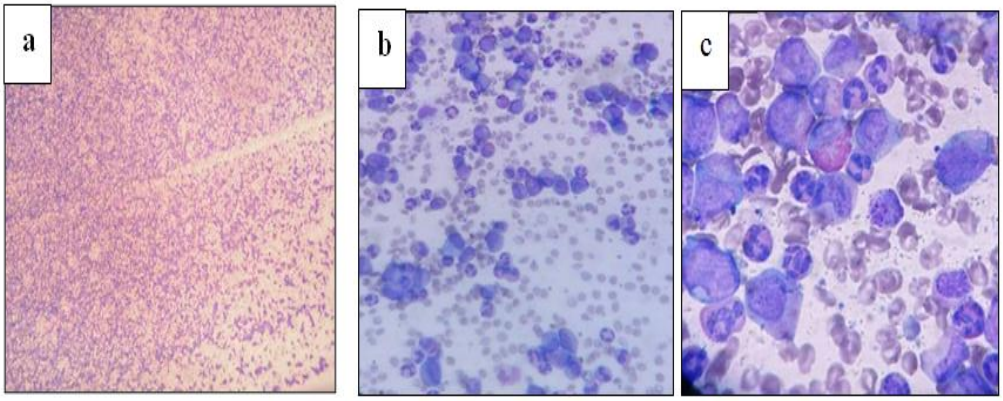

Figure. 2. Bone marrow aspirate smear (a) Low power (X100) leishman stain (b) Higher power (X400) leishman stain (c) At oil immersion (X1000) leishman stain showed granulocytic hyperplasia, majority of late and intermediate stage with eosinophilia & basophilia.

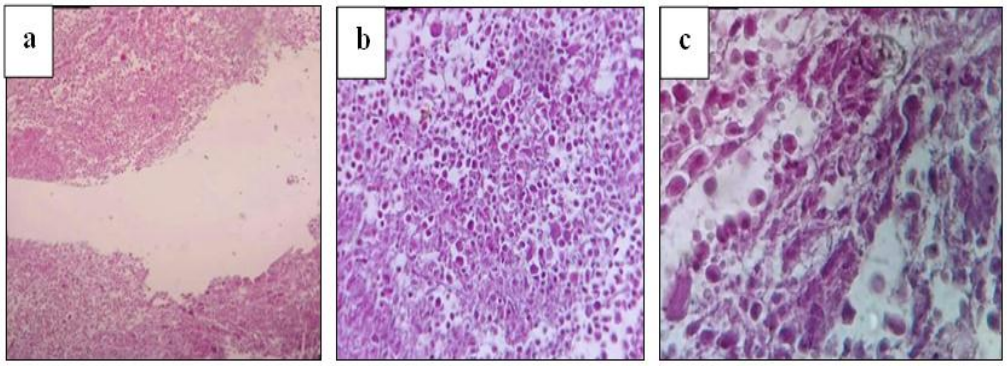

Figure3. Bone marrow core biopsy (a) Lower power (H & E, X100) (b) Higher power (H & E, X400) showed granulocytic hyperplasia and hyper cellular marrow with immature cells (c) At oil immersion (H&E, X1000) showed granulocytic hyperplasia.

DISCUSSION

The condition now known as chloroma was first described by the British physician A. Burns in 1811, although the term chloroma did not appear until 1853.However, because up to 30 % of these tumors can be white, gray, or brown rather than green, the more correct term granulocytic sarcoma was proposed by Rappaport in 1967 and has since become virtually synonymous with the term chloroma.[7,8]

Literature review revealed that myeloid sarcoma mostly misdiagnosed as non-Hodgkin lymphoma, but can also be mistaken for Ewing sarcoma/primitive neuroectodermal tumor and undifferentiated carcinoma. Immunophenotyping by immunohistochemistry for expression of myeloid associated enzymes is essential for identifying myeloid sarcoma.[2,9]

Skin is the most common areas of involvement that is typically appears as violaceous, raised, nontender plaques or nodules, which on biopsy are found to be infiltrated with myeloblasts. Other tissues which can be involved include lymph nodes, colon, small intestine, mediastinum, bone, central nervous system, epidural sites, uterus, testis and the ovaries.[5,8,10-12] In 1902 Dock & Warthin established the link between myeloid sarcoma and acute leukemia which remains the common disease entity associated with this presentation with incidence between 2.5-9% in various series. While myeloid sarcomas was uncommon in chronic myeloid leukemia and other myeloproliferative disorder.[13-15]

There is little data in literature available regarding myeloid sarcoma in chronic phase chronic myeloid leukemia and most cases associated with accelerated phase and blast crisis.[16-19] Tyrosine-kinase inhibitors (TKIs) has improve survival up to 40 months in patient in myeloid sarcoma presenting at chronic phase chronic myeloid leukemia comparing to pre TKIs era. Cases presented in chronic phase less resistance to TKIs and better overall survival than cases presented in accelerated phase or blast crisis.[16,20]

CONCLUSION

Myeloid sarcoma in chronic phase of chronic myeloid leukemia is rare and need to be evaluated as it can be first warning sign of progressive disease. It carry better prognosis in chronic phase than in accelerated or blast phase, clinicians should be aware of this presentation to avoid delay in diagnosis and management.

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