Granulocytic Sarcoma Disguised as Lymphoblastic Lymphoma: A case report

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Abstract

Acute myeloid leukemia (AML) is a malignant clonal disease of hematopoietic stem cells affecting the blood and bone marrow. Granulocytic sarcoma (chloroma) is a rare disease characterized by an extramedullary myeloid cell mass with or without bone marrow infiltration. We hereby report a case of AML presented with granulocytic sarcoma causing cord compression.

background

Acute myeloid leukemia (AML) is a complex genetic disorder characterized by rapid proliferation of myeloid blast cells in the bone marrow resulting in decreased normally differentiated red blood cells, white blood cells and platelets.[1] Clinical presentation of AML is always related to bone marrow failure. Fever, susceptibility to infections and bleeding diathesis are the most common manifestations. Granulocytic sarcoma, also known as myeloid sarcoma (MS) is rare extramedullary tumor of immature granulocytic cells. it may be detected before, simultaneously with or during the course of AML, it is less commonly observed in myeloproliferative neoplasm (MPN) and myelodysplastic syndrome (MDS).[2]

MS occurs in any site of the body, yet most common locations include the soft tissue, bone, periosteum, and lymph nodes; Central nervous system (CNS) involvement is rare. Owing to diverse locations of MS, the clinical presentation of the disease is variegated as well and depends upon the site and the size of the tumor.[3]

Objectives

We present a case report of acute myeloid leukemia with extramedullary intradural granulocytic sarcoma causing cord compression.

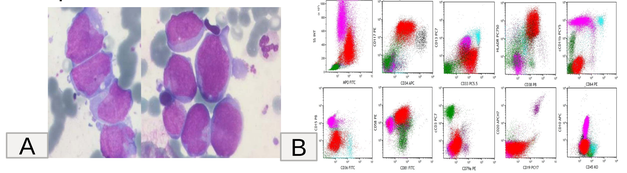
Case report

An 18 –year- old male presented in January 2022 with progressive lower limb motor and sensory impairment followed week later by bladder and bowel dysfunction. Pan CT scan showed anterior mediastinal amalgamated lymph node measures 3.5x6 cm. MRI study detected dorso-lumbar intradural extramedullary lesion 5x8x6cm and heterogeneously enhancing pre-sacral soft tissue mass measures 3.2x7x6 cm. Excision and pathological examination of the dorsal mass showed infiltration by sheets and aggregates of malignant round cell tumor with diffuse positive reaction of the tumor cells by LCA,CD99, PAX5 and BCL2, with 80- 90% of the cells positively stained by Ki 67; picture suggestive of B cell Lymphoblastic Lymphoma. CBC: WBC: 6.5x10^9/l, ANC: 2.4x10^9/l, Hb: 11.9 g/dl, MCV: 88.6 fl, platelets count: 217x10^9/l. bone marrow examination showed 70% blast cells positively stained for MPO,CD117, CD33,CD34, HLADR and CD58 and negatively stained for TdT, CD79, cCD3,CD22. CD19, CD10 and CD20 likewise furthermore immune stain of the spinal mass. Cytogenetic analysis revealed positive inv(16)(p13.1q22) Picture consistent with acute myeloid leukemia.

Treatment commenced and the patient achieved complete remission following 2 induction therapies with “7+3” regimen. On the clinical level, the patient showed noticeable improvement in sensation and motor power. The patient was under high dose Cytarabine while the article is written.

Summary

An -18- year old male presented with dorso-lumbar and pre-sacral extramedullary intradural masses causing cord compression. Excision and preliminary pathological examination of the dorsal mass showed picture consistent of B- cell lymphoblastic lymphoma. Despite normal complete blood count, the bone marrow examination, flowcytometric and cytogenetic analysis revealed acute myeloid leukemia. The patient received “7+3” double induction chemotherapy and complete remission has been achieved with remarkable improvement in both sensory and motor power. Patient was under high dose Cytarabine and continued in complete remission till the article is written.



A: BMA morphology: Leishman-stained bone marrow aspirate (BMA) smear reveals the infiltration with “blast like” cells that resemble non-Hodgkin lymphoma or lymphoblastic leukemia. **B_FC IPT:** Flow cytometric Immunophenotyping of bone marrow aspirate (BMA) specimen showing that the malignant cells (red) express MPO, dim to negative CD13, CD33, CD117, dim CD34, CD38, HLADR, CD58, CD81, and dim CD45 but are negative for CD64, CD11b, CD14, CD36, CD15, CD19, cCD79a, CD10, CD20, and cCD3.Green: T cells, Purple: B-cells, Fuchsia: granulocytes, Aqua: monocytes

Discussion

Myeloid sarcoma is a pathologic diagnosis for an extramedullary proliferation of blasts of one or more of the myeloid lineages that disrupt the normal architecture of the tissue it resides.[4]The presence of MS suggests there might be an aberrant homing signal for the leukemic blasts precluding the more common bone marrow localization. Among the postulated mechanisms of the ability of myeloid sarcoma to invade the surrounding tissues is owed to specific interaction between the matrix metalloproteinase (MMP)-9 and the leukocyte surface beta (2) integrin is required for the migration of AML-derived cells.[5] Diagnosis of MS is a challenge particularly in absence of clinical suspicion of myeloid malignancies. Competent diagnostic clinical, laboratory and radiological imaging of granulocytic sarcoma are crucial for proper management.

References

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