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ABSTRACT
Extramedullary (EM) manifestations of acute leukemia include a wide variety of clinically presentation that often pose difficulty in diagnosis and treatment of Myeloid sarcoma. We present a case of a 5-year-old male with initial complaints of radicular pain of both lower limbs and urinary retention. On MRI, compression by epidural mass was identified, which was shown to be an extramedullary myeloid sarcoma diagnosed on the basis of auer rods containing blasts in peripheral blood smear and bone marrow. Diagnosis was confirmed with flow cytometry and induction chemotherapy was started. Initial neurological presentation of paraplegia due to chloroma is extremely rare in myeloid leukemia with very few case reports published earlier.

Keywords: Myeloid, Sarcoma, Extramedullary, Flowcytometry.
INTRODUCTION
Myeloid sarcoma (MS), also termed extramedullary acute myeloid leukemia, extramedullary myeloid tumor, and Granulocytic Sarcoma (GS) have also been referred to as chloroma secondary to their characteristic green color created by the presence of myleperoxidas [1]. It is a rare manifestation that is characterized by the occurrence of 1 or more tumor myeloid masses occurring at an extramedullary site with effacing the architecture of tissue. Chloroma are most frequently seen in acute myeloid leukemia (AML) and few cases in myelodysplasia and other myeloproliferative disorders. MS present with wide clinical manifestation; GS should be considered in the differential diagnosis of an epidural mass along-with other diagnosis. GS virtually can occur in any organ or tissue, most common sites being - skin (called leukemia cutis) and bones. Other sites are lymph nodes, mediastinum, epidural sites, small intestine, ovary/testis and brain. Treatment for chloroma consists of systemic chemotherapy for the underlying leukemia, and the lesions frequently respond well. If the lesion is refractory to systemic chemotherapy, then surgical debridement or radiation therapy may be considered. Here we present a case of five- year old male with initial presenting symptom of paraplegia due to granulocytic sarcoma which is extremely rare.

CASE REPORT
A 5-year-old boy presented with initial complaints of radicular pain and progressive weakness of both the lower limbs of two months duration followed by difficulty in passing stools and urine for the last one week. There was no history of trauma, significant weight loss, or contact with tuberculosis. On examination, he had pallor; vitals were normal. No lymphadenopathy or hepatosplenomegaly was present. Higher mental function status and cranial nerve examinations were normal. Upper limbs power was normal while in lower limbs, muscle tone was decreased and grade 1/5 power was seen in both the lower limbs. Sensations were intact in both lower limbs. Initial routine blood investigations were done outside and showed hemoglobin 5.7 gm%, total count 14,800 cells/cu.mm, polymorphs 45%, lymphocytes 50%,
eosinophil 02% and monocyte 03%. RBCs Mean cell volume (MCV) and Mean cell haemoglobin (MCH) and Mean corpuscular haemoglobin concentration (MCHC) were reduced as well as platelets count were reduced. MRI examination showed an ill-defined contrast enhancing lesion in posterior epidural space extending from D2 to D10 level, displacing the cord anteriorly with signal intensity alternation and cord compression from D2-D10 level causing compressive neuropathy. Then, peripheral smear was done again in our laboratory which showed [Figure 2] mild reduced of MCV of RBCs. WBC count was normal and showed predominantly blast cells with high nuclear cytoplasmic ratio, moderate cytoplasm, hyperchromatic nuclei with multiple nucleoli. Platelet count was decreased. Bone marrow [Figure 3] examination was ordered following the presence of blast cells in peripheral smear and it showed that the marrow elements were completely replaced with blast cells having high nuclear cytoplasmic ratio, moderate amount of eosinophilic cytoplasm, and hyperchromatic nuclei with multiple nucleoli. Focal Auer rods and few maturing cells of the myeloid series were present. The marrow picture was corresponding to AML M2. Considering the possibility of GS as the cause of paraplegia in acute myeloid leukemia, the patient was started on induction chemotherapy.

DISCUSSION

Myeloid sarcoma (MS) (chloroma, granulocytic sarcoma, extramedullary myeloid tumor), may be defined as extramedullary solid tumor mass of immature myeloid cells or monoblast cell which disrupt the architecture of the tissue associate with or without bone marrow involvement [2]. MS can be primary, which is very rare and secondary when it is associated with acute myeloid leukemia approximately 3-10%, but rarely it can also associated with Myelodysplasia and myeloproliferative disorder. MS can occur in any age group but predominantly 35-50 years of age, as well as MS can occur at any site but most common site skin, periosteum, bone, lymph node, soft tissues, beside this numerous other sites has also been reported [3]. MS has wide differential diagnosis because of various sites of presentation, along with wide variation of age and symptom presentation. Most common differential diagnosis include round cell tumor such as lymphoblastic lymphoma,
medulloblastoma, rhabdomyosarcoma, ewing/ PNET along with other differential such as undifferentiated epithelial cell tumor [4].

Myeloid sarcoma diagnosis requires various diagnostic tools beside morphology in peripheral blood smear, such as special cytochemistry, specific marker for immunophenotyping, and cytogenetic, all play important role in diagnosis of myeloid sarcoma.

On the basis of cytomorphology, myeloid sarcoma classified as blast (which included myeloblasts with little evidence of promyelocyte,) immature (shows an intermediate degree of differentiation contains principally myeloblasts and promyelocytes; eosinophil myelocytes are usually present) and mature myeloid cells (primarily consists of promyelocytes and later stages of maturation with abundant Eosinophil myelocytes). Blast variant of MS has differential diagnosis with lymphoblastic lymphoma, carcinoma, melanoma while immature and mature variant of MS with Hodgkin lymphoma, extramedullary haematopoiesis and infections.

Cytochemistry test include myeloperoxidase, naphol ASD chloroacetate esterase and Non-specific estrase reaction but their role in diagnosis has been replaced by immunophentyping methods by flow cytometry. Immunophenotyping panel for diagnosis of myeloid sarcoma includes myeloperoxidase, lysozyme, CD68(KP-1 and PGM-1), CD34 and CD117,CD 43, CD 3, CD 20 are very useful in differentiation for MS from B and T cell lymphoma [5].

Rarely myeloid cell differentiated into erythroid and megakaryocyte linage which can be differentiated by marker for megakaryocyte CD41, CD61 and for erythroid series glycophorin a, haemoglobin A.

Cytogenetic nowdays play an important role in the diagnosis of MS, however no specific cytogenetic has been still discovered for MS. Cytogenetic usually done on blood or bone marrow samples by fluorescence in situ hybridization (FISH). Most commonly cytogenetic abnormality noted in MS are ranslocation t (8; 21), inversion of chromosome 16 (Inv 16) alongwith with few rare cytogenetic abnormality are monosomy 7 or 5 and trisomy 8 [6].

Myeloid sarcoma either primary or secondary both possess common treatment regimen conventional AML-type chemotherapeutic protocols. The role of other therapy (radiotherapy, haematopoetic stem transplantation, and targeted therapy)
method has not been well established. If local symptoms (e.g., compromise of the spinal cord due to myeloma) than surgery is considered.

Myeloid sarcoma is rare entity hence it has conflict regarding its prognosis. There has been no variation in prognosis of MS either as isolated or associated with AML reported.

**CONCLUSION**

This article is yet another classic example showing that myeloid sarcoma can have varied clinical presentations, based primarily on the site of involvement. Early suspicion of the diagnosis, peripheral blood smear, bone marrow and appropriate investigations can aid in early diagnosis of myeloid sarcoma with timely initiation of chemotherapy for the patient.

**AUTHOR’S CONTRIBUTIONS**

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Conception and design, Acquisition of data, Analysis and interpretation of data
Drafting the article, Critical revision of the article
Final approval of the version to be published.

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Acquisition of data, Analysis and interpretation of data
Drafting the article,
Critical revision of the article

REFERENCES


FIGURE LEGENDS

Figure 1: MRI of the dorsal spine showing an epidural mass lesion D2-D10 level.

Figure 2: Peripheral blood smear showing blast cells with auer X 100 (Leishman stain).

Figure 3: Bone marrow smear showing blast cells s/o acute myeloid leukemia X 100 (Leishman stain).
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