Granulocytic sarcoma of epidural mass with acute myeloid leukemia

**Garima Singh, Ashutosh Kumar, Mili Jain, Rashmi Kushawa,
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ABSTRACT

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CASE REPORT

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Keywords: Extramedullary, Flow cytometry, Myeloid, Sarcoma

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INTRODUCTION

Myeloid sarcoma, also termed extramedullary acute myeloid leukemia, extramedullary myeloid tumor, and granulocytic sarcoma have also been referred to as chloroma secondary to their characteristic green color created by the presence of myeloperoxidase [1]. It is a rare manifestation that is characterized by the occurrence of one 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. Myeloid sarcoma present with wide clinical manifestation, granulocytic sarcoma should be considered in the differential diagnosis of an epidural mass along-with other diagnosis. Granulocytic sarcoma 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

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to systemic chemotherapy, then surgical debridement or radiation therapy may be considered. Here we present a case of five-year-old boy with initial presenting symptom of paraplegia due to granulocytic sarcoma which is extremely rare.

CASE REPORT

A five-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 g/dl, total count 14,800 cells/mm³, polymorphs 45%, lymphocytes 50%, eosinophil 2% and monocyte 3%. The RBCs mean cell volume (MCV) and mean cell hemoglobin (MCH) and mean corpuscular hemoglobin concentration (MCHC) were reduced as well as platelets count were reduced. Magnetic resonance imaging (MRI) scan showed an ill-defined contrast enhancing lesion in posterior epidural space extending from D2–D10 level, displacing the cord anteriorly with signal intensity alternation and cord compression from D2–D10 level causing compressive neuropathy (Figure 1). Then, peripheral smear was done again in our laboratory which showed (Figure 2) mild reduced of MCV of RBCs. White blood cell 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 acute myeloid leukemia M2. Considering the possibility of granulocytic sarcoma as the cause of paraplegia in acute myeloid leukemia, the patient was started on induction chemotherapy.

DISCUSSION

Myeloid sarcoma (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].

Myeloid sarcoma 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.

Figure 1: Magnetic resonance imaging scan of the dorsal spine showing an epidural mass lesion D2–D10 level.

Figure 2: Peripheral blood smear showing blast cells with Auer (Leishman stain, x100).

Figure 3: Bone marrow smear showing blast cells suggested of acute myeloid leukemia (Leishman stain, x100).

Myeloid sarcoma can occur in any age group but predominantly 35–50 years of age, as well as it 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].

Myeloid sarcoma 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 myeloid sarcoma has differential diagnosis with lymphoblastic lymphoma, carcinoma, melanoma while immature and mature variant of myeloid sarcoma with Hodgkin lymphoma, extramedullary hematopoiesis and infections.

Cytochemistry test include myeloperoxidase, naphthol AS-D chloroacetate esterase and non-specific esterase reaction but their role in diagnosis has been replaced by immunophenotyping 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 myeloid sarcoma from B cell and T cell lymphoma [5].

Rarely myeloid cell differentiated into erythroid and megakaryocyte lineage which can be differentiated by marker for megakaryocyte CD41, CD61 and for erythroid series glycoprotein A, hemoglobin A.

Cytogenetic nowadays play an important role in the diagnosis of myeloid sarcoma. However, no specific cytogenetic has been still discovered for myeloid sarcoma. Cytogenetic usually done on blood or bone marrow samples by fluorescence in situ hybridization (FISH). Most commonly cytogenetic abnormality noted in myeloid sarcoma are translocation t(8; 21), inversion of chromosome 16 (Inv 16) along 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, hematopoietic stem transplantation, and targeted therapy) method has not been well established. If local symptoms such as compromise of the spinal cord due to myeloma are present, surgery is considered.

Myeloid sarcoma is rare entity hence it has conflict regarding its prognosis. There has been no variation in prognosis of myeloid sarcoma either as isolated or associated with acute myeloid leukemia 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 Contributions

Garima Singh – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Ashutosh Kumar – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Mili Jain – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Rashmi Kushawa – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Final approval of the version to be published

Arun A. Kumar – Substantial contributions to conception and design, Acquisition of data, Analysis and

interpretation of data, Final approval of the version to be published

Durg Pratap Singh – Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Guarantor

The corresponding author is the guarantor of submission.

Conflict of Interest

Authors declare no conflict of interest.

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