

# Paraplegia due to chloroma as the initial presenting feature of acute myeloid leukemia.

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**Abstract:** Granulocytic sarcoma (or chloroma) consists of neoplastic granulocytic precursors and myeloblasts. Paraplegia due to a spinal cord epidural chloroma is an extremely rare presentation of undiagnosed leukemia. Here, we report a case of 21-year-old-man who presented with acute paraplegia and urine retention due to thoracic epidural mass. The tumor was totally removed by surgery. The histopathological examination was consistent with granulocytic sarcoma. The bone marrow aspirate confirmed the diagnosis of acute myeloid leukemia (AML). After a subsequent systemic chemotherapy, the patient was completely asymptomatic. Granulocytic sarcoma should be considered in the differential diagnosis of an epidural mass in patients with or without acute leukemia, because early diagnosis followed by appropriate combined chemotherapy and radiation may obviate surgical intervention.

**Keywords :** Granulocytic sarcoma, spinal cord compression, acute myeloid leukemia.

## Introduction:

Granulocytic sarcoma (GS) or chloroma has been defined as an extra-medullary tumor mass consisting of myeloid blasts with or without maturation [2]. Paraplegia due to chloroma is an extremely rare presenting neurological manifestation in myeloid leukemia [1]. We are reporting a case of 21-year-old man, who presented with paraplegia due to dorsal epidural mass, as the initial presenting manifestation of acute myeloid leukemia. GS should be considered in the differential diagnosis of an epidural mass in patients with or without leukemia as GS represent a diagnostic challenge, particularly those occurring in patients without evidence of systemic disease [1].

## Case Report:

A 21-year-old man was admitted in neurosurgery department because of paraplegia. He was having progressive weakness associated with paresis of both lower limbs of three days duration. Later, he developed urine retention and paralysis of lower limbs without upper limb weakness or cranial nerve symptoms.

Physical examination on admission revealed a hypoaesthesia under the fourth thoracic level. The tendon reflexes were absent in lower limbs with bilateral extensor plantar response and the Babinski test was positive bilaterally. Cranial nerve examinations were normal. Other system examinations were normal.

Magnetic resonance imaging (MRI) showed extensive epidural lesion extending from D4 to D7, intermediate in T1, hyperintense in T2, dorsally compressing the spinal cord (Figure 1). The patient's blood cell count was pathologic: hemoglobin 8g/dl, white blood cell count  $3100/\text{mm}^3$ , platelet count  $44000/\text{mm}^3$  and 3% neutrophils, 44% lymphocytes and 44% blasts in peripheral smear. The coagulation test was normal. The patient underwent laminectomy of the dorsal spine (D4-D7) and the tumor was totally removed. Histological examination demonstrated fibroadipose connective tissue including diffuse cellular infiltrates. The cells in the infiltrate were middle to large in size, with prominent nucleoli and with slightly basophilic cytoplasm. Mitotic figures were abundant (Figure2). Immunohistochemically, the tumor cells were positive for myeloperoxidase (MPO) (Figure3) and negative for CD3, CD20. Considering the possibility of spine granulocytic sarcoma associated with leukemia, the patient was referred to haematology department. Bone marrow aspirate confirmed the diagnosis of acute myeloblastic leukemia (AML) of the French-American-British M2 subtype (Figure 4). Cytogenetic study of the bone marrow cells revealed a pathological karyotype: 46, XY, t (8, 21) (q22; q22) [9]/ 46, XY [6]. Cerebrospinal fluid cytology for malignant cells was negative. The patient was given a course of systemic chemotherapy according to the protocol of acute myeloid leukemia consisting of rubidomycine ( $45 \text{ mg}/\text{m}^2$  daily for 3days) and cytosine arabinoside ( $200\text{mg}/\text{m}^2$  continuous infusion for 7 days). There was a significant improvement in his paraplegia and the complete remission was achieved.

## Discussion:

GS (also termed myeloid sarcoma or chloroma) is a rare malignant neoplasm resulting from the extramedullary proliferation of myeloid blasts with or without maturation [2]. Macroscopically, chloromas are green in appearance due to the myeloperoxidase in the leukemia cells and fades when exposed to air [1- 4]. Granulocytic sarcoma occurs in 3% to 9% of AML [5]. Rarely chloroma can occur alone without peripheral blood or bone marrow evidence of leukemia [1]. Moreover, 66% -88% of patients with an isolated granulocytic sarcoma will develop AML at a mean of 9-11 months after diagnosis [6]. Granulocytic sarcomas are generally seen in the ribs, sternum, pelvis and orbital bones as well as in the soft tissues, lymph nodes, skin, and gums [3-2]. Involvement of the central nervous system (CNS) is rare, and spinal cord compression by granulocytic sarcoma is even more rare [3-2-7-8]. In our patient, paraplegia due to chloroma as the initial presenting feature of AML is very rare. Chloroma should be considered in the differential diagnosis of any epidural mass in patients with or without leukemia [1]. In the present case, the GS occurs with peripheral blood evidence of leukemia. The correct diagnosis of GS is obtained in only about 50% of non-leukemic patients due to its rareness and the histological and radiological similarities to malign lymphoma [2- 9]. Clinically and histologically, GS should be distinguished from Hodgkin lymphoma, Burkitt's lymphoma, large cell lymphoma, and small round cell tumor, including neuroectodermal tumor[3]. Immunohistochemical studies, especially staining of MPO, are extremely helpful to make a correct diagnosis. The characteristic myeloid sarcoma phenotype is CD45, CD43, CD68 and myeloperoxidase positive and CD20 and CD45 RO negative [2-9-10]. Myeloperoxidase, CD15 and neutrophil elastase are especially effective to

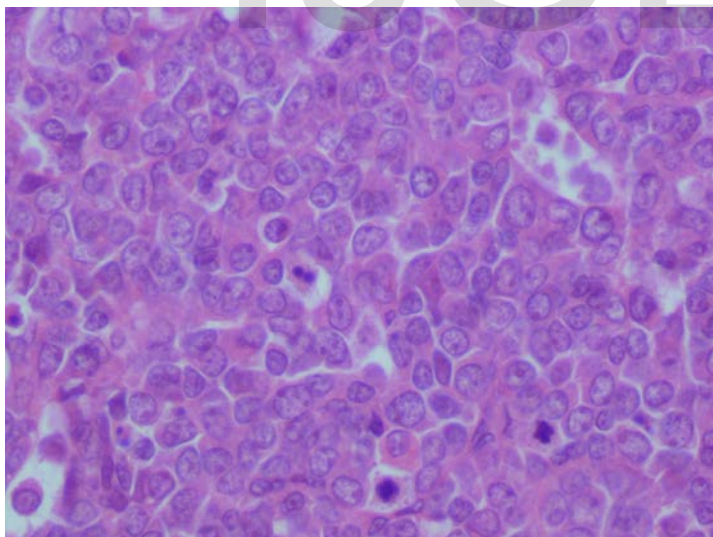
differentiate GS from other tumors. In the present case, the spinal GS was diagnosed by positive staining of MPO.

Chromosome 8 abnormalities were the dominant abnormalities in the bone marrow and/or extramedullary sites. Translocation between chromosomes 8 and 21 is common in granulocytic sarcoma but not requisite [2]. In our case, the cytogenetic study of the bone marrow cells revealed this translocation.

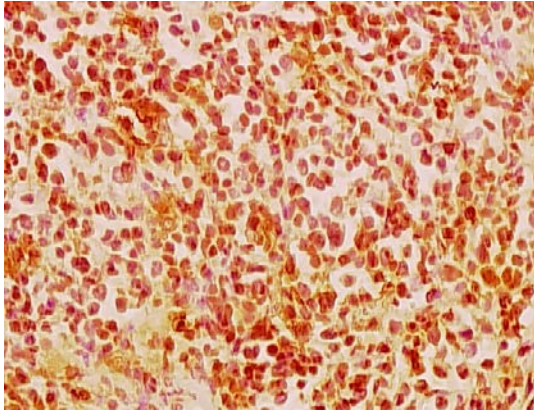
Optimal therapy for patients with GS is not yet well defined. GSs are usually radiosensitive and are often treated with local radiotherapy and chemotherapy [11-12]. Newly diagnosed patients with isolated GS usually treated with aggressive chemotherapy as if they have acute myelogenous leukemia, cures are not attained with radiation therapy alone [11]. Surgery is generally preferred for cases of acute spinal cord compression in cases without systemic evidence of leukemia [12]. In our case, the surgical intervention was occurred despite the evidence of leukemia in peripheral blood. Early diagnosis followed by appropriate combined chemotherapy and radiation may obviate surgical intervention [1]. The early systemic chemotherapy after surgery is beneficial and may induce complete remission as in our case [3-14]. So, prompt diagnosis and adequate treatment are essential to achieve a good outcome. In conclusion, Chloroma, though rare, should be considered in the differential diagnosis of any epidural mass lesion and peripheral smear examination and bone marrow study, if necessary, should be done in all cases.



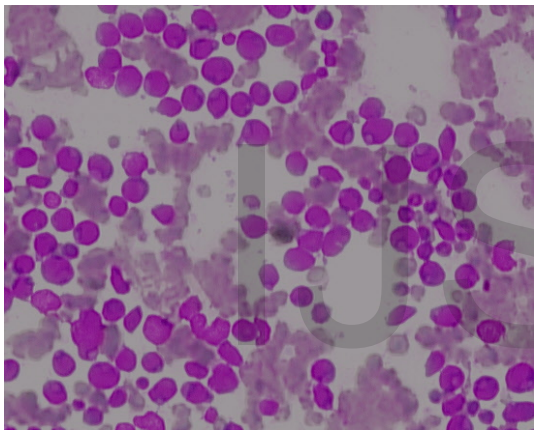
**Figure 1:** MRI of the dorsal spine (sagittal T2) showing extensive epidural lesion extending from D4 to D7.



**Figure 2:** Hematoxylin and Eosin staining of epidural mass specimen showing immature myeloid blastic cell infiltration (original magnification x400).



**Figure 3:** Positive staining for myeloperoxidase in granulocytic sarcoma of the spine (original magnification x400).



**Figure 4:** bone marrow showing blast cells.

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