



## Isolated Spinal Granulocytic Sarcoma in Adolescent Patient: Case Report

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### Abstract

Granulocytic Sarcoma (GS) or what are called chloroma, myeloblastoma and myeloid sarcoma) is an uncommon extramedullary solid tumor of immature myeloid cells. It occurs primarily in Acute Myelogenous Leukemia (AML) but also may arise in patients with other myeloproliferative disorders.

It could occur at any part of the body mostly in the bone, soft tissue or lymph node, rarely involving the Central Nervous System (CNS) and causing spinal cord compression, it could occur during the course of disease, at relapsed or as Isolated Granulocytic Sarcoma (IGS) without bone marrow involvement.

We report a 17 years old boy with IGS of the vertebral body causing spinal cord compression and bilateral lower limb weakness treated with surgical decompression, laminectomy and biopsy followed by chemo-irradiation and currently the he is on regular follow up.

**Keywords:** Granulocytic sarcoma; Chloroma; Cord compression; Acute myeloid leukemia

### Abbreviation

T: Thoracic; L: Lumbar; MRI: Magnetic Resonance Imaging; CD: Cluster of Differentiation; i.v: Intravenously; mg: Milligram; g: Gram

### Introduction

Granulocytic Sarcoma is a rare extramedullary mass composed of immature myeloid cells most commonly found in association with Acute Myeloid Leukemia (AML) [1]. The chloroma may be the initial manifestation of leukemia and can occur in different anatomic sites. MS is most commonly found in the orbit or periorbital and the diagnosis can be difficult especially when they precede the bone marrow involvement [2]. About 47% of MS are initially misdiagnosed mostly as malignant lymphoma but also as Ewing's sarcoma, rhabdomyosarcoma and neuroblastoma.

Is a rare form (<1%) of extramedullary Acute Myeloid Leukemia (AML) presenting without marrow involvement [3].

We reported a case of isolated spinal GS which was successfully treated with decompression surgery, irradiation, and chemotherapy with intrathecal injection.

### Case Presentation

A 17 year old adolescent boy was referred to us by neurosurgeon for further assessment and treatment.

He was presented with localized severe back pain, asthenia in the legs and inability to walk for 12 days associated with numbness and paresthesia, no history of trauma to back. There is no consanguinity of parents, neither family history of malignancy nor similar condition, and high socioeconomic status, neither previous medical nor surgical problems. At presentation neurological examination revealed a 1/5 power in both legs, loss of pain and sensory perception below the T4 level, hyperactive deep tendon reflexes and positive Babinski reflex. There were neither other neurological abnormalities nor organomegaly. At causality MRI of the whole spine revealed hyperintense bony lesions on the L1 to L5 vertebral bodies, spinal cord compression by multiple extradurally masses were also observed between the T2 and T4, and between the T12 and L2 vertebral levels (Figure 1A and 1B).

After starting him on Dexamethasone 8 mg intravenously four time daily, he was underwent

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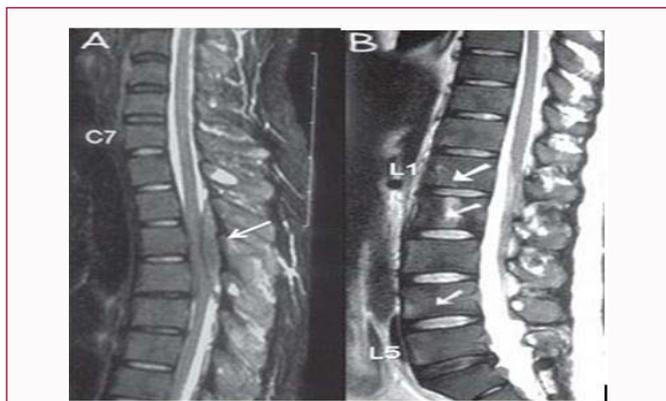
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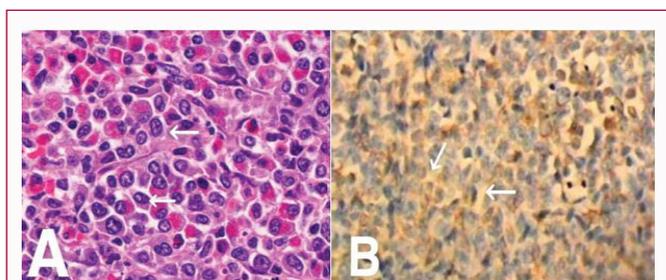
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**Figure 1A and B:** Whole spine MRI: Shows spinal cord compression by multiple extradurally masses were also observed between the T2 and T4 and between the T12 and L2 (white arrow), hyperintense bony lesion L1-15 (white arrow).



**Figure 2A:** Pathological examination revealed involvement by a myeloid neoplasm of atypical immature cells with irregular nuclei and delicate chromatin with many immature eosinophils by Hematoxylin-Eosin-stain (H&E), (B) Myeloperoxidase (MPO) stain was positive(original magnification x200).

surgical decompression and open biopsy.

Post-operatively the patient was stable, oriented, afebrile, and well looking with normal vital Signs.

Complete blood count revealed hemoglobin content was 12.7 g/dl, the platelet count was 350,000/ml, and a leukocyte count of 8,000/ml with 55% neutrophils, 44% lymphocytes, 1% monocytes, 0% eosinophils, and 0% basophils with normal blood film.

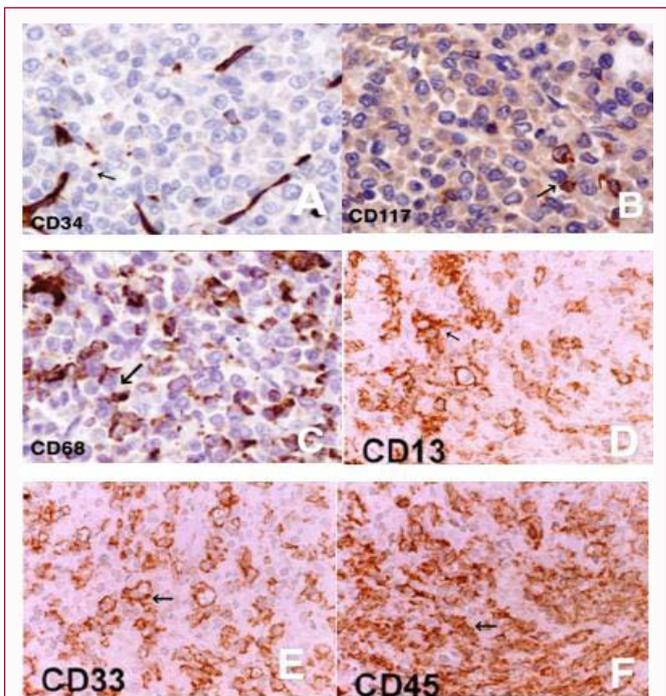
Bone marrow aspirate and biopsy were cellular without any other abnormalities; cerebrospinal fluid cytology was negative, with normal chest X-ray.

The histopathology evaluation of the mass revealed malignant cell infiltration with oval nuclei, eosinophilic cytoplasm by Hematoxylin-eosin-stain. Tumor cells with eosinophilic cytoplasm are diffusely infiltrating the bone tissue (original magnification x100) Myeloperoxidase (MPO) stain was positive (Figure 2A and 2B).

Immunohistochemical stains panel was positive staining in the atypical cells with CD34, CD117, CD68, CD13, CD33 and CD45 tested positive (Figure 3) and negative for CD20, CD79a, CD138, CD15, CD3 and CD5.

Based on normal bone marrow finding and immunohistochemistry stains the diagnosis was isolated granulocytic sarcoma of the spinal cord.

PET scan was not done because it was locally not available and radiotherapy was started urgently at a dose of 20 Gy, administered



**Figure 3:** Immunohistochemical study positive for (A) CD34, (B) CD117, (C) CD68, (D) CD13, (E) CD33 and (F) (CD45) (original magnification x200).

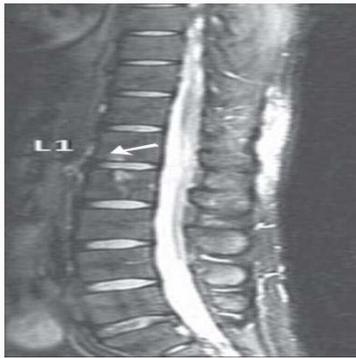


**Figure 4:** MRI 4 weeks after radiation showing no of mass intraspinally (A); but few bony lesions on T and L vertebral bodies were persist (white arrows in B).

in 1.5 Gy to 3.5 Gy fractions with systemic induction chemotherapy according to the MRC AML protocol consisted of daunorubicin 60 mg/day intravenous (i.v.) on days 1, 2 and 3 and cytarabine (200 mg/day continuous i.v. on days 1 to 10 for 1 course, followed by daunorubicin 60 mg/day intravenous (i.v.) on days 1, 2 and 3 and Ara-C (200 mg/day continuous i.v. on days 1 to 8). The patient also received 2 intrathecal injections containing 15 mg hydrocortisone, 30 mg Ara-C and 15 mg methotrexate each time.

Clinically, the patient's lower limbs motor power gradually improved with aggressive physiotherapy and was became 5/5; the patient started walking and sensory deficits disappeared. After finishing of radiotherapy and the induction chemotherapy, MRI of the spine showed no evidence of the masses intraspinally but regressing bony lesions persist (Figure 4A and 4B).

According to the chemotherapy protocol the patient receive



**Figure 5:** Spinal MRI after completion therapy showing a few residual bony lesions on L1 vertebral bodies (white arrows).

two additional consolidation courses of high dose cytarabine (1.5 g/day i.v. over 3 h on days 1, 3 and 5) without further intrathecal chemotherapy, the treatment course was uneventful.

Spinal MRI was done 1 month after completion of the consolidation chemotherapy, showed that the all bony lesions on vertebral body disappeared with few residual bony lesion on the L1 and L2 (Figure 5).

The family refused to do neither new biopsy nor additional investigations so the patient kept on regular follow up and he is now 6 years after finished his chemo-irradiation without any complications or disease recurrence.

## Discussion

Myeloid sarcoma was first described in 1811 and later named chloroma by King because of the green color caused by the presence of myeloperoxidase granules in the malignant myeloid cells [4-7]. Overall, myeloid sarcoma represents a rare hematologic phenomenon with an incidence of 2.5% to 9.1% in patients with AML more frequently in those with myelomonocytic and monocytic morphology (M4 and M5 French-American-British subtypes) [8-11]. It could occur concomitantly, following, or, rarely, before the onset of systemic bone marrow leukemia [4,9,10]. The prevalence of myeloid sarcoma in the spine is estimated to be less than 1% among all patients with acute and chronic myeloid leukemia [7].

Granulocytic sarcomas have approximately the same rate of occurrence in both sexes. Children are more often affected than adults: 60% of patients are younger than 15 years old [12].

In this case report, we presented an isolated granulocytic sarcoma which initially manifested as inability to walk, bilateral legs weakness and sensory deficit. Spinal MRI of our patient revealed hyperintense bony lesions on the L1-L5 vertebral bodies. Extradurally masses were also observed between the T2 and T4, and between the T12 and L2 vertebral levels causing bilateral lower extremity weakness that quickly progressed to paraplegia. Patient underwent surgical

decompression of tumor and biopsy revealed isolated granulocytic sarcoma for which he putted immediately on chemo-irradiation and high dose steroid, he respond well to the treatment and the time this manuscript was prepared the patient was undergoing regular follow up for the last uneventful 6 year.

## Conclusion

Isolated Granulocytic Sarcoma (IGS) should be considered in the differential diagnosis of patient experiences localized severe backache with motor and sensory deficit and. When Granulocytic sarcoma located in spinal column was suspected radiologically, an urgent MRI of the spine should be done with high dose steroid for those with evidence of cord compression diagnosed, surgical decompression, irradiation with chemotherapy that used for acute myeloid leukemia postoperatively constituted the treatment of choice for this tumor. Notably, treating GS in the same manner as AML may benefit patients with isolated spinal GS but large studies and new agents are needed for the treatment of isolated AML patients.

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