

Chronic myeloid leukemia with myelopathy: Rare complication of common malignancy

Faheem Arshad, Bashir Ahmad Shah¹, Showkat Ahmad Mufti¹, Irshad Ahmad Parry¹

Departments of Internal Medicine and ¹Emergency Medicine, Sher-i-Kashmir Institute of Medical Sciences, Srinagar, Jammu and Kashmir, India

ABSTRACT

Myelopathy due to a spinal cord epidural mass is an extremely rare presentation of hematological malignancies. We report a case of 50-year-old male who was diagnosed with chronic myeloid leukemia 2 years back and was on imatinib. He presented with paraplegia due to spinal epidural mass likely chloroma. Chloroma or granulocytic sarcoma should be considered in the differential diagnosis of an epidural mass in patients with hematological malignancy. Chronic myeloid leukemia (CML) is a common chronic myeloproliferative disorder, both bleeding and thrombosis occur less frequently during stable phase. Our patient presented with paraplegia with an epidural mass likely chloroma which settled after emergent treatment, receiving radiotherapy and steroids. We believe this is the first case report of myelopathy in a patient of previously diagnosed CML in stable phase showing marked improvement after treatment.

Key words: Chronic myeloproliferative disorders, extramedullary tumor, Granulocytic sarcoma

INTRODUCTION

Chloromas (granulocytic sarcomas) arise from myeloid blast cells and infiltrate the spinal extradural area. They are rare manifestations chronic leukemia and can occasionally precede the development of systemic disease by weeks to years.^[1] They may occur in the extramedullary spinal canal and cause spinal cord compression or cauda equina syndrome.^[2] Only a few cases of granulocytic sarcoma presenting with symptoms of spinal cord compression in patients without myelogenous leukemia have been reported.^[3] Myelopathy due to chloroma is an extremely rare presenting neurological manifestation in myeloid leukemia.^[4] Although chloromas are most frequently seen in acute myeloid leukemia (AML), it may signal the onset of accelerated phase of chronic myeloid leukemia (CML) or blastic transformation of a myeloproliferative disorder.^[2] It should be considered in the differential diagnosis of an epidural mass in patients with or without leukemia as

they represent a diagnostic challenge, particularly in stable phase of chronic myeloid leukemia in which it is very rare^[5] as in our patient and timely diagnosis is important for appropriate treatment. Chloromas have been reported to be more common in blast phase of leukemia. Aim of presenting this case is to remind an emergency physician about basic pathogenesis and urgent management of spinal cord compression in myeloproliferative disorders with radiotherapy, which has excellent results. We believe this the first case report of CML in stable phase presenting with spinal cord compression due spinal mass likely chloroma.

CASE REPORT

50-year-old male diagnosed as a case of CML 2 yrs back, had BCR/ABL translocation positive by reverse transcriptase polymerase chain reaction (RT-PCR) and was started on imatinib, presented to our tertiary institute with 1 week history of gradual onset progressive weakness lower limbs with worsening for last one day and urinary retention. On general physical examination he had pallor and vitals were normal. Systemic examination revealed moderate splenomegaly 4 cm below costal margin with neurological examination revealing truncal weakness, power was grade 1/5 MRC (Medical Research Council) both lower limbs and bilateral extensor plantars and complete sensory level below T-10. Cranial nerve examination including fundus examination were normal.

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Address for correspondence: Dr. Faheem Arshad, 133 Housing Colony, Sanat Nagar, Srinagar - 190 005, Jammu and Kashmir, India.
E-mail: faheem2285@gmail.com

Investigation revealed hemoglobin of 11.5 gm%, total cell count 6500 with neutrophils 72% and lymphocytes 18%, and platelets count 98000, there were no blasts in Peripheral blood film (PBF), suggesting a stable phase of CML. Kidney function test (KFT) and liver function test (LFT) was normal. T1-weighted MRI [Figure 1] whole spine revealed multiple lesions with hypointense signals suggestive of marrow infiltration, with evidence of intraosseous extension of neoplastic infiltration into prevertebral space at T3 and T10 level, in addition there was also evidence of epidural mass at T3 level causing cord compression [Figure 2 showing T3, T11 and T12 vertebral bodies show mild compression]. Invasive procedure for knowing the exact nature of epidural mass was not performed because of patient refusal. Final diagnosis was CML with myelopathy likely chloroma. Patient was managed with steroids and radiotherapy and there was improvement in truncal weakness and power of lower limbs improved to MRC grade 3/5.

DISCUSSION

CML is a chronic myeloproliferative disorder characterised by a reciprocal translocation between chromosomes 9 and 22 and thereby formation of the Philadelphia chromosome. Spinal cord involvement in chronic myeloproliferative disorders, is a rare neurological complication. The common clinical manifestations of chronic myeloproliferative disorders are hemorrhage from the gastrointestinal or genitourinary tract and thrombosis of blood vessels in the leg, central nervous system (CNS) or spleen. Spinal stroke has been reported in case of polycythemia vera (PV). Although CML is the common of the chronic myeloproliferative disorders but both bleeding and thrombosis occur least frequently in during stable phase of CML. They can rarely occur alone without peripheral blood or bone marrow evidence of leukemia.^[6]



Figure 1: T1 weighted MRI whole spine revealed multiple lesions with hypointense signals suggestive of marrow infiltration, with evidence of intraosseous extension of neoplastic infiltration into prevertebral space at T3 and T10 level

Grunberg *et al.*, first time described an acute myelopathy that developed in a 53-year-old woman with PV in whom spinal venous thrombosis was demonstrated at autopsy. Since then many cases of paraplegia in polycythemia vera have been reported cause being extramedullary hematopoiesis as duramater has functional hematopoietic capacity.^[7] The most common sites of extramedullary hematopoiesis are the liver and spleen. Other sites which can also be involved include kidneys, adrenal glands, pericardium, mediastinum, and sclera. Although extramedullary hematopoiesis is a common compensatory phenomenon to chronic hemolytic anemia it may also be seen in other conditions such as leukemia, spinal cord compression being a rare complication. Spinal cord compression secondary to extramedullary hematopoiesis especially of thoracic spine is very uncommon especially in patients with a chronic myeloproliferative disorder.^[8] Granulocytic sarcomas (GS) also known as chloroma, consist of myelocytic precursor cells forming an extramedullary tumor, occur most frequently in AML. They may signal the onset of the accelerated phase of CML or the blastic transformation of a myeloproliferative disorder. Chloromas of the spine presenting as paraplegia have been reported as an initial manifestation of acute leukemia or found in rare cases of aleukemic leukemia. In review of literature from 1902-2006 there have only four reports of paraplegia due to chloroma of the spine as the presenting manifestation of undiagnosed CML.^[9] It is distinctly rare for such tumors to cause epidural compression as a first manifestation of disease. We believe ours is the first case of a diagnosed CML in stable phase (may lead to accelerated phase) presenting with spinal cord compression due to chloroma. Critchley and Greenfield in their case series delineated neurologic manifestations of chloroma, in one of the cases at autopsy they found green deposits (chloromas) on duramater and also noticed cord vessels enclosed by chloromas appeared constricted and pale and on cross section proved to be anemic and gray matter could not be distinguished from the white columns and concluded that the most common

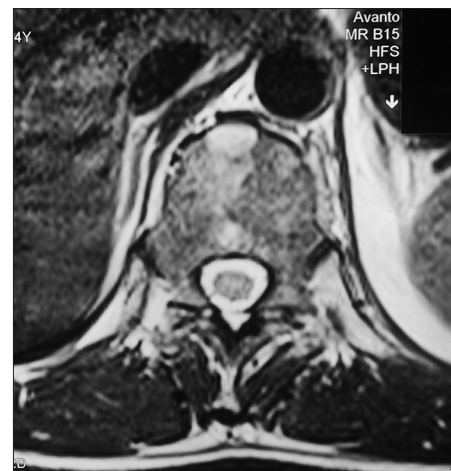


Figure 2: Epidural mass at T3 level causing cord compression

factor in the production of the paraplegia in chloroma was obliteration of the blood supply, or actual compression by cellular masses, which lowered its nutrition and led to the disintegration.

Diagnosis of spinal cord compression due to chloroma has been reported using conventional and CT myelography, CT, and MRI. But MRI, now the recommended test of choice,^[10] is able to visualize cord compression and paraspinal masses without contrast administration. It is also useful in differentiating chloroma from epidural abscess, neoplastic invasion and vertebral fracture from trauma. Because of its rarity, no guidelines for the treatment of chloroma exist. Management options include blood transfusion, radiotherapy, surgical decompression, hydroxyurea, or a combination of these modalities. Therapy usually depends on the severity of symptoms, size of the mass, patient's clinical condition, and previous treatment. Chloroma is highly sensitive to ionizing radiation, requiring a dose of only 10-30 Gy. Radiotherapy results in excellent local disease control and palliation of symptoms without significant toxicity, thus used now as the current recommended treatment.^[11] Excellent results have been reported with low-dose radiation as a monotherapy with 50% of patients having neurological improvement observed as soon as 3-7 days after initiation of treatment. Hematopoietic tissue being extremely radiosensitive undergoes shrinkage after radiotherapy. Dosages reported range 900-3,500 cGy. The main drawback of radiotherapy is risk of recurrence.^[12] In the past, surgical excision followed by radiation had been recommended. Surgery has the advantage of a definitive histologic diagnosis as well as immediate decompression. However, an invasive approach may be an unacceptable risk for the patient as was in our case. Our patient received radiotherapy within 24 hrs and there was a marked improvement in his power and currently on our follow-up.

To conclude paraplegia caused by CML as a result of granulocytic sarcoma is a rare presentation and an emergency physician should know basic pathogenesis and urgent management of spinal cord compression

myeloproliferative disorders to prevent permanent disability.

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