

Granulocytic Sarcoma with Compressive Myelopathy: A Rare Presentation of Chronic Myelogenous Leukemia

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ABSTRACT

Granulocytic sarcoma occurs most commonly in acute myelogenous leukemia. The appearance of granulocytic sarcoma in chronic myelogenous leukemia signals accelerated phase/ blast transformation. This is a rare case of undiagnosed chronic myelogenous leukemia with granulocytic sarcoma causing cord compression, which went into tumour lysis syndrome requiring dialysis after starting of steroids and radiotherapy. A 43-year-old male presented in emergency department with acute onset of flaccid paralysis. On clinical examination, there was hepatosplenomegaly and lower motor neuron paralysis in the lower limbs. The peripheral smear was consistent with chronic myelogenous leukemia in chronic phase. The MRI spine revealed para-spinal and epidural masses causing cord compression and the biopsy from the paraspinal mass was consistent with granulocytic sarcoma.

Keywords: Cord compression, Granulocytic sarcoma, Tumour lysis syndrome.

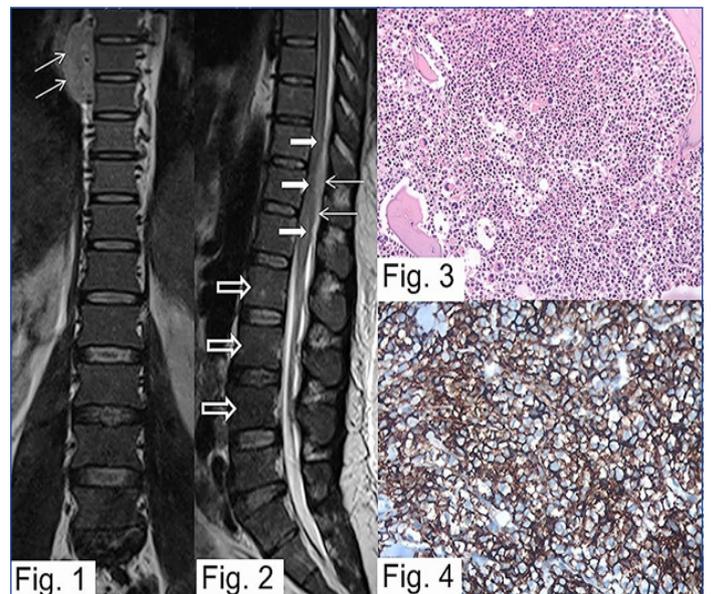
CASE REPORT

A 43-years-old male presented with a history of progressive back pain for last 4 days unrelieved by analgesics. He noticed progressive unsteady gait for two days and sudden onset of paraplegia on the day of admission. On clinical examination there was liver of 2cm and spleen of 10cm. Neurological examination showed flaccid paralysis of lower limb with bladder and bowel involvement. MRI spine showed para-vertebral soft tissue lesions at multiple levels, largest being on the right side at T6-T8 level [Table/Fig-1]. Epidural soft tissue component at the T9-11 level compressing and displacing the cord to the left side was associated with cord oedema [Table/Fig-2]. The biopsy from the paraspinal mass was composed of immature cells with round to irregular nuclei, fine chromatin and inconspicuous nucleoli, with moderate amount of eosinophilic cytoplasm. Few eosinophilic myelocytes were also seen. There is high endothelial venular proliferation. The immunohistochemistry on the malignant cells were as follows MPO+, CD 43+, CD20-, CD3-, focally positive CD 34 and MIB-75% [Table/Fig-3,4] The examination of the peripheral count revealed a leukocyte count of 2,52,600/mm³ with neutrophils- 58%, promyelocytes- 2%, myelocytes- 13%, metamyelocytes- 10%, band forms- 11%, basophils- 2%, eosinophils- 3%, blasts- 1% with 2 nucleated RBCs/100 WBCs. Hemoglobin was 11.6 grams/dl and the platelet count was 166,000/mm³. This profile was consistent with chronic myelogenous leukemia in chronic phase. The bone marrow aspiration done was consistent with chronic myeloid leukemia in chronic phase. The Fluorescent in Situ Hybridization was positive for BCR-ABL fusion (98% positive), reverse transcriptase polymerase chain reaction showed b3 a2 transcript and conventional cytogenetics revealed 46, XY, t(9;22) (q34;q11.2) (20). He was started on allopurinol, sodabarbonate, dexamethasone and cytoreductive doses of hydroxyurea. He received 30 gray RT in 10 fractions for reduction of the cord compression due to tumour. He went into tumour lysis syndrome (TLS) following steroids and radiotherapy with biochemical parameters as follows calcium-7mg/dl, phosphorus-6 mg/dl, creatinine 2.7mg/dl, potassium-6 mille-equivalent/liter. He required one sitting of dialysis on second day of starting the therapy following which the tumour lysis settled. The paraparesis recovered over next five weeks and currently he has lower limb power of 5/5.

He was later initiated on Imatinib and two months on treatment has achieved complete hematological response.

DISCUSSION

Chronic myeloid leukemia in chronic phase (CML-CP) presenting with compressive myelopathy is very rare [1-4]. Here, we report a patient with chronic myeloid leukemia in chronic phase (CML-CP) who presented with compressive myelopathy due to spinal granulocytic sarcoma (GS). Malignant spinal cord compression (MSCC) is an uncommon event occurring in cancer patients with



[Table/Fig-1]: T2-weighted coronal image through the lower thoracic and lumbar spine showing T2 hyperintense right paravertebral soft tissue (thin white arrows) at T6-8 level **[Table/Fig-2]:** T2-weighted sagittal image of the lower thoracic and lumbar spine shows diffusely hypointense marrow signal (hollow white arrows). Hyperintense soft tissue (thin white arrows) in the posterior epidural space compresses the lower thoracic cord at T10-12 level with swelling and hyperintensity of the cord extending from T9 upto the conus (solid white arrows) **[Table/Fig-3]:** Haematoxylin and Eosin (20X) section of tissue showing monotonous population of malignant cells **[Table/Fig-4]:** Immunohistochemistry showing CD43 positive cells in tissue at 40 X

Reference	Year Reported	Patient age/gender	Tumor location	Length of survival
Present case	2013	43/M	T6-T8 Paravertebral mass	3 months alive on follow
			T9-11 Epidural mass	
Barbara Bryant et al.,[1]	2006	53/F	T4-T6	4 months
Widhyde et al.,[2]	1963	12/F	T6	8 months
Rand and Rand et al.,[3]	1960	6/F	T1-T5	5 months
Critchley and Greenfield et al.,[4]	1930	14/M	T2, T7	4 months

[Table/Fig-5]: Paraplegia due to GS of the spine at the initial manifestation of undiagnosed CML

cumulative incidence of 2.5% during the course of the disease [5]. The commonest tumour associated with MSCC is multiple myeloma with the incidence being 1.95% [5]. MSCC is a rare event in leukemia with cumulative incidence of 0.3% along the course of the disease [5]. Acute myeloid leukemia rarely presenting as GS causing cord compression has reported in literature [6,7]. There have been only four case reports of CML presenting as paraplegia at presentation due to GS of spine and this probably is the fifth case to the best of our knowledge in literature [1-4] [Table/Fig-5]. Patient with GS in CML have poor prognosis [1-4,8] [Table/Fig-5]. In cases with cord compression related to metastatic deposit, surgical decompression compared to radiotherapy has better chance of achieving ambulation [9]. However, in our case we gave radiotherapy as there were tumour deposits at multiple levels, so surgical decompression was not possible. He responded to radiotherapy and regained his lower limb power, currently is ambulatory with normal bowel, bladder functions. The tumour lysis syndrome requiring dialysis, which occurred post RT and steroids was probably due to steroid responsiveness of the tumour with superadded effect of RT.

CONCLUSION

Unique features of our case are CML presenting with compressive myelopathy due GS and patient later went into TLS after he was

exposed to steroids and RT. While clinically evaluating a case of CML with focal neurological deficit, GS as cause must be kept in differential diagnosis.

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