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Extradural Granulocytic sarcoma causing acute paraparesis

Arshad A. Siddiqui

Naim-ur Rehman

CASE REPORT

EXTRADURAL GRANULOCYTIC SARCOMA CAUSING ACUTE PARAPARESIS

Arshad A. Siddiqui and Naim-ur-Rehman

ABSTRACT

A case of 9 years old female presenting with rapidly progressive paraparesis during remission phase of acute myeloblastic leukemia is reported. Radiological imaging revealed an extradural mass in the upper dorsal spine producing significant cord compression. The patient showed a dramatic neurological recovery after spinal cord decompression and subsequently treated with appropriate chemotherapy and local radiotherapy.

KEY WORDS: *Spinal cord compression. Spinal epidural tumor. Chloroma. Granulocytic sarcoma. Extramedullary myeloid leukemia. Acute myeloid leukemia.*

INTRODUCTION

Granulocytic sarcoma is a localized malignant neoplasm composed of immature cells of the granulocytic series. It was first called chloroma by King because of the greenish color of the fresh specimens, which was thought to be secondary to the presence of myeloperoxidase.[1,2] The lesion was first described by Burns in 1811 and Rappaport renamed these tumors as granulocytic sarcoma to emphasize their mesodermal origin.[3] More recently the term extramedullary myeloid cell tumors has been proposed to include both leukemia cell infiltrate that do not form destructive tumor mass and true granulocytic sarcoma.[4] Granulocytic sarcoma usually appears in association with the leukemia or myeloproliferative disorder and may rarely present as first manifestation in aleukemic patients.[4,5] This case report describes granulocytic sarcoma presenting with acute spinal cord compression as the first manifestation during remission phase of acute myeloblastic leukemia.

CASE REPORT

A 9 years old female child, diagnosed as a case of acute myeloid leukemia one year back, presented in the emergency room with 6-month history of intermittent dull pain in the interscapular region. A week prior to presentation, she started having numbness, stiffness and weakness in both her lower limbs, which rapidly progressed to the extent that she was unable to walk without support. There were no bladder or bowel symptoms.

On clinical examination, there was mild kyphus deformity in the upper dorsal spine associated with marked tenderness in the interscapular region. There was marked spastic paraparesis (the weakness being more pronounced distally) and bilateral sustained ankle clonus. The sensory level was at the xiphisternum and tendon reflexes were exaggerated in the both lower limbs. Abdominal reflexes were also diminished but peri-anal sensations were intact. The position sense was impaired and the plantar response was extensor bilaterally.

The white cell count was 4700/cu.mm and the differential count revealed no blast cells. The hemoglobin level was 11.3 gm/dl and platelets count was also normal. The bone marrow biopsy revealed no hypercellularity and abnormal cells. Plain x-ray dorsal spine showed erosion and destruction of the pedicles of the upper dorsal spine, most prominent from 2nd to 6th thoracic vertebral level. Magnetic resonance imaging of the dorsal spine revealed a spindle shaped mass in the extradural space, extending from the 2nd to the 6th thoracic vertebrae, causing marked compression of the cord. The mass was isointense on T-1 weighted images and showed marked enhancement after gadolinium administration (Figures 1 and 2).

Emergency decompressive laminectomy was performed, from 2nd to 7th thoracic vertebral levels with adequate decompression of the spinal cord. The tumor was firm and greyish white occupying the epidural space, extending laterally upto the vertebral pedicles. It was not adherent to the periosteum but to the underlying dural sac, which was intact. Peroperative fresh frozen analysis of biopsied tissues revealed myeloid deposits.

Detailed histopathology revealed differentiated morphological pattern characterized by presence of promyelocytes, eosinophilic myelocytes and mature granulocytes including neutrophils. Immuno-staining

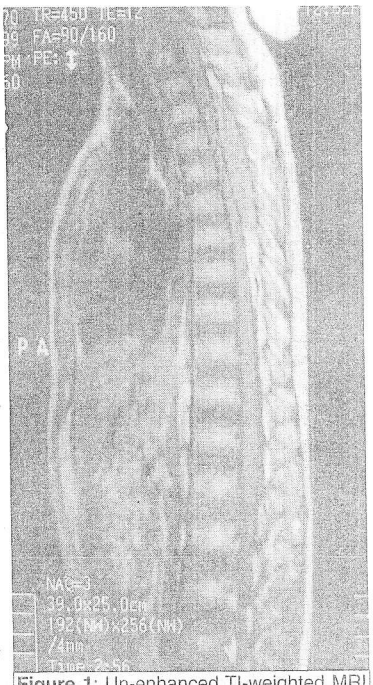

Figure 1: Un-enhanced TI-weighted MRI (sagittal view) showing a lesion at the upper dorsal spine causing cord compression from posterior. The lesion is isointense to the spinal cord.

Department of Neurosurgery, The Aga Khan University Hospital, Karachi.

Correspondence: Dr. Arshad A. Siddiqui, Chief Resident Neurosurgery, The Aga Khan University Hospital, Stadium Road, P.O.Box 3500, Karachi. E-mail: doc4brains@yahoo.com

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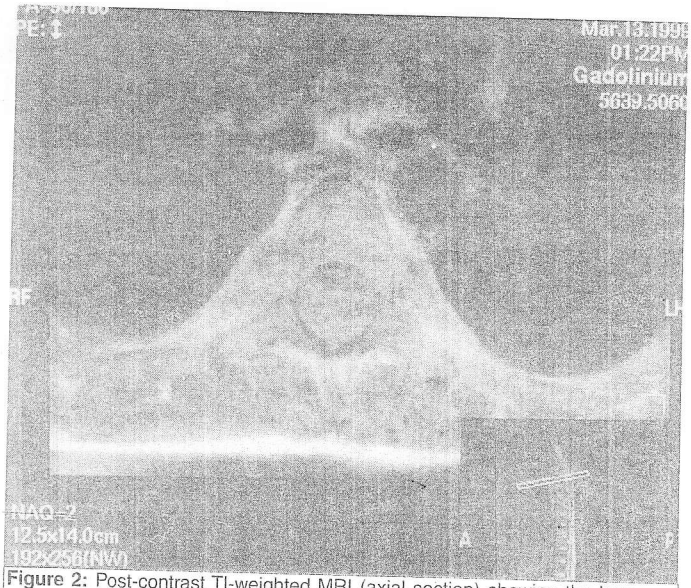

Figure 2: Post-contrast TI-weighted MRI (axial section) showing the homogeneously enhanced lesion causing spinal cord compression and the cord is flattened anteriorly.

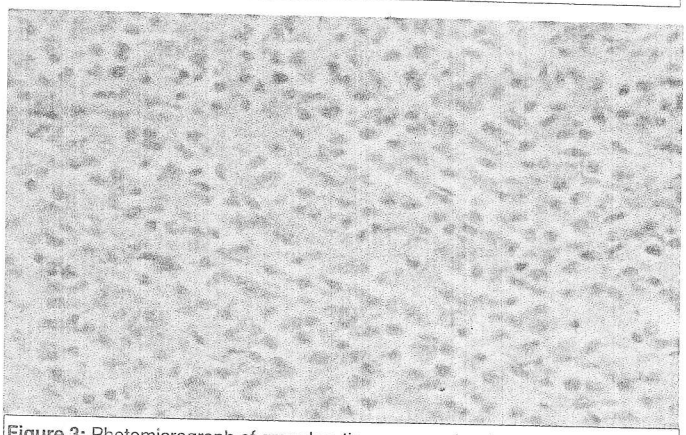

Figure 3: Photomicrograph of granulocytic sarcoma showing mature and immature granulocytes (H and E stain: Mag: X200).

of tumor cells showed positivity for CD45 (leucocyte common antigen) antibody. These features were compatible with the diagnosis of granulocytic sarcoma, differentiated type (Figure 3).

Postoperatively, the patient showed very dramatic improvement and she started walking on 3rd postoperative day. Chemotherapy was continued and she was discharged after one week of hospital stay for the local radiotherapy. She was quite well without any neurological deficits after one year of follow-up.

Discussion

Granulocytic sarcoma is a rare malignant neoplasm, with a reported incidence of 2.9% to 3.1% in patients with myeloid leukemia or myeloproliferative disorders.[5,6] The association of granulocytic sarcoma and acute leukemia was first established by Dock in 1893.[7] Granulocytic sarcoma may occur in association with acute myeloid leukemia,[2] myelodysplastic disorders with leukemic transformation, chronic myelogenous leukemia with impending blast crisis and in non-leukemic patients with no evidence of acute myeloid leukemia, as a myeloproliferative disorder, or a myelodysplastic syndrome at presentation.[4,5] Granulocytic transformation heralds the presence of a myelogenous leukemia or reflects an enlarging tumor cell burden with a potential to set up a blast crisis.[4,7] During remission, when the disease process is well-controlled, appearance of granulocytic sarcoma with acute presentation with rapidly progressive paraparesis, as in our case, is quite unusual.

Granulocytic sarcomas may involve any part of the body either concurrently, or sequentially. The orbit and subcutaneous tissue are the most common tumor sites while the spinal cord is a very infrequent site.[6,8,9] In spinal MR imaging, the tumors are isointense relative to gray matter, muscle, bone marrow or both muscle and bone marrow on T-1 weighted images, and isointense relative to white matter, muscle, bone marrow or both muscle and bone marrow on T-2 weighted images.[8-10] All granulocytic tumors enhance homogeneously with intravenous gadopentate dimeglumine.[8]

The histogenesis of granulocytic sarcomas of the spine is undetermined. Embryonic hematopoietic nests in the spinal dura mater could be the cells of origin.[3,5] An origin from myeloid precursor cells in the bone marrow is less likely because of the localized nature of the disease.[4,7]

The occurrence of granulocytic sarcoma, in patients with myeloproliferative disorders, has a great prognostic significance. The appearance of granulocytic sarcoma is the initial manifestation of blast transformation, which prelude to rapid progression within a few months.

The infrequent presentation of these tumors as acute cord compression may become a diagnostic dilemma; upto 75% of these tumors may initially be misdiagnosed, usually as malignant lymphoma.[5-7,9] The identification of eosinophilic myelocytes, which are present only in well-differentiated granulocytic sarcomas, is a helpful morphological clue.[3,4,7] Chloracetate esterase and antilysozyme immunoperoxidase staining and electron microscopy are used to confirm the diagnosis.[3,4,7] Decompressive laminectomy is required to establish the histological diagnosis and to relieve spinal cord compression immediately as is done in our case. But the mainstay of therapy is local radiotherapy with adjuvant chemotherapy as these tumors are highly radio-chemosensitive with a very good clinical outcome of 3.5-16 years.[2,5] The patient with granulocytic sarcomas is treated with a chemotherapeutic regimen similar to that used for acute myeloid leukemia.[5,6] The chemotherapeutic protocols for intermediate-grade, large cell malignant lymphoma, with which granulocytic sarcomas are commonly confused, is entirely different.[7]

This case report emphasizes that presentation of granulocytic sarcoma, as first manifestation during the remission phase, may become a diagnostic dilemma, being a rare tumor of the spine. So these tumors should be included in the differential diagnosis. Urgent surgical intervention is critical for institution of an immediate appropriate therapy as these tumors have a very good prognosis.

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