CASE REPORT

Granulocytic sarcoma, an undiagnosed leukemia, initially manifested as paralysis

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Abstract

Granulocytic sarcoma (GS) may be a presenting sign of myelogenous leukemia. Occasionally, an extramedullary neoplasm composed from myelocytic precursor cells occurs in patients without evidence of leukemia. Rarely, undiagnosed leukemia occurs initially manifesting with paralysis to spinal cord GS. We present a case report of 20-year-old girl with an undiagnosed leukemia, initially manifesting as paralysis. En bloc spondylectomy with chemotherapy postoperatively constituted the treatment of choice for this tumor. After two courses of chemotherapy, the patient made a good postoperative recovery with notable bilateral lower extremity improvement.

INTRODUCTION

Granulocytic sarcoma (GS), firstly described by Burns in 1811, is a tumor composed of myelocytic precursor cells in extramedullary sites. Based on a degree of maturation we can distinguish three types of tumors such as blastic, immature and differentiated. The term "chloroma" was given to the lesion by King (1853) because of the green color after exposure to air what is caused by high concentration of myeloperoxidase in myelocytic cells (King 1853). Appearance of tumor has been reported to arise in three clinical settings: (1) as a harbinger of acute myelogenous leukemia (AML) in nonleukemic patients, (2) as a signal of the accelerated phase of chronic myelocytic leukemia (CML), (3) in association with blastic transformation of a myeloproliferative disorder (Neiman *et al.* 1981; Landis & Aboulafia 2003). According to most medical literature, the lesion can be found in any anatomical site, including the skin, lymph nodes, bone, soft tissue (Saito *et al.* 1998), and brain (Tsimberidou *et al.* 2003). Without presence of serious abnormality of peripheral hemogram or bone marrow picture, GS can be easily misdiagnosed as non-Hodgkin's lymphoma or other carcinomas.

Rarely, GS may be the manifestation of undiagnosed leukemia. The length of time from the diagnosis of GS to the development of leukemia may vary from months to years. Among all tumors, causing compressive myelopathy, acute and chronic leukemia represent <3%. Malignant spinal cord compression is a rare event in leukemia with a prevalence of 0.07% at diagnosis and a cumulative incidence of 0.3% along the course of the disease (Loblaw *et al.* 2003).

The Granulocytic sarcoma we present was a large mass, filling the entire spinal canal. The unusual appearance was initially manifested with paralysis.

CASE REPORT

A 20-year-old Chinese girl with previously undiagnosed leukemia presented with a chief complaint of chronic low back pain with referral into the buttocks and posterior thigh for 4 months. The pain has been described as sharp and lancinating, and relieved by acetaminophen or bed rest. One month prior to hospitalization, she noted the unsteady gait with right lower extremity weakness, numbness, and tingling. So did the left extremity later. It quickly progressed to complete bilateral lower extremity paralysis and urinary and fecal incontinence. No major problems in her previous medical history were reported. Physical examination revealed flaccid paralysis presented in both lower extremities upon admission to the orthorpedics department. Bilateral knee jerk, posterior tibial tendon reflex and ankle jerk were all absent. Babinski reflex and Oppenheim reflex of right lower extremity were positive, while negative in the left one. Straight leg-raising test: positive in left, negative in right. The liver and spleen were not palpable.

Magnetic resonance images of her spine revealed a soft tissue mass (8.0×6.0×2.0 cm^3) involving the L4–L5 vertebral levels and extending into the spinal canal, resulting in severe spinal stenosis and cord compression (Figure 1).

Examination of her blood revealed a leukocyte count of 9,000/ml with 38% neutrophils, 41% lymphocytes, 1,890/ml monocytes, 2% eosinophils, and 1% basophils. Hemoglobin content was 8.9 g/dl and the platelet count was 52,000/ml.

Emergency spinal decompression surgery (en bloc spondylectomy) was performed with tumor debulking and L4–L5 laminectomy. The tumor removed during surgery was composed of multiple light green masses of soft tissue with some fragments of bone. No evidence of leukemia was found after examination of the peripheral blood or bone marrow smears. Microscopically, the tumor consisted of non-cohesive sheets of undifferentiated cells of uncertain lineage (Figure 3). The tumor cells were monotonous with blastic morphology and virtually lacking differentiation. Further immunohistochemical examination of the tumor showed blast cells positive for MPO, CD43, CD99, Fli-1, and weakly positive for CD68 (Figure 4). The tumor blast cells were negative for CD3, CD20, CD79a, and MyoD1. The CD43 is a very sensitive marker for Granulocyte Precursor Cells and fragments of bone with active marrow showing granulocytic hyperplasia. Therefore, this tumor was diagnosed as GS.

The patient was transferred to the Hematology Department and treated with DA therapeutic schedule (daunorubicin + cytarabine). Bone marrow morphology revealed bone marrow hyperplasia with 16% of myeloblasts, 51% of R2 inner cell, 13% of $CD13^+$, 41% of $HLDR\text{-}DR^+$, 60% of $CD33^+$, 15% of $CD71^+$, 14% of $CD34^+$. POX strain Bone marrow aspirate revealed 100% positive blast cells (Figure 2). Reexamination of her blood was following: leukocyte count of 5,600/ml, content of hemoglobin was 7.3 g/dl and the platelet count was 57,000/ml. Autoantibody test was following: antinuclear antibody: 1:320, and normal since. Shilling classification was following: 11% of myelo-

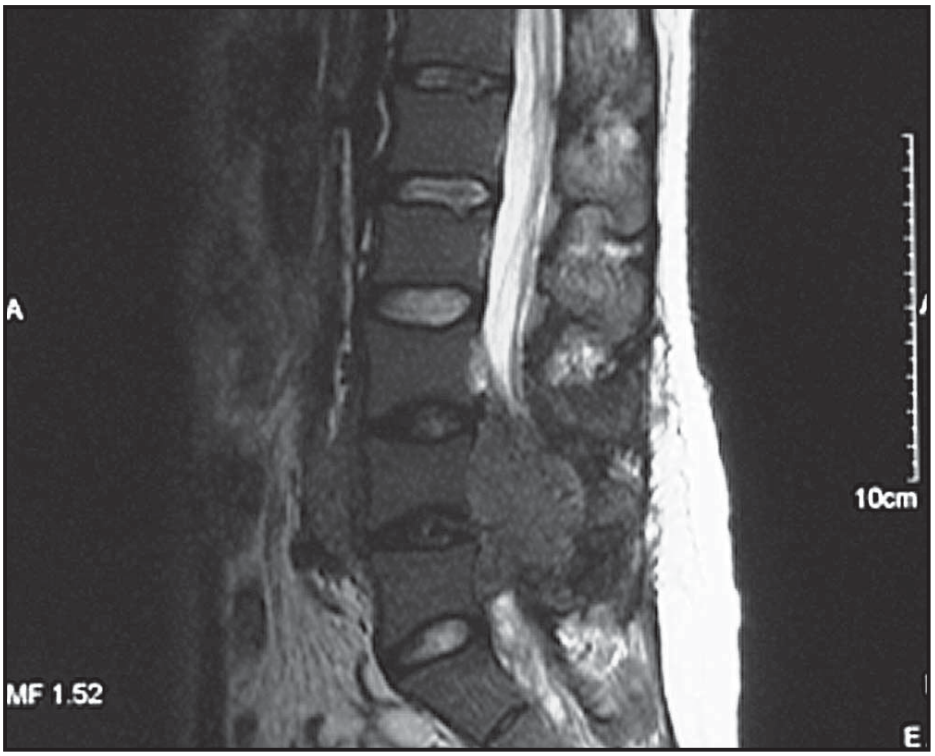

Fig. 1. Spinal MRI revealing soft tissue mass at L4–L5.

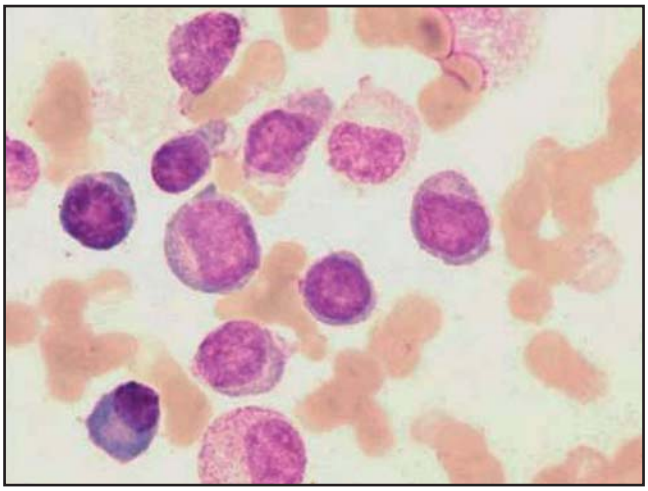

Fig. 2. Bone marrow aspirate performed at the time of initial diagnosis of GS. (Wright's stain, × 500)

blast, 1% of premyelocyte, 11% of myelocyte, 6% of metagranulocyte.

After two treatment courses, a remission was achieved. Bilateral lower extremity improved notably. Muscle power was: Grade 3~4 in left, Grade 1 in right. Reexamination of her blood revealed leukocyte count of 4,600/ml, content of hemoglobin of 9.8 g/dl and the platelet count was 331,000/ml.

DISCUSSION

In this case report, we presented an unusual Granulocytic sarcoma which initially manifested as paralysis of lower extremities. Spinal MRI of our patient revealed a L4–L5 paraspinal mass causing bilateral lower extremity weakness that quickly progressed to paraplegia.

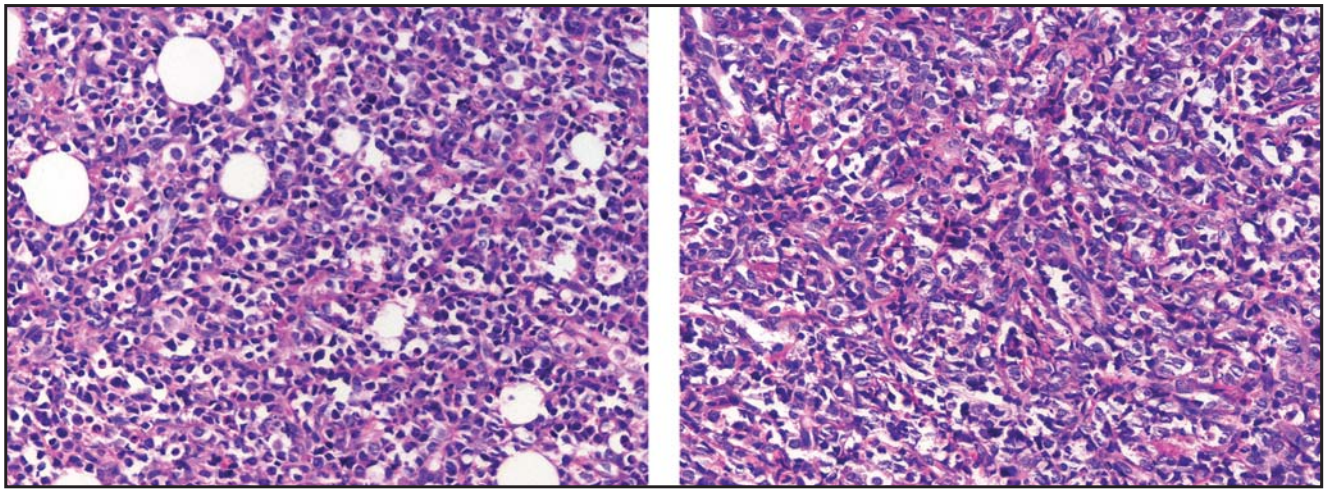

Fig. 3. Section of the tumor illustrating typical appearance of GS, containing non-cohesive sheets of undifferentiated cells of uncertain lineage (H&E stain, × 200 left, × 400 right)

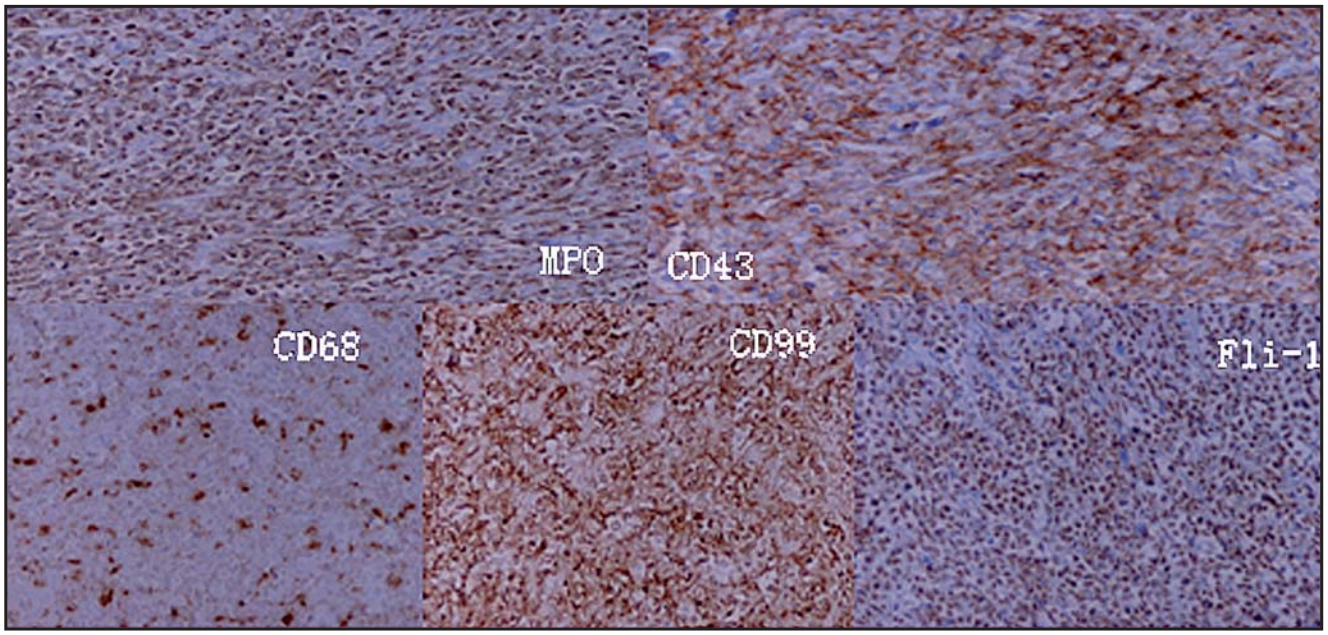

Fig. 4. Immunohistochemical examination of the mass (positive parts, × 200).

Emergency surgery to remove the mass and decompress the spinal cord was necessary. Her preoperative blood count revealed previously undiagnosed AML/CML in the chronic phase. Lumbocrural pain fast proceeded into paraplegia.

The optimal strategy for most extramedullary tumor is surgical excision. The extent of resection is determined by the anatomy of the lesion, the surgeon's experience, and the preliminary histological examination of frozen sections (Parsa *et al.* 2004). During the surgery, histhological diagnosis revealed minicell tumor. Due to minicell tumor sensitivity to chemotherapy, en bloc spondylectomy with chemotherapy after constituted the treatment of choice for this tumor.

Spinal cord compression secondary to GS is extremely rare. The main symptoms of patients are pain, numbness, or motor deficits. In a recent review of GS, the frequency of presenting symptoms in spinal cord compression was as follows: pain in 69%, motor deficits in 67%, and numbness or dysethesias in 27%. To date, we only found two reports of paraplegia due to just lumbar GS (Table 1). Our report is the third reported case of paraplegia due to GS of lumbar spine presented as paraplegia.

Granulocytic sarcoma (GS) should be considered in a list of differential diagnosis if patient experiences lumbocrural pain and paraplegia. When Granulocytic sarcoma located in spinal column was diagnosed, en bloc spondylectomy with chemotherapy postoperatively constituted the treatment of choice for this tumor.

Tab. 1. Paraplegia due to GS of lumbar at the time of initial tumor diagnosis.

References	Year reported	Age/ Gender	tumor location
Sauer (1914)	1914	36/M	Lumbar, Sacral
Mieremet (1914)	1914	15/M	L1-L3
Critchley & Greenfield (1930)	1930	11/M	T10-Lumbar
Laurie (1937)	1937	11/M	Lumbar
Griffin & Brindley (1942)	1942	26/F	T9-L1
Roger & Gastant (1947)	1947	13/M	T10-L2
Stransky & Campos (1955)	1951	14/M	Thoracic, Lumbar

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