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Granulocytic sarcoma with long spinal cord compression: A case report

Shao YD et al. GS with long spinal cord compression

Abstract

BACKGROUND

As an extramedullary form of proliferating myeloblasts, granulocytic sarcoma (GS) is

common in patients with acute myeloid leukemia (AML). GS in the central nervous

system is rare, and an intraspinal space-occupying lesion caused by GS is even rarer.

Surgical decompression is often necessary to remove the intraspinal space-occupying

lesion. To the best of our knowledge, we report, for the first time, a case of GS which

caused extensive compression in the spinal canal without surgical decompression

treatment.

CASE SUMMARY

A 15-year-old male suddenly developed numbness and weakness in his lower limbs for

10 d, which affected his walking ability. AML was later diagnosed in the Department of

Hematology. Magnetic resonance imaging (MRI) revealed that multiple segmental

space-occupying lesions were causing severe spinal cord compression in the thoracic

spinal canal. As a result, the patient received routine chemotherapy before surgery.

Interestingly, the intraspinal space-occupying lesions completely diminished on MRI

after a course of chemotherapy, and the sensation and strength in his lower limbs

markedly recovered.

CONCLUSION

An intraspinal space-occupying lesion could be the first symptom of AML, causing

spinal nerve compression without any other symptoms. Following standard

chemotherapy, spinal canal compression can be quickly relieved, and the spinal cord

and nerve function restored, avoiding emergency surgery.

Key Words: Granulocytic sarcoma; Acute myeloid leukemia; Spinal cord compression;

Intraspinal tumor; Chemotherapy; Case report

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Core Tip: This case is important as not all intraspinal granulocytic sarcoma in acute myeloid leukemia or chronic myeloid leukemia appear after systemic symptoms. We suggest that if conditions permit, relief of spinal cord compression can be achieved by standard chemotherapy. However, if hematological diseases are excluded, emergency surgery or biopsy is necessary to preserve the spinal cord and nerve function.

5 INTRODUCTION

Acute myeloid leukemia (AML) remains the most common acute leukemia in adults, including all non-lymphocytic acute leukemia^[1]. Approximately 2.5%-9.1% of AML patients suffer from granulocytic sarcoma (GS), also termed myeloid sarcoma, which is a solid tumor composed of mature or immature myeloid cells^[2]. The most frequent sites of GS are bone. GS of the brain and spinal cord is rare. An intraspinal space-occupying lesion caused by GS is even rarer. The treatment regimen for AML is induction chemotherapy to obtain complete remission, followed by consolidation and reinforcement treatment or stem cell transplantation^[3]. We here describe a patient with GS-associated spinal cord compression who was later diagnosed with AML during preoperative multi-disciplinary consultation by hematology, oncology, imaging, and neurosurgery experts prior to emergency surgery.

4 CASE PRESENTATION

Chief complaints

A 15-year-old male patient was admitted to hospital due to 10 d of lower back pain, and 4 d of numbness and fatigue of both lower limbs.

History of present illness

Ten days previously, the patient suffered from persistent chest and back pain without obvious inducement and no relief after rest. The degree of pain gradually worsened and was not relieved by oral analysics. Six days later, the patient experienced numbness and fatigue in both lower limbs which affected normal walking.

1 History of past illness

The patient had no relevant medical history.

Personal and family history

The patient had no significant family history.

Physical examination

The patient had abnormal sensation below the xiphoid process. Muscle strength in both lower limbs was weak, muscular tension in both lower limbs was increased, knee jerk reflex of both lower limbs was (+++), bilateral Achilles tendon reflex was (++++), bilateral Babinski sign was positive, and bilateral ankle clonus was positive.

Laboratory examinations

Preoperative routine blood tests showed abnormalities in white blood cells 23.43×10^9 /L, red blood cells 2.87×10^{12} /L, and platelets 101.00×10^9 /L. Tests for serum tumor markers, including alpha fetoprotein, carcinoembryonic antigen, carbohydrate antigen (CA) 19-9, CA 72-4 were all negative and ferritin level was significantly increased (> 2000 ng/mL) (Table 1).

Imaging examinations

Magnetic resonance imaging (MRI) showed a T3-8 spinal space-occupying lesion and marked spinal cord compression (Figure 1).

MULTIDISCIPLINARY EXPERT CONSULTATION

Doctors from the Department of Hematology suggested the possibility of hematological diseases and extramedullary hematopoiesis, and advised bone puncture and systemic treatment. Considering the long space-occupying segment, intraoperative bleeding and short medical history, the patient's family refused surgical treatment and agreed to transfer him to the Department of Hematology for bone puncture. The immunohistochemical results were as follows: CD34 small vessels (+), a few round nucleus cells (+), CD117 (+), CD61 megakaryocytes (+), monocytes (+) myeloperoxidase (MPO) (+), Lyso (+); CD3 was positive occasionally; CD20 was positive occasionally. The results of flow cytometry showed that CD117+ cells accounted for 24.61% of the total number of nuclear cells, and the immunophenotypes were CD13+, CD33+, CD34+, CD38+, CD56+, CD117+, HLA-DR+, MPO+, CD15-, CD64-, CD7-, CD11b-, CD19-, CD10-, CD16-, and the relative proportion of granulocytes was normal. The immunophenotypes of CD13, CD16, CD15, and CD11b were disordered and the expression of CD38 and CD56 was abnormal (Figures 2 and 3).

FINAL DIAGNOSIS

According to the above test results combined with clinical manifestations, the patient was diagnosed with AML-M2b.

TREATMENT

Systemic chemotherapy (IA scheme: Idarubicin + cytosine arabinoside) was administered. During treatment, the patient's neurological function in the lower limbs was closely observed, which gradually improved.

OUTCOME AND FOLLOW-UP

Lower limb muscle strength gradually returned to normal, chest and back pain disappeared, and the patient's body temperature was normal. One month after treatment and one year after treatment (Figure 4), an MRI scan showed that the intraspinal space-occupying lesions had completely disappeared.

DISCUSSION

As a form of myeloid leukemia, GS is a localized infiltration of abnormal white blood cells in subperiosteal or soft tissue^[4]. It is a localized solid tumor formed by immature granulocytes outside the bone marrow, which is very rare in clinical practice^[5]. The central nervous system, orbit, and bone marrow are common sites of GS^[2]. Preauricular and submandibular lymph nodes are often enlarged. There is also green pigmentation on the surface of the skin. In the late stage of the disease, all the essential organs and limbs are involved, and patients often die of anemia or infection^[8]. The incidence of GS in AML is 2.5%-9.1% [6], and is more common in male AML patients. The incidence of GS in chronic myeloid leukemia (CML) is lower compared with AML. In particular, GS in the central nervous system is rare, and spinal cord compression caused by GS is even rarer^[4]. Cord compression mainly occurs in the thoracic spinal canal. In previous literature reports^[7], most patients developed intraspinal compression symptoms after the diagnosis of AML or CML and were diagnosed as intraspinal space-occupying lesions by MRI examination^[8]. Most of these cases were treated with intraspinal decompression surgery, and the symptoms of intraspinal compression were significantly relieved[4]. AML patients often have solid tumors in other areas, especially intraspinal tumors, and these tumors often develop rapidly during the treatment of leukemia or when the disease is stable^[9]. In order to relieve pain and preserve spinal cord and nerve function, spinal surgeons often choose spinal canal decompression and tumor resection in an emergency situation. However, patients could suffer much more blood loss during surgery due to the rich blood supply and poor blood coagulation index.

CONCLUSION

Our case further confirmed that intraspinal GS can suddenly appear in previously healthy patients who had not previously been diagnosed with hematological diseases. Our patient had sudden onset of clinical symptoms of spinal cord compression. GS should be considered in patients with spinal canal space-occupying lesions and abnormal blood tests. Standard chemotherapy in combination with simultaneous monitoring of spinal cord function could be an alternative treatment regimen to surgery. The advantages and disadvantages of surgery in these patients require further study.

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