CASE REPORT

Acute myeloid leukemia presenting with complete paraplegia and bilateral total blindness due to central nervous system involvement

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We report a case of central nervous system (CNS) involvement in a 29-year-old man with acute myeloid leukemia (AML). Although leukemic cell invasion of the CNS in patients with AML has been reported in ~3% cases, multiple invasions of the CNS are rare. A 29-year-old man presented with rapidly progressive flaccid paralysis of the lower extremities. Laboratory findings showed blast cells, accounting for 79% of his white blood cell count. Thoracic spine magnetic resonance imaging showed a T2-hyperintense intramedullary lesion at the T6 level. Peroxidase staining was positive in cells isolated from a paravertebral mass as well as the bone marrow aspirate. The patient was diagnosed as having AML, with CNS involvement. We performed an emergency laminectomy between T6 and T9 with tumor resection. Following chemotherapy, although the patient achieved complete remission, the paraplegia did not improve. The patient showed signs of involvement of the cranial nerves and bilateral total blindness because of the involvement of visual areas in the cortex; these were considered to be caused by another lesion. Furthermore, heterotopic ossification resulted in left hip contractures. Despite the patient achieving complete remission of AML and improvement in overall prognosis, the patient's physical function remained limited.

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Acute myeloid leukemia (AML) with central nervous system (CNS) involvement has been described in ~3% patients.^{1,2} The prognosis of patients with this condition is poor, with a median overall survival of ~3 months.¹ Leukemic cell invasion may occur in any part of the CNS; however, invasion of the spinal cord and the cranial nerves and the medullary area controlling ocular muscles is extremely rare.^{1–4} We present the case of a 29-year-old man with CNS involvement, who presented with complete paraplegia and bilateral total blindness. Surgical treatment and chemotherapy were effective in achieving complete remission, but the patient's symptoms did not improve.

A 29-year-old man with difficulty in walking due to pain in both the lower extremities was transported to the emergency department from a local clinic. Physical examination revealed a loss of sensation to pinprick and vibration sensations from T10 to S5 levels, along with flaccid paralysis of the lower extremities and the absence of anal sphincter tone and sensation. Laboratory investigations revealed a white blood cell count of $4040 \mu\text{l}^{-1}$ (blast cells accounted for 79%) and his lactate dehydrogenase level 3292 U l^{-1} . Whole-body computed tomography (CT) showed bilateral pleural effusion. Thoracic spine magnetic resonance imaging (MRI) showed an intramedullary lesion at T6 and a paravertebral mass extending from T6 to T9 (Figures 1a and b). The blasts obtained by bone marrow aspiration were positive for peroxidase staining. Thus, AML with CNS involvement and a spinal cord injury scale A at T9 (The American Spinal Injury Association (ASIA) impairment scale) was diagnosed.

As the patient had progressive paralysis due to CNS invasion, surgical treatment for decompression of the spinal canal was planned before chemotherapy. We performed laminectomy

between T6 and T9, along with tumor resection. After surgery, the patient received daunorubicin hydrochloride (50 mg m^{-2} per day) for 5 days and cytarabine (200 mg m^{-2} per day) for 7 days for remission induction. During this therapy, the patient developed a central scotoma in the right visual field. A head CT showed a high-density area in the pituitary body and low-density masses in both orbital cavities (Figure 2a). Pleural fluid cytological examination confirmed disease progression; therefore, the patient received high-dose cytarabine therapy (8400 mg m^{-2} per day) for 5 days. At this point, the patient achieved complete remission of AML.

After a second course of consolidation therapy with high-dose cytarabine therapy (7800 mg m^{-2} per day) for 5 days, the complete remission was maintained, but there was no improvement in the paraplegia. At 2 months after the patient's admission, the patient lost his bilateral pupillary reflexes and developed bilateral total blindness. A head MRI and orbital contrast radiography MRI showed no disease progression (Figures 2b and c). Because the patient had developed a left hip flexion and abduction contractures caused by heterotopic ossification, he could not use a standard-type wheelchair because of the restricted hip joint range of motion. The patient was discharged from our hospital after 10 months of inpatient treatment, when his Functional Independence Measurement motor category scores totaled 18 (eating, 4; grooming, 3; and all other motor category scores, 1 each) and his cognitive category scores totaled 35. The patient required total assistance in activities of daily living. He has experienced no recurrence in the first year after discharge.

Figure 1. (a, b) Axial and sagittal T2-weighted thoracic spine MRI shows a high-intensity region in the spinal cord at T6 and a paravertebral mass from T6 to T9 on emergency admission. (c, d) Axial and sagittal T2-weighted thoracic spine MRI shows a high-intensity region in the spinal cord at T4 and T6 after the third course of chemotherapy.

To our knowledge, we report an extremely rare case of AML with CNS involvement, which presented with complete paraplegia and a progressive visual disturbance. Complete remission of AML was achieved with chemotherapy; however, the complete paraplegia and bilateral total blindness did not improve. Although the patient's overall prognosis improved, his physical function was increasingly restricted because of left hip contractures caused by heterotopic ossification (Figure 3) and a lack of improvement of the lower-extremity paralysis.

AML with CNS involvement has been described in ~3% patients. The major symptoms are back pain, lower back pain and neck pain associated with CNS involvement at a specific spinal cord level.² One case of facial nerve paralysis with bilateral extremity weakness has been reported previously; however, visual disturbance was not reported in that patient.³

We performed spinal canal decompression surgery and complete resection of the paravertebral mass, and the patient achieved complete remission with chemotherapy. However, the neurons in the spinal cord were irreversibly damaged because of CNS involvement (Figures 1a–d). The abnormal findings on the head MRI disappeared after chemotherapy courses, but the total blindness did not improve (Figures 2a–c). This lack of improvement suggested that the neurons in the medullary area that controls the ocular muscles and cranial nerves may have been damaged, as there were no abnormal MRI T2-weighted imaging findings in these areas that would suggest that blindness was a side effect of chemotherapy or the result of the so-called posterior reversible encephalopathy syndrome.

Heterotopic ossification commonly occurs in patients with paraplegia or tetraplegia caused by spinal cord injury. The cause of heterotopic ossification after spinal cord injury is unknown. In this case, the heterotopic ossification of the hip joint was diagnosed as myositis ossificans, which usually follows muscle injury.⁵ It is possible that the myositis ossificans was caused by trauma-related minor postural changes during the operative period. Although we carefully implemented a passive hip joint motion range training in order to prevent hip joint contracture, we were unable to prevent left hip joint contractures due to heterotopic ossification induced by myositis ossificans. Consequently, our patient cannot sit on a standard-type wheelchair. Our patient initiated rehabilitation in the activities of daily living, and in particular self-catheterization, in order to return home; however, the patient was not able to carry out self-catheterization because of hip contracture and bilateral total blindness. In the future, we should consider resecting the heterotopic ossification area or release hip contracture if it interferes with the transfer from bed to wheel chair or could lead to other restrictions in the activities of daily living or untreatable bedsores; especially, left greater trochanter pressure ulcer or left femoral fracture occurs because of progressive myositis ossificans.

AML with CNS involvement has been reported to have a poor survival prognosis. In our case, the patient initially presented with paraplegia. Although chemotherapy was effective for achieving AML remission, the complete paraplegia did not improve and the visual disturbance was progressive. In addition, the patient

Figure 2. (a) The head CT scan demonstrates a high-density area in the pituitary body and low-density masses in both orbital cavities on the eighth postoperative day. (b, c, d) Head MRI and orbital contrast MRI show no abnormal findings.

Figure 3. The hip joint X-ray image demonstrates the hip joint heterotopic ossification.

developed left hip contractures because of myositis ossificans. Despite improvements in the overall prognosis, the patient's physical function remained limited.

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COMPETING INTERESTS

The authors declare no conflict of interest.

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