Spontaneous spinal epidural hematoma as the initial presentation of leukemia

Hidetoshi Nojiri · Sungdo Kim · Takaaki Tsuji ·
Soichi Uta

Received: 10 September 2008 / Revised: 2 December 2008 / Accepted: 14 December 2008 / Published online: 6 January 2009
© Springer-Verlag 2008

Abstract We present a case of a 55-year-old male with progressive neurological deficits that appeared dramatically. MRI detected a spinal epidural hematoma at the cervicothoracic junction and blood tests showed leukocytosis, mild anemia, and thrombocytosis. Spontaneous spinal epidural hematoma (SSEH) as the initial presentation of leukemia was diagnosed. Urgent posterior decompression was performed after 28 h from acute onset of backache, and the patient experienced remarkable improvement in neurological findings.

Keywords Spontaneous spinal epidural hematoma · Chronic myeloid leukemia

Introduction

Spinal epidural hematoma is a potentially devastating problem that requires rapid diagnosis and surgical intervention. Two types of spinal epidural hematoma have been described in the literature: traumatic and nontraumatic [16]. Traumatic spinal epidural hematoma has been attributed to spinal surgery, epidural catheter placement, lumbar puncture, and chiropractic manipulation. In most

previous reports, nontraumatic spinal epidural hematoma has been termed spontaneous spinal epidural hematoma (SSEH). SSEH has been associated with anticoagulation and antiplatelet medication, pregnancy, and hemophilia, but few reports have described an association of SSEH with malignancy [2, 8]. In this report, we describe a case of cervicothoracic epidural hematoma that occurred as the initial presentation of leukemia.

Case report

A 55-year-old male presented to the emergency room complaining of an acute onset of backache. There was no history of trauma, disease, medication, or constitutional symptoms. Since the physical examination revealed no focal neurological deficits and radiographs of the spine revealed no abnormality, he was prescribed NSAIDs and went home on foot. Five hours later, he returned complaining of gait disturbance due to bilateral leg weakness. Within 10 h, he became progressively weak in both legs and a physical examination revealed complete paraplegia with a sensory level at T1. Within the same 10 h, bilateral arm weakness appeared, and he became unable to flex and extend his fingers (Frankel Grade B status).

On admission, magnetic resonance imaging (MRI) of the cervicothoracic spine detected the presence of a lesion compressing the spinal cord that extended from C5 to T3. The lesion was observed as an extradural space-occupying lesion with homogeneous isointensity to the spinal cord on T1-weighted images, and a mosaic of low- and high-intensity regions on T2-weighted images (Fig. 1). Computed tomography (CT) of the brain and cervical spine and MRI of the brain showed no abnormalities. In the

H. Nojiri (✉) · S. Kim · S. Uta
Department of Orthopaedics, Saisei Hospital,
800-1 Kashiwai-cho, Hanamigawa-ku,
Chiba 262-8506, Japan
e-mail: hnojiri@juntendo.ac.jp

T. Tsuji
Department of Orthopaedics,
Juntendo Tokyo Koto Geriatric Medical Center,
Juntendo University, Tokyo, Japan

Fig. 1 Preoperative MRI of the cervicothoracic spine detected the presence of a lesion compressing the spinal cord that extended from C5 to T3. The lesion was observed as an extradural space-occupying lesion with homogeneous isointensity to the spinal cord on T1-weighted images, and a mosaic of low- and high-intensity regions on T2-weighted images. The boundary between the spinal cord and the epidural hematoma was shown (arrowheads)

peripheral blood, leukocytosis (white blood cell count, $42.3 \times 10^9/L$) and mild anemia (hemoglobin 11.6 g/dL) were present, and the platelet count was increased to $596 \times 10^9/L$. D-dimer was elevated to 2.3 $\mu g/mg$. Coagulation function such as prothrombin time and activated partial thromboplastin time was not delayed.

The next day, the patient was transported to an associated hospital for emergency spinal surgery because neurological impairment was observed during the night, and urgent posterior decompression was performed under general anesthesia with intraoperative autotransfusion and red cell concentrates transfusion. After a C5–T5 left hemilaminectomy, all of posterior epidural hematoma extending from C5 to T4 could be evacuated easily because it was not so sticky. Prior to wound closure, a closed suction drain was placed deep into the paravertebral

muscle. The interval between the initial backache and decompression of the spinal cord was 28 h.

In a few days after surgery, the patient experienced remarkable improvement in neurological findings so that he could move his extremities against gravity (Frankel Grade C status). He could walk himself slowly with bilateral elbow crutches and a short leg brace at the last follow-up examination at 2 years after surgery (Frankel Grade D status). Urination was supported by clean intermittent catheterization. Neutrophil leukocytosis and basophilia were present, and the neutrophil alkaline phosphatase (NAP) score was low in the peripheral blood. Bone marrow aspiration for morphology and cytogenetic analysis was performed after surgery. Diagnosis was BCR-ABL gene positive chronic myeloid leukemia (CML). Beginning at 1 month after surgery, chemotherapy using imatinib, a small molecule tyrosine-kinase inhibitor, was administered effectively. Blood cell and platelet counts were within normal limits under continuous oral administration of imatinib (100 mg/day). Postoperative MRI one year later demonstrated a complete radiological resolution of the epidural hematoma and a complete decompression of the spinal cord (Fig. 2).

Fig. 2 Postoperative MRI demonstrated a complete radiological resolution of the epidural hematoma and a complete decompression of the spinal cord

Discussion

Spinal epidural hematoma is a well recognized, but rare event. The incidence has been estimated to be 0.1 patients per 100,000 patients per year [10], and fewer than 300 cases have been reported [8]. The etiology of SSEH has been examined, and possible causes include minor trauma, whooping cough, sneezing, voiding, vomiting, and lifting, all of which are assumed to produce an acute increase in vascular pressure [5]. In addition, pregnancy or bleeding diathesis has been mentioned as etiological factors [5]. The majority of SSEH may result from a rupture of the vascular network, and the rupture sites include epidural veins, arteries, cryptic angiomas, vascular malformations, hemangiomas and spinal angiomas. However, no statistical analyses or anatomical evidence has been presented. In addition, no relationship has been established between SSEH and arterial hypertension [7].

C5 to T2 comprises one third of the total number of involved segments reported. A second high rate of occurrence appears at T12. Patient age affects the vertebral level distribution of SSEH, with the hematoma in patients under the age of 40 years having an increased incidence around the cervicothoracic junction [7]. Reports of cases with coagulation disorders are increasing due to the worldwide increase in the use of oral anticoagulants for brain and/or circulatory disease. However, there have been no reports of spontaneous onset as an initial presentation of leukemia. Malignancy-associated spinal epidural hematoma was reported previously in two cases. One was a case of secondary onset in the blast crisis phase of CML that interrupted medication for 3 years [2]. The other case was ascribed to myelomatous vertebral lesions. The patient was administered prophylactic low molecular weight heparin therapy to prevent thrombophlebitis, but became worse neurologically and completely lost sensation and motor function. Tumor-related epidural inflammation, fragility of epidural venous plexuses caused by the tumor process and hemorrhagic pathologic microfracture of vertebral myeloma lesions have been suspected [8].

CML is a clonal hematopoietic disorder due to an acquired genetic defect in the pluripotential stem cell population. CML accounts for 6 to 15 percent of adult leukemia cases, and has an incidence of 1 to 1.5 per 100,000 per year [17]. CML usually behaves as a biphasic illness. The patient is diagnosed in a relatively indolent chronic phase that remains stable for a number of years until a more aggressive advanced phase intervenes. Ninety percent of CML patients, including our case, present in a chronic phase and the blastic stage of the advanced phase terminates with the patient's death [20]. Clinical features in CML patients are asymptomatic or generally common such as fatigue, fever, bone pain, or weight loss. Patients in

advanced phase are more likely to be symptomatic, with fever, bone pain, and/or bleeding being more prominent [20]. Bleeding and thrombosis are frequent, but relatively uncommon complications (20 and 6% of CML patients, respectively) compared to splenomegaly, which is present in more than three quarters of CML patients and is associated with severe organ damage and a high mortality [19]. The initial presentation of spontaneous mediastinal hematoma [12] or iliopsoas muscle hematoma [3] has been reported as rare cases in CML patients, but, in these cases, SSEH was not found.

Successful management of SSEH requires a rapid diagnosis. MRI is the most valuable tool in diagnosing the presence, location and extent of spinal hematoma. Inflammation [11], infection [4], tumor including epidural lymphoma [18], hemangioma [1], neurinoma, meningioma, as well as metastasis [15] are considered as differential diagnoses. MRI showed a hyperacute spinal hematoma, isointense to the spinal cord on T1- and hyperintense on T2-weighted images. An early subacute SSEH manifests with heterogeneous signal intensity and with areas of high signal intensity on T1- and T2-weighted images. A late subacute SSEH shows high signal intensity on T1- and T2-weighted images, while SSEH in the late chronic phase shows as hypointense on T1- and T2-weighted images [5]. Our MRI demonstrated that the patient was in the hyperacute-acute transitional phase because of the appearance of a mosaic of low- and high-intensity on T2-weighted images.

A relationship between surgical timing and neurological outcome was discussed previously. Immediate drainage is recommended by most surgeons, and surgical decompression within 24 h of complete sensorimotor deficit or within 48 h of incomplete sensorimotor deficit will achieve the optimum outcome [9]. Liao et al. [14] showed that good neurological recovery in patients with complete neurological deficits can be achieved when the interval between initial onset and surgery is less than 48 h and when the duration of the complete neurological deficits is less than 12 h. Lawton et al. [13] recommended immediate surgical evacuation within 12 h, but even patients with complete motor and sensory deficits or long-standing compression can improve with surgery. On the other hand, conservative (nonoperative) management of SSEH may be appropriate in those instances in which there is early and sustained neurological recovery confirmed by radiological resolution of the lesion [6]. In patients with mild or improving deficits, careful observation may be appropriate. However, when progressive neurological deterioration occurs, decompression surgery should be performed as soon as possible without hesitation. In the present case, the patient achieved neurological recovery in the Frankel Grade from B to D, but there were residual symptoms such as gait

difficulty and bladder symptoms. Therefore, we consider that the surgical treatment should have been performed earlier.

In conclusion, CML can be the cause of SSEH. The precise diagnosis and correct timing of decompression are essential for the management of SSEH, and can change the prognosis and quality of life of CML patients.

Conflict of interest statement None of the authors has any potential conflict of interest.

References

1. Aoyagi N, Kojima K, Kasai H (2003) Review of spinal epidural cavernous hemangioma. *Neurol Med Chir (Tokyo)* 43:471–475. doi:[10.2176/nmc.43.471](https://doi.org/10.2176/nmc.43.471) discussion 476
2. Banerjee R, Stanley J, Palumbo M (2004) Spinal epidural hematoma induced by leukemia. *Orthopedics* 27:864–866
3. Bauduceau O, De Revel T, Fogelman J, Amadou S, Souleau B, Nedellec G (2003) Spontaneous hematoma of the iliac psoas muscle in chronic myeloid leukemia. A case report. *Ann Med Interne (Paris)* 154:183–186
4. Boukobza M, Guichard JP, Boissonet M, George B, Reizine D, Gelbert F, Merland JJ (1994) Spinal epidural haematoma: report of 11 cases and review of the literature. *Neuroradiology* 36:456–459. doi:[10.1007/BF00593683](https://doi.org/10.1007/BF00593683)
5. Braun P, Kazmi K, Nogues-Melendez P, Mas-Estelles F, Aparici-Robles F (2007) MRI findings in spinal subdural and epidural hematomas. *Eur J Radiol* 64:119–125. doi:[10.1016/j.ejrad.2007.02.014](https://doi.org/10.1016/j.ejrad.2007.02.014)
6. Clarke DB, Bertrand G, Tampieri D (1992) Spontaneous spinal epidural hematoma causing paraplegia: resolution and recovery without surgical decompression. *Neurosurgery* 30:108–111. doi:[10.1097/00006123-199201000-00019](https://doi.org/10.1097/00006123-199201000-00019)
7. Groen RJ, Ponssen H (1990) The spontaneous spinal epidural hematoma. A study of the etiology. *J Neurol Sci* 98:121–138. doi:[10.1016/0022-510X\(90\)90253-J](https://doi.org/10.1016/0022-510X(90)90253-J)
8. Hayem G, Deutsch E, Roux S, Palazzo E, Grossin M, Meyer O (1998) Spontaneous spinal epidural hematoma with spinal cord compression complicating plasma cell myeloma. A case report. *Spine* 23:2432–2435. doi:[10.1097/00007632-199811150-00015](https://doi.org/10.1097/00007632-199811150-00015)
9. Hejazi N, Thaper PY, Hassler W (1998) Nine cases of nontraumatic spinal epidural hematoma. *Neurol Med Chir (Tokyo)* 38:718–723. doi:[10.2176/nmc.38.718](https://doi.org/10.2176/nmc.38.718) discussion 723–4
10. Holtas S, Heiling M, Lonntoft M (1996) Spontaneous spinal epidural hematoma: findings at MR imaging and clinical correlation. *Radiology* 199:409–413
11. Kuker W, Schaade L, Ritter K, Nacimiento W (1999) MRI follow-up of herpes simplex virus (type 1) radiculomyelitis. *Neurology* 52:1102–1103
12. Kuo YC, Wang HC, Chu KA, Lu JY (2000) Chronic myeloid leukemia initially presenting with spontaneous mediastinal hematoma and hemothorax. *Zhonghua Yi Xue Za Zhi (Taipei)* 63:170–174
13. Lawton MT, Porter RW, Heiserman JE, Jacobowitz R, Sonntag VK, Dickman CA (1995) Surgical management of spinal epidural hematoma: relationship between surgical timing and neurological outcome. *J Neurosurg* 83:1–7
14. Liao CC, Lee ST, Hsu WC, Chen LR, Lui TN, Lee SC (2004) Experience in the surgical management of spontaneous spinal epidural hematoma. *J Neurosurg* 100:38–45
15. Matsumura A, Namikawa T, Hashimoto R, Okamoto T, Yanagida I, Hoshi M, Noguchi K, Takami M (2008) Clinical management for spontaneous spinal epidural hematoma: diagnosis and treatment. *Spine J* 8:534–537. doi:[10.1016/j.spinee.2007.01.009](https://doi.org/10.1016/j.spinee.2007.01.009)
16. Mattle H, Sieb JP, Rohner M, Mumenthaler M (1987) Nontraumatic spinal epidural and subdural hematomas. *Neurology* 37:1351–1356
17. Morrison VA (1994) Chronic leukemias. *CA Cancer J Clin* 44:353–377. doi:[10.3322/canjclin.44.6.353](https://doi.org/10.3322/canjclin.44.6.353)
18. Singer MB, Atlas SW, Drayer BP (1998) Subarachnoid space disease: diagnosis with fluid-attenuated inversion-recovery MR imaging and comparison with gadolinium-enhanced spin-echo MR imaging—blinded reader study. *Radiology* 208:417–422
19. Wehmeier A, Daum I, Jamin H, Schneider W (1991) Incidence and clinical risk factors for bleeding and thrombotic complications in myeloproliferative disorders. A retrospective analysis of 260 patients. *Ann Hematol* 63:101–106. doi:[10.1007/BF01707281](https://doi.org/10.1007/BF01707281)
20. Yong A, Goldman JM (2000) Chronic myeloid leukemia, 2nd edn. Churchill Livingstone, New York