

Case report

Thunderclap headache as a presentation of spontaneous spinal epidural hematoma with spontaneous recovery

Pornchai Sathirapanya, Suwana Setthawatcharawanich, Kitti Limapichat, Kanitpong Phabphal

Department of Internal Medicine, Faculty of Medicine, Prince of Songkla University, Hat Yai, Songkhla, Thailand

Background: Spontaneous spinal epidural hematoma (SSEH) is a rare spinal cord compression disorder. Thunderclap headache mimicking subarachnoid hemorrhage as an initial manifestation of this disorder has been scarcely reported. Moreover, the neurological outcome of conservative treatment is satisfactory only in some clinical settings. The unusual presentation and mechanisms for spontaneous recovery of SSEH are discussed.

Objective: To report a case of SSEH that presented with acute severe headache that mimicked subarachnoid hemorrhage. After a period of neurological deficit, spontaneous improvement was seen, which progressed to full recovery without neurosurgical intervention.

Findings: A 62-year-old man presented with acute occipital headache and neck stiffness for which subarachnoid hemorrhage was initially suspected. An emergency computed tomographic brain scan and cerebrospinal fluid analysis excluded subarachnoid hemorrhage. Later, he developed acute paraparesis, hypoesthesia up to the fourth thoracic dermatomal level (T4) as well as bowel and bladder dysfunction. The magnetic resonance imaging eventually revealed spinal epidural hematoma, located anterior to C7 through T4 spinal level. He had no previous history of spinal injury, systemic, and hematological disorders. The neurological recovery began 20 hours after the onset and continued until complete recovery, 4 months after the onset without neurosurgical intervention.

Conclusion: SSEH could sometimes imitate subarachnoid hemorrhages when it located in the cervical region. Spontaneous recovery of SSEH without surgical intervention might be possible in cases with continuing neurological recovery, although the recovery began much later in the clinical course.

Keywords: Headache, Spontaneous spinal epidural hematoma, Subarachnoid hemorrhage, Paraparesis

Introduction

Spontaneous spinal epidural hematoma (SSEH) is an uncommon cause of spinal cord compression. The annual reported cases were estimated at two to six cases.¹ However, more usage of magnetic resonance imaging (MRI) for diagnosis of spinal disorder has made it more detectable in present days.¹ SSEH usually presents with acute spinal pain associated with acute or progressive spinal cord compression syndrome similar to the other causes of spinal cord compression. Therefore, a spinal MRI is necessary to differentiate

the etiology. The actual pathogenesis of SSEH has not been clearly understood. Thunderclap headache with stiffness of neck, mimicking subarachnoid hemorrhages are very unusual presentations of SSEH. Although emergency surgical spinal decompression is now the indicated treatment of SSEH, satisfactory response to conservative measures has been reported as well only in a few cases with neurological predictors for favorable outcomes.

Case report

A 62-year-old man with well-controlled mild essential hypertension had an abrupt onset of a severe occipital headache, posterior neck pain, and near syncope while he was watering his garden. He was immediately

Correspondence to: Pornchai Sathirapanya, Division of Neurology, Department of Internal Medicine, Faculty of Medicine, Prince of Songkla University, Hat Yai, Songkhla 90110, Thailand.
Email: sporncha@medicine.psu.ac.th

examined at the emergency department of a local hospital, where only stiffness of neck was found during neurological examination. A subarachnoid hemorrhage was the provisional diagnosis. Emergency computed tomographic scan of the brain (CT brain) was unremarkable. Lumbar puncture, for cerebrospinal fluid (CSF) analysis to exclude subarachnoid hemorrhage, was performed. The CSF analysis revealed clear CSF, open pressure of 10 cm H₂O, and no white or red blood cells were found. CSF protein was 43 mg/dL and CSF sugar was 68 mg/dL. No CSF xanthochromia was present. He was admitted for a cerebral angiography. Two hours after the onset, he experienced rapidly progressive paraparesis and loss of pain and temperature sensations below his chest. Bowel and urinary bladder dysfunction developed simultaneously. At this moment, he still had intolerably severe cranial and neck pain for which multiple doses of parenteral potent analgesics were needed. Repeated neurological examinations revealed flaccid paraparesis of grade 1/5 with areflexia on both lower limbs. Loss of pain, temperature, light touch, and vibratory sensations were detected up to the fourth thoracic dermatomal level (T4). Weakness of anal sphincter contraction, loss of anal sensation, and absence of bulbocavernosus reflex were associated as well. Owing to a delay in the patient transfer process, an emergency MRI of spinal cord was performed in our center 6 hours after the onset of paraparesis. The MRI revealed a spinal epidural hematoma located in the anterior spinal epidural space, extending from the C7 to T4 spinal vertebral level, with intrinsic spinal cord T2 hyperintensity down to the T8 level (Figs. 1 and 2). He denied the history of previous spinal injuries and bleeding disorders. The complete hematological assays including coagulogram were unremarkable. A subsequent spinal angiography showed a small arteriovenous fistula (AVF) at the lumbar spinal level, which was considered as an uncorrelated abnormality. While waiting for the patient's consent for immediate surgical decompression

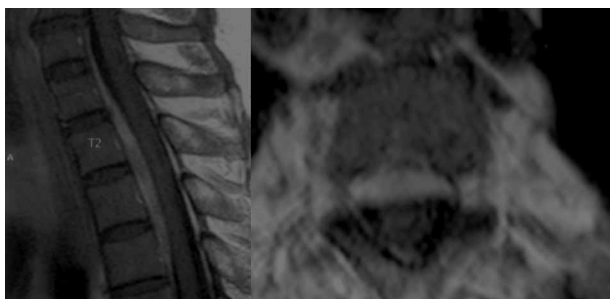


Figure 1 Sagittal and axial T1W MRI of cervico-thoracic spinal cord show a spinal epidural hematoma at anterior epidural space, extending from C7–T4 with spinal cord compression.

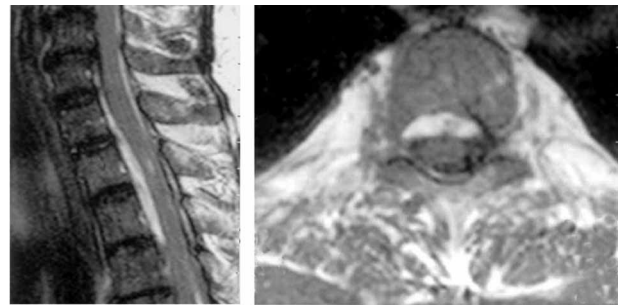


Figure 2 Corresponding sagittal and axial T2W MRI of cervico-thoracic spinal cord show a spinal epidural hematoma at anterior epidural space and an intra-medullary spinal cord hyperintensity, which is suggestive of myelopathy.

after the preoperative evaluations were completed, the neurological deficits unexpectedly started to improve spontaneously 20 hours after the onset. The patient could move his feet, and his headache had disappeared. Finally, the patient decided to undergo non-operative treatment, under closed neurological monitoring by the authors and a consultant neurosurgeon. Two weeks after the onset, only urinary dysfunction and weakness of anal muscle contraction remained unchanged, the motor power of the lower extremities had recovered to grade 3–4/5 and he was discharged. He achieved complete neurological recovery 4 months after discharge with intensive physical rehabilitation, but without surgical intervention.

Discussion

A constellation of symptoms with abrupt and intractable localized spinal pain, immediately followed by rapidly progressive paraplegia, and bowel and urinary dysfunction are highly suspected of acute spinal cord compression. Ruptured spinal epidural abscess, ruptured spinal arteriovenous malformation, and complicated spinal metastatic neoplasm were common etiologies of such a rapidly progressive compression myelopathy in clinical experience. Rarely, SSEH was found as the etiology. The more rapidity of neurological deterioration within a few hours might differentiate SSEH from other spinal cord compressions such as spinal metastasis, meningioma, neurofibroma, and other benign spinal cord compression causes. The hematomas of SSEH are commonly located in the posterior part of cervical and thoracic spinal canal followed by the cervico-thoracic spinal canal but less frequently found in the lumbo-sacral spinal region.^{2,3} Two cadaveric studies of the morphology of internal spinal venous plexus, one in the fetuses and the other in the aged humans, showed that the posterior internal spinal venous plexus was more prominent from C5 to T2 in

both of them. These findings correspond to the common site of SSEH found.^{4,5} In a rare case of SSEH, when the hematoma involved the cervical spinal level (as in this reported case), the presenting symptoms were unusual for spinal cord compression. Bleeding into the epidural space, which is a potential space, caused a space-occupying lesion (hematoma) within the spinal canal and suddenly raised intraspinal canal pressure. This resulted in compression and displacement of cervical spinal meninges by the hematoma; with subsequent stretching and tension on cervico-cranial meninges, meningeal irritation signs ensued. Therefore, it was potentially a mimicry of meningeal signs found in a subarachnoid hemorrhage. Unlike a previous reported case by Labeodan⁶ in which acute headache, stiffness of neck associated with obviously severe thoraco-lumbar pain occurred together during bending and lifting weights, and SSEH was found by MRI at the T5–T7 spinal level. The exertional activity with Valsalva's effect combined with the reduction of intraspinal space during bending might be the co-contributing factors to raise the spinal venous plexus pressure and finally spinal venous plexus rupture in such a case.⁷ This patient showed only a severe headache and neck stiffness during a neutral body position and during non-exertional activity as well. Initially, subarachnoid hemorrhage was highly suspected based on the presenting symptoms until both the CT brain and CSF analysis were unremarkable. The emerging of acute paraparesis combined with bowel and bladder malfunction subsequently suggested a spinal cord disorder.

The etiologies of spinal epidural hematoma varied from idiopathic to various identifiable causes such as spinal trauma, coagulopathy, pregnancy and child delivery, and spinal invasive procedures.⁸ Spontaneous rupture of Batson's venous plexus was attributed in idiopathic or spontaneous cases.⁹ There was no identifiable causes or notable precipitating factors of spinal epidural hematoma in the presented case. According to the findings of spinal angiography, the small spinal AVF was found at the lumbar level, and there was no draining veins bridged between it and the site of spinal epidural hematoma demonstrated. Arterial hypertension and a small AVF in the lumbar region were considered as unrelated conditions to SSEH in previous reports.^{9,10} Spinal AVFs are mostly solitary lesions that present at thoracolumbar region. In addition, they rarely cause spinal hematoma, and chronic progressive myelopathy is a common presentation.¹¹ Therefore, SSEH was the final diagnosis in this case. The emergency MRI of the spinal cord was an investigation of choice to confirm and differentiate the diagnosis of spinal cord

compression. In some occasions, a follow-up MRI study with T1W, T2W, gradient echo, and fat suppression sequences were necessary for a definite diagnosis, since the results of an early spinal MRI in the hyperacute and acute stage of SSEH may be non-differentiable.¹²

Most of the clinical studies supported emergency surgical decompression in order to achieve a satisfactory neurological outcome.^{13–16} Factors determining a favorable outcome were less severe preoperative neurological deficits and shorter interval time from the onset of neurological dysfunction to surgery.^{13,17} Neurological recovery from conservative treatment of SSEH was reported in some cases, in which neurological improvement started early after the onset and when an initial neurological deficit was minor.¹⁸ Recently, the previously undiagnosed and mild cases of SSEH have been diagnosed due to the broadly available MRI in diagnosis of spinal cord disorders. Early detection of such mild SSEH cases will facilitate the use of non-operative treatment when there is no further neurological deterioration. A review study comparing operative to non-operative treatment of SSEH cases (474 vs. 62 cases) showed that cases with successful non-operative treatments had a significant longer length of hematoma on the MRIs.¹⁹ Early spontaneous regression and subsequent resolution of the hematomas was possible, because the hematoma was considered a venous blood mass. Furthermore, the longer length of spinal epidural hematoma, in correlation with the cylindrical shape of the spinal canal, would exert less pressure effect on the spinal cord and eventually had less severe neurological disorders. These may be the reasons for spontaneous recovery of SSEH. The current reported case had continuing neurological improvement starting 20 hours after the onset of neurological deterioration. Therefore, the patient decided to receive a conservative treatment and complete neurological recovery occurred.

Conclusion

The authors reported a rare cause of spinal cord compression with unusual initial presentations imitating subarachnoid hemorrhage, and with spontaneous neurological recovery without surgery. Although the neurological deficits were severe initially and the improvement started late, the patient unexpectedly had a continuing spontaneous and complete recovery of neurological impairments without surgery.

Acknowledgement

The authors thank Prof. Dr Siriporn Hirunpat, Dr Kittipong Riabroi, Dr Nuttha Sanghan, and Sakrai

Thongchuea, Department of Radiology, Faculty of Medicine, Prince of Songkla University for their assistance in reviewing the MRIs and formatting the image files. The authors also thank David Brown and David Leather, International Affairs Unit, Faculty of Medicine, Prince of Songkla University for editing the manuscript.

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