Spontaneous Spinal Epidural Hematoma: A Case Report

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Abstract
Spontaneous spinal epidural hematoma (SSEH) manifests from blood accumulating in the epidural space, compressing the spinal cord and leading to acute neurological deficits. MRI provides the most valuable visualization of the location and hematoma mass as well as the presence of the spinal cord compression. SSEH can occur in any segments of the spinal cord but predominantly at the posterior cervicothoracic (C5–T2) and thoracolumbar (T10–L2) levels. The source of hemorrhage SSEH can be both vertebral venous plexus system or arterial source. Decompressive laminectomy and hematoma evacuation are the standard surgical procedures upon diagnosis of SSEH, although spontaneous recoveries have been reported. The degree of preoperative neural deficit is a major prognostic factor. Conservative management has proven effective, although feasible only if spontaneous recovery is manifested. Decompressive laminectomy should continue to remain readily available, given the inverse correlation between operative interval and recovery.
Introduction

Spontaneous spinal epidural hematoma (SSEH) is a relatively rare pathology characterized by blood accumulating in the epidural space and compressing the spinal cord. SSEH was first described by Jackson in 1869 [1]. The incidence of SSEH is considered to be 0.1 per 100,000 per year [2]. Symptoms may vary clinically from radicular symptoms to quadriplegia depending on the extent of cord compression [3]. The exact etiology of SSEH remains unknown in 40–50% of the patients, although vascular malformations, coagulopathy, epidural catheter and surgical trauma, anticoagulants, and the conditions causing increased intrathoracic-intra-abdominal pressure such as cough and Valsalva maneuver have been reported as the predisposing factors for SSEH [4, 5]. Both the venous and arterial theories provide plausible explanations for SSEH with most cases probably resulting from a venous bleed [6, 7]. Magnetic resonance imaging (MRI) has emerged as the diagnostic study of choice for SSEH [8]. SSEH patients with progressive neurological deficits require urgent decompression of the spinal cord and the evacuation of the hematoma [2]. On the other hand, there are several studies suggesting that patients with no neurological deficits or patients showing spontaneous recovery can be conservatively followed up [9]. The degree of pretherapy neural deficit is a major prognostic factor [10]. An early and individualized rehabilitation program after recovery of the operation is beneficial for SSEH patients [11].

In this report, we present a previously healthy young woman who was admitted to the emergency department with weakness of bilateral lower extremities caused by SSEH, in whom prompt surgical treatment and immediate rehabilitation proposal prevented definitive neurological sequelae.

Case Report

A previously healthy 31-year-old female patient presented to our emergency department with weakness in bilateral lower extremities after waking up in the morning. The patient deteriorated with an acute onset of flaccid quadriplegia, graded as an American Spinal Injury Association (ASIA) score of A. There was no recent medical history of trauma, infection, surgery, or anticoagulant use. The initial laboratory tests of blood, such as blood routine test, liver function test, blood creatinine, electrolytes, activated partial thromboplastin time, and international normalized ratio, were within normal limits. The blood pressure was normal. The neurological examinations revealed loss of strength of bilateral lower extremities and the muscle strength was grade 0/5. The loss of skin sensation was below the T4 vertebral level with saddle anesthesia. Babinski reflex of bilateral lower extremities was positive and bilateral tendon reflex was were reduced.

MRI of the cervical and dorsal spines revealed a posterior epidural hematoma from the T2 to T3 level with spinal cord compression. The mass had isointensity to the spinal cord on T1-weighted images and hyperintensity on T2-weighted images (Fig. 1).

The patient was hospitalized in the orthopedic department after diagnosis of SSEH and an emergency posterior approach decompression bilateral laminectomy and hematoma evacuation operation was performed from T1 to T3. Thick, clotted blood and prominent epidural hematoma were noted in the epidural space, but no obvious vascular malformation was found during operation (Fig. 2). The hematoma was clearly evacuated and the cord was adequately decompressed. The pathological report confirmed no evidence of vascular malformation or neoplasm. After the healing of the incision, the patient was transferred to the department of
rehabilitation medicine. A detailed rehabilitation schedule was established for the early rehabilitation program, which included passive joint range of motion training of lower limbs, active joint range of motion training of upper limbs, muscle strength enhance training, pulmonary function training, and position change training. After gaining enough upper limb strength, the next stage of training began, which included seat balancing training, tilting table training, transfer training (bed to wheelchair), and activities of daily living training. The patient finally could handle basic living activities, complete wheelchair locomotion, and transfer from bed to wheelchair independently.

Postoperative MRI showed complete resolution of the hematoma (Fig. 3). The patient recovered well with prompt return of neurological function. On one-year follow up, she was doing well with no residual neurological deficits.

Discussion

SSEH accounts for less than 1% of all spine epidural lesions, with an estimated annual incidence of only 1 per million [2]. Classic SSEH can present with features ranging from simple back or neck pain to complete paraplegia or quadriplegia, depending on the site and severity of spinal cord compression [3]. However, the initial presentations of SSEH are often miscellaneous and atypical. The initial symptoms can occasionally mimic disc prolapse and even rarely be misdiagnosed as transient ischemic attack or stroke [12, 13].

SSEH can be difficult to establish prior to the appearance of neurological deficits. Therefore, the differential diagnosis of SSEH should include pulmonary emboli, spontaneous pneumothorax, and acute myocardial infarction in the absence of neurological deficit and should include transverse myelitis, Guillain-Barré syndrome (GBS), epidural subarachnoid bleeding, and acute spinal cord ischemia in the presence of a neurological deficit [14, 15].

The ASIA developed a set of standards that assess the degree and level of spinal cord injury to an individual, on a scale of A – for most severe – to E – for normal. A hallmark indicator of neural function that has long been used in longitudinal studies, the ASIA score serves as an accurate marker for SSEH patients’ preoperative functioning and subsequent prognosis [16]. Therefore, a score should be immediately collected upon presentation.

Upon suspicion of SSEH, the imaging modality of choice to confirm is MRI. MRI provides the most valuable visualization of the location and hematoma mass as well as the presence of the spinal cord compression. The early MRI recognition provides the opportunity for early appropriate treatment and therefore leads to better neurological recovery. When compared to the spinal cord within 24 h from symptom onset, the hematoma typically appears isointense on T1-weighted and hyperintense on T2-weighted MRI. The hematoma often appears hyperintense on both T1- and T2-weighted images 24 h later. Chronic hematomas may present hypointense on both T1- and T2-weighted images. Fat suppression images may be used to distinguish hematoma from epidural fat [8]. Sometimes active bleeding into the hematoma will reveal a central area of enhancement when contrast is used. A computed tomography scan should be obtained if MRI is unavailable.

SSEH can occur in any segments of the spinal cord but predominantly at the posterior cervicothoracic (C5–T2) and thoracolumbar (T10–L2) levels. Some authors noted an association between hematoma location and outcome. A plausible explanation might be that the thoracic spinal canal is narrower than the cervical and lumbar spinal canals, making the thoracic spinal cord more susceptible to hematoma compression-induced ischemia [17]. Some authors
believe that the spine segments which have more mobility produce more tension on the epidural veins, which may explain why SSEH commonly occur either at the cervicothoracic or thoracolumbar regions [18].

The dorsal epidural space is more often affected by SSEH than the ventral space since the dural sac is firmly attached to the posterior longitudinal ligament as well as the fact that the dorsal epidural plexus is larger than the ventral epidural plexus. In addition, there may also be an area of “locus minoris resistentiae” that is more susceptible to rupture with minor changes in intravenous pressure [19]. Anterior SSEH has also been reported, though very rarely. Similar to the literature, the source of bleeding in our patient was posterior.

The etiology of SSEH or more specifically the source of hemorrhage has been debated for decades. The considerable evidence points to the venous system as the most likely source of hemorrhage. The vertebral venous plexus, a low-pressure valveless system, is in continuity with the abdominal and thoracic venous system. Lack of valves in the epidural venous plexus makes the area especially vulnerable to any intrinsic change in pressure. The bleeding can occur after daily activities such as voiding, sneezing, bending, coughing, and coitus. Such activities of normal living can cause fluctuations in the intrathoracic and intra-abdominal pressures, which may cause blood flow reversal resulting in rupture of a delicate vein in the valveless epidural plexus [6].

However, the hypothesis of an arterial source of bleeding has also been well supported. Given that hematomas usually form quickly and given the rapid onset of SSEH, greater arterial flow is indicative of an arterial source, as the pressure of the epidural venous plexus is lower than the intrathecal pressure. Furthermore, although this arterial versus venous distinction may be of great academic interest, the source of bleeding is not a major prognostic factor once the epidural space has been infiltrated with blood [7]. Therefore, some authors raised an underlying and overlooked question: why must one source of bleeding preclude another? Perhaps an SSEH patient is just as likely to bleed from an artery as they are from a vein. Furthermore, venous bleeding may be more insidious due to less profusely bleeding and slow symptom development than arterial bleeding. This might be a major cause of primary misdiagnosis delaying operative care.

Decompressive laminectomy and hematoma evacuation are the standard surgical procedures upon diagnosis of SSEH. Urgent decompression of the spinal cord especially in the high-risk SSEH patients with severe neurological deficit should be the initial step for the neurological deficit recovery and the hematoma classification [2]. However, there are several studies suggesting that low-risk cases with no neurological deficits or patients showing spontaneous resorption or spontaneous recovery can be conservatively followed up by dynamic MRI [9]. In the current case, surgery was performed due to the presence of severe neurological symptoms on initial presentation.

Two salient factors have been shown to have major prognostic implications: the degree of preoperative neural deficit and the time interval between ictus and surgery [20]. An emphasis has been placed on establishing an acceptable surgical time frame between ictus and intervention. Prompt decompression of the spinal cord is of prime importance because prolonged compression of the spinal cord may lead to irreversible injury of the spinal cord. Yet, some studies have failed to prove a statistically significant difference in outcomes based solely on the time as it is highly difficult to define the most favorable time for the application of decompression surgery. Given that most of the available case reports are based on small numbers, many have suggested that for optimal neurological improvement, patients should undergo surgical decompression within 12–48 h of symptom onset [20, 21].
More importantly for long-term outcomes is the preoperative neurological status of the patient which is the major prognostic indicator [10]. Patients with minimal symptoms prior to therapy are more likely to acquire complete recovery than those with major deficits in sensorimotor functioning. A hallmark indicator of pretherapy neural deficits is the ASIA score [16]. The ASIA score should be immediately collected upon presentation of SSEH-like symptoms, as it aids in developing a prognosis and determining the time and type of intervention needed. Therefore, one could argue that SSEH can, when causing a large preoperative neural deficit of ASIA score A or B, require priority emergency care. In contrast, when the neural deficit is of ASIA score C or D, urgent care is recommended but time is not as strong a factor.

**Conclusion**

SSEH have extremely low incidence rates in the adult population. Immediate diagnosis and surgical intervention is typically recommended as it can cause catastrophic consequences. However, clinical manifestations of the SSEH are nonspecific. Therefore, it should be kept in mind especially in patients presenting with neurological deficits and a sudden onset of neck or back pain. MRI is still the gold standard for the differential diagnosis of patients suspected to have SSEH. Neurogenic claudication may occur secondary to the compression of the spinal cord with no invasion into the neural tissue. Therefore, surgical intervention is the ideal method for achieving complete recovery. However, spontaneous resolution of SSEH can also occur in the absence of neurological deficits, demonstrating that surgical intervention is not always indicated for these patients. As in our case, multilevel spinal cord compression secondary to hematoma after prompt surgical intervention can benefit from inpatient rehabilitation that specializes in spinal cord injury. This may lead to significant improvement in functional outcomes.

**Statement of Ethics**

The authors have no ethical conflicts to disclose.

**Disclosure Statement**

The authors have no conflicts of interest to declare.

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**Author Contributions**

Bin Zuo collected the data and wrote the paper, YueHui Zhang, Jing Zhang, and Jia Song reviewed the literature, and Shao Jiang and XiaoLing Zhang checked the manuscript.
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Fig. 1. Preoperative thoracic sagittal T1 and T2 MRI as well as axial T2 MRI.

Fig. 2. Thoracic spinal epidural hematoma images during surgery.
Fig. 3. Postoperative thoracic axial T2 MRI fields marked hemilaminectomy.