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Spontaneous Spinal Epidural Hematoma: A Case Report and Literature Review

Yangliang Huang*, Houqing Long
Department of Spine Surgery, the First Affiliated Hospital, Sun Yat-Sen University, Guangzhou, 510700, China

***Corresponding author**
Yangliang Huang

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Abstract: Spontaneous spinal epidural hematoma (SSEH) is a rare neurologic emergency of the spinal cord, which is caused by a hemorrhage in the epidural space. Patients usually experience sudden severe neck or back pain accompanied by muscle weakness and bladder dysfunction. The mean age of SSEH patients is about 30 in Chinese population. Thus, early recognize of these patients and urgent surgical decompression is critical for a satisfied outcome. The risk factors for poor clinical outcome included a short progression interval, no symptom relief after admission, hypesthesia, complete spinal cord injury, and hematoma below the T4 level.
Keywords: Spontaneous spinal epidural hematoma; arteriovenous fistula; surgical treatment.

INTRODUCTION

Spontaneous spinal epidural hematoma (SSEH) is a rare neurosurgical condition with potentially devastating consequences. It is defined as a spontaneous hemorrhage in the spinal epidural space without overt traumatic or iatrogenic causes [1]. The incidence of SSEH is around 1/ 100,000[2]. The symptoms may be similar with cervical myelopathy or cerebral infarction, which makes the clinical diagnosis not always obvious.

The source of bleeding is believed to be the posterior internal vertebral venous plexus or arteriovenous malformation, a structure that is more prominent in adults than in children [3, 4] .The most common locations for SSEH are in the middle to lower cervical region (C4-T1) and the lower thoracic upper lumbar region (T9-L1). Delay of surgical intervention is probably the most important modifiable prognostic factor for recovery after SSEH, on the other hand, a short progression interval, no symptom relief after admission; hypesthesia, complete spinal cord injury, and hematoma below the T4 level are reported as unmodifiable prognostic factors [5].

CASE REPORT

History

A 22-year-old young lady without history of past illness presented to the emergency department in our hospital with 4 hours of sudden onset of muscle weakness and urinary retention without trauma. Except a neck pain, she denied any other complaints and any systemic diseases. She was conscious and did not have any difficulty of communication. Initially, she was referred to the neurology clinic for medical attention and misdiagnosis as transverse myelitis. Before in-patient admittance, a routine further consultation of surgical specialist was carried out, then emergency MRI scan was suggested to identify the condition of spinal cord.

Examinations

Vital signs were stable and there was no fever. Physical examination revealed absent motor and sensory function below shoulders of both side. Disappear of deep tendon reflex was noted. Babinski sign was negative bilaterally. Muscle tonus of all extremities was highly decreased, there was no voluntary muscle contraction. Fortunately, proprioceptive sense was preserved of both lower limbs. The emergency MRI revealed an extradural mass compressing the spinal cord at C4-6 levels (Fig. 1,2,3). The mass almost had the similar signal intensity as cerebrospinal fluid on T1- and T2- weighted images, and contrast enhancement was not preformed. Pre-operative laboratory examinations consisted of complete blood count, electrolytes, liver, renal function tests, urinalysis. They were all normal. After main differential diagnoses were excluded, with the presence of typical MRI findings, the diagnosis of Spontaneous spinal epidural hematoma was concluded, a rare neurologic emergency of the spinal cord, which is caused by a hemorrhage in the epidural space.

Surgical treatment

The C4-6 levels of epidural hematoma compression were considered to be responsible of clinical symptoms. Surgical posterior decompression was performed 4 hours after admission. After standard single door laminoplasty of C4-6, immediately obvious

was the extent of sponge like vascular structure in this area, causing a mass effect on the spinal cord. However, there were no fibrous adhesions. The hematoma was removed completely without difficulty. To preserve cervical movement, internal fixation was carried out (Fig. 4,5). Then the wound was closed in the usual multi-layered fashion. She was treated postoperatively with a rigid hard plastic collar that had to be worn constantly for 4 weeks.

Pathological findings and Postoperative course

Histopathological examination confirmed the lesion to be vascular structures without evidence of arteriovenous teratism. 3 days after surgery her muscle tonus was inceased and muscle strength was improved. The manual muscle test demonstrated 3/5 strength on the lower limbs and 2/5 on the upper limbs. The dysuria function was not impaired. After 1 week of rehabilitation, the patient can stand up depended on herself. After an uncomplicated hospital course, he was discharged home 2 weeks later. After follow-up for 3 months, there was no recurrence of neck pain, neurologic examination was normal. Cervical range of motion 40° flexion, 40° extension, 65° left rotation, and 65° right rotation(Fig. 6,7).

The design and performance of this study conformed to ethical standards of Helsinki Declaration and our national legislation. It was approved by Medical Ethical Committee of our institution. The patient was enquired whether or not willing to take part in a scientific research and informed consent forms were signed by her.

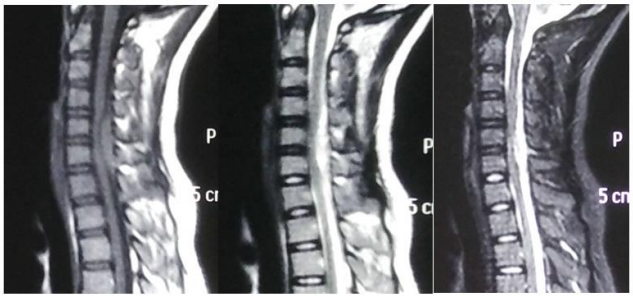

Fig-1, 2, 3 T1, T2 and lipid suppressor MR images of spontaneous spinal epidural hematoma

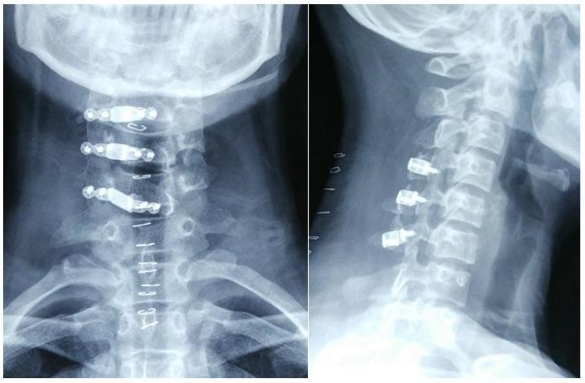

Fig-4, 5 post-operative X-ray pictures of single door laminoplasty

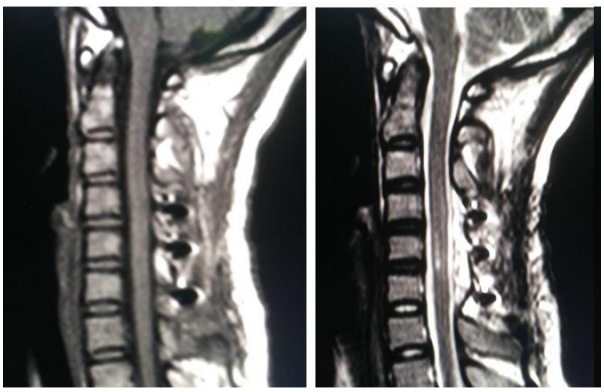

Fig-6, 7 T1, T2 MR images post-operation

DISCUSSION

The incidence of SSEH has been reported to be 1 in 100,000, which is a rare neurosurgical emergency [1]. The most common symptoms include sudden severe neck or back pain accompanied by plegia of the extremities, bladder, and bowels [1,4,5]. SSEH affects mainly adults, including elderly adults, and is rare in children [1,3,5-7]. The majority of these studies included cases with coagulation disorders, thrombolytic therapy, vascular malformation, hemophilia, and pregnancy[1,3,8,9]; the abnormal vascular structure is the source of bleeding in SSEH cases, from the posterior vertebral venous plexus[1,3]. A sudden increase in venous epidural pressure in the posterior internal vertebral venous plexus caused by valsalva maneuver, such as sneezing, voiding, vomiting, and lifting, may be predisposing factors. Reports of pure spontaneous SSEH without any such predisposing factors are uncommon [3,5-11]. Importantly, in children, hemophilia is a notable risk factor for SSEH. However, in this report, the above predisposing factor was not detected.

The locations commonly seen in SSEH are C5-T2 and T12[1], where the majority of SSEHs are located dorsal to the sac[3,4]. Acute local neck and back pain with progressive hemiparesis or tetra paresis should promptly warrant further investigation to exclude the possibility of SSEH. Magnetic resonance imaging of the spinal cord is the gold standard for diagnosing SSEH., with many of these patients only mildly symptomatic[1,3,4,12]. The signal changes depend on the stage of blood (acute/subacute/chronic). Peripheral enhancement is the most common pattern noted on contrast administration [4]. Heterogeneous hyper intensity with focal hypo intensity on T2-weighted MRI sequences should suggest the probability of acute spinal extradural hematoma [4]. In acute stages of blood where the signal will be isointense, computed tomography pyelography can be helpful for confirmation of diagnosis [3].

Preoperative motor function, time to surgery, symptom progression interval, and presence of spinal cord edema, thoracic hematoma, hematoma size, and number of levels involved are suggested to be factors influence outcomes [1, 3, 4, 8, 9, 11, 12-14]. Since the canal diameter is narrowest in the thoracic segment, making the thoracic spinal cord more susceptible to compression-induced ischemia caused by the hematoma [1,14,15]. More than 4 levels hematoma, rapid progression with a shorter progression interval (<12 hours) and spinal cord edema are associated with a worse prognosis[1,8,11,12-14]. Nevertheless, time to surgery is a critical factor. Hejazi et al[3] analyzed 9 cases of SSEH, All patients underwent multilevel laminectomy, and only 3 patients experienced full improvement (2 with incomplete deficits and 1 with complete deficits), 2 of whom were operated on within 18 hours, whereas the remaining 6 patients operated on after 24 hours showed only slight or no improvement. Other authors have reported similar findings[8,16]. On the other hand, there is no significant differences in age between patients with good and poor neurologic outcomes[1,12]. However, preoperative neurologic status has been reported to be the most consistent and critical factor correlating with the prognosis and outcome in almost all studies [9, 11, 12-14].

Conservative treatment should be considered for patients with a benign clinical process, many studies have reported that conservative treatment is suitable with mild clinical presentations and relatively rapid improvement, which can yield a preferable clinical outcome. Coagulation status should be normalized in patients on oral anticoagulant therapy to minimize progression of the hematoma[5,8,17,18].Although conservative treatment of SSEH may recover spinal cord function, it does not prevent the recurrence of an episode or hemorrhage[2,4,11,14,19-21]. 34.5% of these patients will experience multiple episodes. It is shown that severe complications are often followed by re-bleeding. 30% of the patients experienced residual deficits or even death after an SSEH attack [22] .Thus; we should pay attention that the re-occurrence of an episode with such a tragic end is highly probable.

In contrast, after receiving operations, none of the patients experienced subsequent episodes or re-bleeding. The relatively simple procedures of laminectomy/multilevel laminectomy over the most affected segment is the treatment of choice in the majority of cases[3,19]. In addition, complications caused by such operations are quite rare. Nevertheless, Early diagnosis and emergency surgery helps to maximize the recovery [8,11].

CONCLUSIONS

When sudden onset of neurological deficits is observed in clinical practice, Spontaneous spinal epidural hematoma should be taken into consideration. MRI scan is essential in differential diagnosis and surigcal planning. Although conservative treatment is a choice for patients with a benign clinical process or mild clinical presentations, surgical decompression should be considered to stop subsequent episodes or rebleeding.

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