

Acute Spontaneous Cervical Epidural Hematoma Mimicking Cerebral Stroke: A Case Report and Literature Review

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Spontaneous cervical epidural hematoma (SCEDH) is a rare disease, but can cause severe neurologic impairment. We report a case of a 68-year-old female who presented with sudden onset, posterior neck pain, right shoulder pain, and progressive right hemiparesis mimicking stroke with no trauma history. Initial brain CT and diffusion MRI performed to rule out brain lesion did not show any positive findings. Laboratory examination presented only severe thrombocytopenia ($45,000/\text{mm}^3$). Subsequent cervical MRI revealed a cervical epidural mass lesion. We confirmed that it was pure hematoma through C5 unilateral total laminectomy and C6 partial hemilaminectomy. She achieved complete neurologic recovery with active rehabilitation. Early surgical decompression for SCEDH with neurologic impairment should be recommended for better outcome.

Key Words: Spontaneous cervical epidural hematoma (SCEDH) • Stroke • Liver cirrhosis

INTRODUCTION

Spontaneous cervical epidural hematoma (SCEDH) is an uncommon disorder and can develop signs and symptoms of spinal cord compression. Clinically, it can present as a wide range of neurological deficits from simple cervical radiculopathy to complete quadriplegia depending on the severity and rapidity of compression. Such neurological deficits are reported to be associated with blood coagulopathies, anti-coagulant treatment, infection, tumor genesis, pregnancy, herniated discs, Paget's disease, and vascular malformations⁸⁾. Although several predisposing factors have been proposed, the exact bleeding source and mechanism are still uncertain. Recently, we successfully treated a case of spontaneous cervical epidural hematoma mimicking epidural mass-like lesion with different MRI findings in a liver cirrhosis patient.

CASE REPORT

A 68-year-old female presented with sudden onset posteri-

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or neck and right shoulder pain with progressive right side weakness while falling asleep. During consult, she had reported taking anti-hypertensive medication for 20 years and was followed regularly for hepatitis C. She reported no trauma history or physical exertion for the past few weeks. Neurological examination resulted in alert mental status and the detection of right hemiparesis. Motor power of her right upper limb was grade III and ipsilateral hand grasping power was grade II. In her right lower extremity, motor power was grade IV, and her left whole extremities were intact. Right side hypoesthesia was also observed. Based on her medical history and clinical symptoms, the physician working at the emergency department ordered brain CT to rule out intracranial hemorrhage. However, brain CT did not show any positive findings (Fig. 1-A). Subsequent brain diffuse MRI was taken to rule out cerebral infarction, but it was also negative (Fig. 1-B, C). Cervical MRI was performed to rule out a cervical lesion based on the patient's report of posterior neck pain and revealed a mass like lesion in the right posterior epidural

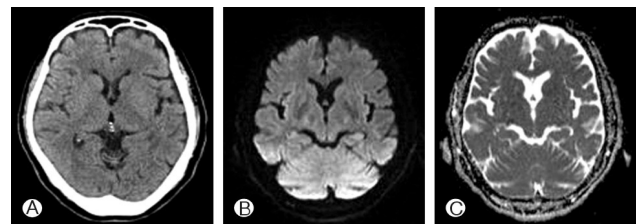


Fig. 1. Brain CT (A) and MR diffusion images (B, C) showing no intracranial lesion.

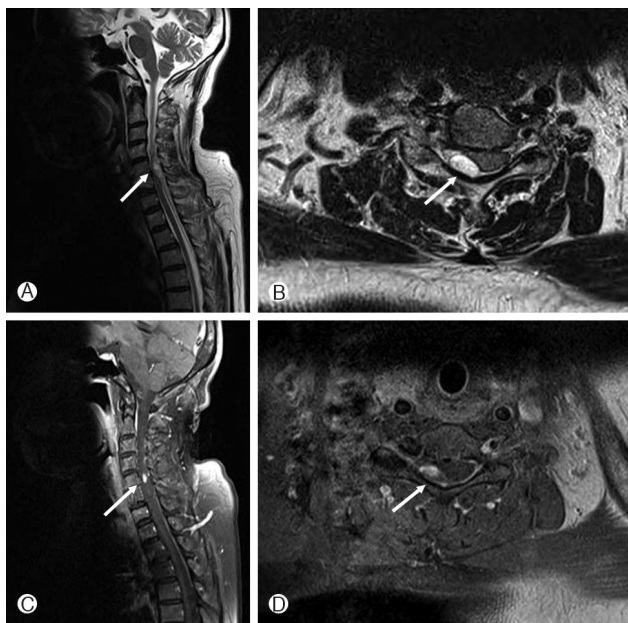


Fig. 2. Mid sagittal (A, C) and axial MRI (B, D) of the cervical spine showing a mass like lesion with enhancement.

space at the C3-6 level compressing the right side of the spinal cord. The mass was isointense to the spinal cord on T1WI and heterogeneously hyperintense with central high signal foci on T2WI. At the C5 level, the MRI showed an ovoid, enhancing, nodular lesion, which was a suspected tumorous condition such as hemangioma or hemangiopericytoma (Fig. 2). Initial laboratory tests showed only severe thrombocytopenia ($45,000/\text{mm}^3$). As her neurologic signs were getting worse with time, we decided to perform surgical intervention. Under general endotracheal anesthesia via light wand intubation, right C5 unilateral total laminectomy and C6 partial hemilaminectomy were performed. The mass lesion was pure hematoma and was removed (Fig. 3). We could find neither vascular malformation or an active bleeding point. She went into rehabilitation after her operation and finally achieved complete neurologic recovery and relief of neck and shoulder pain.

DISCUSSION

Spontaneous spinal epidural hematoma is a rare disease and a total of 470 cases have been reported worldwide since 1869⁹. Beatty et al. described SCEDH as idiopathic spontaneous spinal epidural hematoma⁴. Lonjon et al. used “nontraumatic spinal epidural hematoma” to describe idiopathic SCEDH with liver cirrhosis secondary to some other pathology¹³. Because various descriptions have been used for idiopathic spinal EDH, we used “spontaneous” to emphasize “the absence of any trauma or iatrogenic injury”. Risk factors of SCEDH

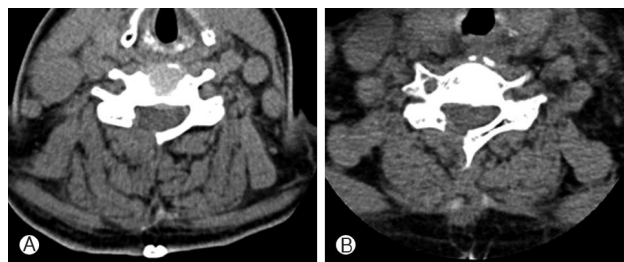


Fig. 3. Cervical axial CT images after surgery showing complete hematoma evacuation (A: C5 level, B: C6 level, respectively).

include hypertension, coagulopathy, anti-coagulant use, Paget’s disease, tumor genesis, and vascular malformations. However, an exact bleeding source and mechanism of hematoma have not yet been revealed.

It is generally accepted that most cervical epidural hematomas arise from rupture of the epidural venous plexus^{3,8,10}. However, it is still being debated whether the origin of bleeding in acute SCEDH is arterial or venous. Some authors have favored an arterial origin as the bleeding source. For example, Beatty and Winston postulated that the source of bleeding was the ‘free’ anastomotic arteries in the epidural space that connect with radicular arteries⁴. The probability of arterial bleeding is also supported by the rapid development of spinal cord compression¹⁶. Conversely, Bruyn and Bosma theorized that spontaneous spinal epidural hematoma occurs due to local pooling within valve-less, thin-walled epidural veins and brief increases in intravenous pressure (caused by intra-thoracic and intra-abdominal pressure elevations) leading to epidural vein rupture⁵. This theory of how venous pooling and pressure causes vessel vulnerability could explain why bleeding occurs in SCEDH with liver cirrhosis, and possibly accounts for the reported associations with voiding, bending, turning in bed at night, straining while toileting, sneezing, and coitus^{5,8}. Liver cirrhosis is a systemic disease that affects the entire circulatory system and causes abnormalities in venous plexuses in the cervical region. In addition to coagulation abnormalities, liver cirrhosis causes portal vein hypertension, resulting in the development of collateral venous flow from splanchnic circulation. The epidural veins, like the azygous or hemiazygous thoracic veins, would be swollen and their walls would become thin. Thus, in patients with liver cirrhosis, the epidural venous plexus can easily be damaged with neurologic deficits becoming more readily apparent with less blood pooling in epidural spaces. However, our case did not show any evidence of definite venous plexus or vascular malformation, grossly.

The clinical manifestations of SCEDH can resemble those of a ruptured cervical disc, an epidural mass, or a dissecting aortic aneurysm¹⁵. Typical clinical features are sudden, localized, intensive pain around the involved vertebrae, often show

radiculopathy which mimic disc rupture. Some authors insisted that the characteristics clinical symptoms of SCEDH are acute onset of radicular paresthesia and localized pain and progressive paraplegia and loss of sensory function within minutes or hours¹⁵. However, it is not always easy to diagnose SCEDH because various symptoms may be often confused with myelitis, polyradiculitis, myelocompressive disease including tumor, intracranial lesions such as aneurysmal rupture, or thalamic hemorrhage. SCEDH may sometimes present as hemiparesis or hemiplegia without neck pain. Hsieh et al. reported a case of SCEDH in a 65-year-old man complaining of acute onset unilateral hemiparesis the initial presentation¹². Patients with hemiparesis due to SCEDH have often been misdiagnosed as having cerebral infarction and had been treated with anti-platelet therapy of anti-coagulants^{1,12,19}. The symptoms of the present case were enough to lead to misdiagnosis even though the epidural hematoma compressed the cervical cord and neural foramen directly.

MRI is the most useful method for positive diagnosis of spontaneous and secondary cervical epidural hematoma. Many authors reported various signal intensities with differing time intervals. Matsumura et al. reported that SCEDH usually exhibits an isointense signal to the spinal cord within 24 h after symptom onset and a hyperintense signal after 36 hours on T1WI¹⁴. Groen et al. reported a characteristic hyperintensity with hypointense foci on T2WI⁸. Melanie et al. insisted that SCEDH showed various intensities including hyper-, iso- or hypointensity on T1WI and did not correlate with time from symptom onset to imaging. They also reported that it showed heterogeneously hyperintense to spinal cord with foci of hypointensity on T2WI⁷. However, MRI findings in the present case showed a different pattern compared to previous reports. It showed isointensity to spinal cord on T1WI, but heterogeneous hyperintensity with central foci of hyperintensity on T2WI that did change with time. Nodular enhancing ovoid lesion was suspected with tumorous condition such as hemangiopericytoma or hemangioma.

Several cases of SCEDH have been reported to treat conservatively^{2,11,18}. All cases involved mild neurological symptoms and quadriplegia at examination. However, many neurosurgeons still advocate rapid diagnosis and surgical hematoma as the standard management to recover neurologic deterioration. Choi et al. emphasize rapid surgical decompression in cases of symptomatic epidural hematoma⁶. Groen et al. reported significantly better outcomes for patients with complete neurologic deficit that underwent decompression within 36 h of symptom onset and within 48 h in patients with incomplete deficit⁸. Shin et al. emphasized the importance of early decompression at less than 12 h for incomplete neurological deficits associated with ischemic change, and recommended a shorter

time to surgical intervention to promote positive neurological outcomes¹⁷. In our case, surgery was performed within 24 h of the development of initial neurologic signs. Although our patient had bleeding tendency due to severe thrombocytopenia, her postoperative state was favorable with no evidence of complication. Our experiences and the literature suggest that recovery of neurological impairment is related to the time elapsed between symptom onset and surgical decompression. Accurate and early diagnosis of SCEDH and prompt surgical intervention, within 24 h, alleviates sensory motor deficits and may contribute to complete neurologic recovery¹⁷.

CONCLUSION

Spontaneous cervical epidural hematoma is a rare disorder and can present various clinical patterns and easily be misdiagnosed. As was shown by our case, early diagnosis based on MRI findings and hematoma evacuation within 24 h of symptom onset can lead to full neurologic recovery in patients with spontaneous cervical epidural hematoma.

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