Brown-Sequard syndrome: a rare presentation of spontaneous cervical spinal epidural haematoma

脊髓半切綜合徵：頸椎自發性脊髓硬膜外血腫的一種罕見表現

SY Hsu 許劭遠, CJ Chang 張志儒, CT Hsieh 謝政遠

Spontaneous spinal epidural haematoma is a rare but well-known entity that has been considered a surgical emergency. Brown-Sequard syndrome resulting from spontaneous cervical spinal epidural haematoma is rare. Only 11 cases have been described in the literature. We reported an 85-year-old female who presented with Brown-Sequard syndrome at the right C5 level, and an acute haematoma located from the C3 to C6 level was diagnosed from the series images. Within two weeks after decompressive surgery, the previous neurological deficits had gradually improved. (Hong Kong j.emerg.med. 2014;21:107-111)

自發性脊髓硬膜外血腫是一種罕見但眾所周知的外科急症。頸椎自發性脊髓硬膜外血腫造成脊髓半切綜合徵是罕見的。在文獻中只有 11 個病例。我們報告一個 85 歲的女性病人，有右側 C5 疼痛的脊髓半切綜合徵，一系列影像診斷為位於從 C3 到 C6 水平的急性血腫。減壓手術後兩個星期內，先前的神經功能缺損已逐漸改善。

Keywords: Functional laterality, magnetic resonance imaging, spinal cord compression, stroke

關鍵詞：功能偏側、核磁共振成像、脊髓受壓、中風

Introduction

Spontaneous spinal epidural haematoma (SSEH) is a rare spinal disease with unknown aetiology, and is a surgical emergency.\(^1\) With the use of advanced imaging techniques, such as magnetic resonance (MR) images, early identification of this disease is made easier.\(^2\)

Although the clinical presentation varies, Brown-Sequard syndrome as a presentation of SSEH has been infrequently discussed in the literature. Herein, we report a case of cervical SSEH with Brown-Sequard syndrome and the relevant literature was reviewed.

Case report

An 85-year-old female developed acute neck pain while she was sawing the branches of trees in her garden. Within 30 minutes, the pain aggravated and radiated to the right shoulder, combining with tingling and numbness, and she developed weakness in her right arm. She felt that the arm did not belong to her, and the symptoms would worsen when she flexed her neck forward. She was brought to our emergency department (ED) 2 hours after the onset of symptoms. On arrival, she was alert and oriented, with blood
pressure of 240/105 mmHg, pulse 74 beats/min, respiration rate 22/min, and temperature of 36 degrees Celsius. Neurological examination revealed normal mental functions and no significant deficit of the cranial nerves. Neck movement was limited by pain. Sensory examination revealed diminished pinprick and temperature sensations in the left limbs below the C5 level. Proprioceptive and vibration sensations were diminished in the right limbs below the C5 level. Muscle power was grade 3 over 5 in the right limbs and 5 over 5 in the left limbs. The deep tendon reflexes increased in the bilateral lower limbs and diminished in the bilateral upper limbs. The clinical presentations were consistent with Brown-Sequard syndrome at the right C5 level. Laboratory examinations, including the coagulation profiles, were non-contributory. The radiographs of the cervical spine revealed no obvious fractures or subluxations. Computed tomographic (CT) scan of the cervical spine within 2.5 hours of symptom onset revealed a high-density extradural mass located in the right posterior-lateral aspect of the spinal canal, extending from the C3 to C5 level and compressing the spinal cord. On the subsequent MR images of the cervical spine within 6.5 hours of symptoms onset, the mass appeared as an iso- to hypointensity on T1-weighted images and hyper-intensity on T2-weighted images, locating from C3 to T1. With gadolinium administration, no obvious enhancement was noted. The majority of the mass located from C3 to C6 and at the spinal cord was deviated to the left side (Figures 1 & 2).

A diagnosis of cervical SSEH was made based on the clinical presentation and radiological images. Mega-dose steroid therapy with methylprednisolone (a bolus dose with 30 mg/kg followed by a maintenance dose with 5.4 mg/kg for 48 hours) was prescribed at the ED. She underwent emergency decompressive laminectomies from C3 to C7 within 8 hours of symptom onset. At operation, a blood clot located in the right posterior-lateral aspect of the spinal canal was completely removed. No obvious vascular abnormalities or neoplasms were found. After haematoma evacuation, the spinal cord re-expanded and had pulsatile movement. The sensory impairments resolved from the first day after operation, and the patient recovered completely from the weakness within two weeks after operation.

**Figure 1.** T2-weighted magnetic resonance images of the cervical spine. The sagittal image reveals the high-intensity mass located at the posterior aspect of the spinal cord from C3 to T1.

**Figure 2.** T2-weighted magnetic resonance images of the cervical spine. The axial image revealed the mass located in the right posterior-lateral aspect of the spinal canal, which compressed the spinal cord.
Discussion

SSEH is a rare but devastating entity with unknown definite aetiologies. The annual incidence is estimated to be approximately 0.1 per 100,000 people. The majority of SSEH cases occur in middle-aged or older patients. The haematomas were commonly found from the C5 to T2 level. A higher predominance of male patients has been noticed. Although this bleeding event is frequently associated with haemorrhagic conditions, anticoagulant administration, or vascular malformation, it may also occur in healthy subject. Hypertension was also considered as an important contributing factor.

The mechanism of SSEH remains unclear. Since the majority of haematomas are located on the posterolateral aspect of the spinal canal, bleeding from the posterior valveless epidural venous plexus during the transient raising of venous pressure induced by

<table>
<thead>
<tr>
<th>Author/Year</th>
<th>Gender/Age (years)</th>
<th>Medical history</th>
<th>Coagulation profile</th>
<th>Location of haematoma</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Russman, et al./1971</td>
<td>Female/53</td>
<td>Hypertension</td>
<td>Normal</td>
<td>C6</td>
<td>Decompressive laminectomy, C4-C8</td>
<td>Complete recovery within 10 days</td>
</tr>
<tr>
<td>Murata, et al./1984</td>
<td>Male/75</td>
<td>Hypertension</td>
<td>Normal</td>
<td>Left dorsolateral, C2-C5</td>
<td>Decompressive laminectomy, C2-C5</td>
<td>Unknown</td>
</tr>
<tr>
<td>Mustafa, et al./1987</td>
<td>Female/71</td>
<td>Factor XI deficiency</td>
<td>PT prolong</td>
<td>Right dorsolateral, C5-C6</td>
<td>Decompressive laminectomy, C5-C6</td>
<td>Complete recovery within 4 months</td>
</tr>
<tr>
<td>Chou, et al./1988</td>
<td>Female/75</td>
<td>Hypertension</td>
<td>Normal</td>
<td>Right dorsolateral, C3-6</td>
<td>Decompressive laminectomy, C3-C7</td>
<td>Mild residual numbness in the left leg one month after the operation</td>
</tr>
<tr>
<td>Ehsan, et al./1996</td>
<td>Male/69</td>
<td>Haemorrhagic lymphoid follicular hyperplasia of the small bowel</td>
<td>Normal</td>
<td>Left dorsolateral, C5-C6</td>
<td>Conservative</td>
<td>Complete recovery within 17 days</td>
</tr>
<tr>
<td>Adamson, et al./2004</td>
<td>Male/63</td>
<td>Hypertension</td>
<td>Normal</td>
<td>Right dorsolateral, C3-C5</td>
<td>Decompressive laminectomy, C3-C5</td>
<td>Good recovery within 4 months</td>
</tr>
<tr>
<td>Gill, et al./2004</td>
<td>Female/71</td>
<td>Hypertension</td>
<td>Normal</td>
<td>Right dorsolateral, C5-C7</td>
<td>Decompressive laminectomy</td>
<td>Slow recovery</td>
</tr>
<tr>
<td>Spengos, et al./2005</td>
<td>Female/66</td>
<td>Hypertension</td>
<td>Normal</td>
<td>Left dorsolateral, C4-C7</td>
<td>Decompressive laminectomy, C4-C6</td>
<td>Good recovery within 6 days</td>
</tr>
<tr>
<td>Neetu, et al./2006</td>
<td>Male/55</td>
<td>Hypertension</td>
<td>Coagulopathy</td>
<td>Right dorsolateral, C1-C5</td>
<td>Conservative</td>
<td>Weakness improved within 24 hours and bladder symptoms resolved within 15 days</td>
</tr>
<tr>
<td>Ofuoglu, et al./2009</td>
<td>Male/63</td>
<td>Unknown</td>
<td>Normal</td>
<td>Right dorsolateral, C4-C7</td>
<td>Decompressive laminectomy, C5-C6</td>
<td>Complete recovery within 2 months</td>
</tr>
<tr>
<td>Panciani, et al./2009</td>
<td>Male/52</td>
<td>Unknown</td>
<td>Normal</td>
<td>Left dorsolateral, C2-C5</td>
<td>Conservative</td>
<td>Complete recovery within 2 days</td>
</tr>
<tr>
<td>Our case/2012</td>
<td>Female/85</td>
<td>Hypertension</td>
<td>Normal</td>
<td>Right dorsolateral, C3-C6</td>
<td>Decompressive laminectomy, C3-C6</td>
<td>Complete recovery within 2 weeks</td>
</tr>
</tbody>
</table>
activities such as cough, sneezing or holding of heavy baggage has been considered the main cause of SSEH.\textsuperscript{5}

The typical presentation of cervical SSEH is characterised by sudden onset neck pain followed by rapid motor paralysis or anaesthesia.\textsuperscript{6} Chest, pain, flank pain, back pain, monoparesis or hemiparesis have been infrequently reported as atypical presentations of SSEH.\textsuperscript{7} Brown-Sequard syndrome is an incomplete spinal cord injury characterised by ipsilateral hemiplegia with contralateral pain and temperature deficits, and that resulting from cervical SSEH is a rare entity, with only 11 cases described in the literature (Table 1).\textsuperscript{8-18}

MR imaging is the "gold-standard" diagnostic method for SSEH.\textsuperscript{2,4} Within 24 hours of onset, the haematoma appears as an iso-intensity on T1-weighted images and a heterogeneous intensity on T2-weighted images.\textsuperscript{2} Twenty-four hours after onset, hyperintensity on both T1- and T2-weighted images is demonstrated. Because of the various clinical presentations of SSEH, different therapeutic strategies have been hypothesized.\textsuperscript{1,19-22} Conservative treatment may be reasonable for those with trivial neurological deficit or if the haematoma is small and not compressing the spinal cord.\textsuperscript{6} Early decompressive laminectomy with removal of the haematoma is advocated for patients presenting with significant and progressive neurological deficits.\textsuperscript{1,19,20} The prognosis is associated with the preoperative neurological condition, symptoms progression, present of spinal cord oedema, and the extension and vertebral level of the SSEH.\textsuperscript{1,19}

In published cases of Brown-Sequard syndrome caused by cervical SSEHs (Table 1), including our case, the mean age was 66.5 years. The prevalence in males and females was equal. A history of hypertension was noted in 8 patients (67%). All haematomas (100%) occurred in the posterior aspect of the spinal canal. Although three patients (25%) did not undergo decompressive surgery, a good recovery was noted in all patients. However, several patients with atypical presentation of SSEH were misdiagnosed as having ischemic stroke and were treated with antithrombolytic agents.\textsuperscript{23} Bleeding from the SSEH would then be worse and more devastating. As in our patient, normal cranial nerve function and the presence of neck pain (or back pain) are important clues to localise the level of lesion at the spinal level. However, with the atypical presentations of cervical SSEH, high clinical suspicion, detailed history-taking, complete neurological examinations, and prompt imaging is the key for early diagnosis.

References