Acute Spinal Subdural Hematoma in a Patient with Active Systemic Lupus Erythematosus: A Case Report and Literature Review

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Abstract

We herein describe a case of acute spinal subdural hematoma (SSDH) during the administration of high-dose corticosteroids and intravenous heparin for the treatment of active lupus nephritis. After SSDH was promptly diagnosed using magnetic resonance imaging (MRI), the patient recovered well with conservative treatment involving the discontinuation of heparin sodium. Although SSDH is a rare complication, it should be considered as a cause of neurological manifestations in patients with active systemic lupus erythematosus.

Key words: MRI, spinal subdural hematoma, systemic lupus erythematosus

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Introduction

Complications involving the spinal cord are rare in patients with systemic lupus erythematosus (SLE) (1). Transverse myelitis, a demyelinating syndrome resulting from the high disease activity of SLE, is the most frequent spinal complication and has been suggested to have a strong association with the presence of anti-phospholipid antibodies (1). However, symptoms that are similar to transverse myelitis may be mimicked in cases of space occupying spinal cord lesions, such as abscesses, hematomas and neoplasms. Intrathecal bleeding should be considered as a differential diagnosis in such cases.

Acute spinal subdural hematoma (SSDH) is an exceedingly uncommon and potentially life-threatening condition. SSDH usually occurs as a result of trauma or an accident on lumbar puncture and rarely occurs spontaneously in cases of hematological disorders, vascular malformations, tumors or anticoagulant use (2, 3).

We herein describe a case of SSDH that occurred during the administration of high-dose corticosteroids and intravenous (IV) heparin for the treatment of active lupus nephritis. In addition, we provide a review of the literature.

Case Report

A 30-year-old woman presented to our hospital with general fatigue, digital swelling and dyspnea on exertion lasting for three months. A hematological examination revealed pancytopenia (red blood cell count, 314×10⁴/mm³; hemoglobin level, 8.5 g/dL; hematocrit, 26.3%; white blood cell count, 2,900/mm³; platelet count, 11.8×10⁴/mm³). A urinalysis revealed protein 2+ and occult blood 2+. Blood chemistry and serological tests showed an elevated creatinine level of 1.09 mg/dL, a C-reactive protein level of 1.1 mg/dL, a decreased C3 level of 32 mg/dL, a C4 level of 4 mg/dL, an elevated antinuclear antibody (ANA) level of ×81,920 (peripheral pattern) and an anti-double-stranded (ds) DNA antibody level of >400 IU/mL. A chest X-ray scan indicated cardiomegaly (cardiothoracic ratio: 60%), and echocardiography revealed a medium amount of pericardial effusion. Although a prolonged activated partial thromboplastin time (aPTT) of 41 seconds and an elevated D dimer value of 3.7 ng/dL were observed, other coagulation parameters, including the levels of protein S, protein C and anti-

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thrombin III, were within the normal ranges. Anticardiolipin (CL) antibodies and lupus anticoagulant were later proven to be negative.

The patient was diagnosed with SLE, fulfilling the 1982 American College of Rheumatology (ACR) criteria, with non-erosive arthritis, pericarditis, a renal disorder, a hematological disorder and the presence of anti-ds DNA antibodies with positive ANA. The SLE disease activity index (SLEDAI) score was 23, indicating a very high disease activity.

The administration of intravenous pulse methylprednisolone (1,000 mg/day for three days) followed by oral prednisolone (60 mg daily) was started to treat the lupus nephritis and pericarditis. Considering for the possibility of antiphospholipid antibody syndrome (APS), as indicated by the prolonged aPTT, and high-dose corticosteroid-induced accelerated coagulability, IV heparin sodium (15,000 units/day) was also administered.

On the ninth day of treatment, the patient presented with sudden back and chest pain and, a few hours later, experienced a headache and stiffness in her neck. A physical examination revealed tachycardia and a high blood pressure of 220/146 mmHg; however, no apparent objective neurological findings were detected. Blood examinations, electrocardiography, echocardiography and contrast-enhanced computed tomography (CE-CT) did not demonstrate any signs of acute coronary syndrome, aortic dissection, pulmonary embolism or intracranial bleeding. However, a small lesion of increased signal density was observed in the spinal cord at the seventh thoracic vertebral level on a CE-CT scan (Fig. 1).

The possibility of the prodromal stage of transverse myelitis due to the activity of SLE and intrathecal bleeding were considered as differential diagnoses. An MRI scan of the spine revealed a small, round lesion with a highly intense signal on T2-weighted images and an isointense signal on T1-weighted images; the lesion appeared to compress the spinal cord at the seventh thoracic vertebral level (Fig. 2). These results were consistent with the findings of the acute phase of SSDH (2, 3). No signs of transverse myelitis were observed (1). Bloody cerebrospinal fluid was noted on a spinal tap. A coagulation test showed a prolonged aPTT of 53 seconds induced by the administration of heparin sodium.

Conservative management comprised discontinuation of the heparin sodium infusion. Blood pressure control and pain relief were also included because the patient did not exhibit any motor, sensory or autonomic nervous disorders. Her back pain and headache were palliated within six days after onset. She recovered well with no neurological sequelae. An MRI scan performed 26 days after onset indicated that the hematoma had almost completely regressed. A contrast-enhanced MRI scan did not show any signs of vascular malformation or tumor-like structures at the site of bleeding.

We herein reported a case of acute SSDH that occurred during the administration of high-dose corticosteroids and heparin sodium infusion in a patient with SLE.

Spinal involvement is a very rare manifestation of SLE; however, this condition is intractable and often refractory, resulting in neurological sequelae. A prompt diagnosis and appropriate treatment are required.

Transverse myelitis, a demyelinating syndrome that results from the high disease activity of SLE, is the most frequent spinal complication (1); however, similar symptoms may be mimicked by space-occupying spinal cord lesions. Intrathecal bleeding should be considered as a differential diagnosis. Only nine reports of intrathecal bleeding during the clinical course of SLE have been published in the literature (4-12) (Table). In these cases, the bleeding sites covered a wide range of the spine. Of the nine reported cases, hematomas were observed in the thoracic spine in three cases and in the cervical spine in four cases. No unique spinal bleeding sites associated with SLE were noted among the small number of examples. Interestingly, six of the nine reported cases involved hemorrhagic events when the patients exhibited a high level of disease activity of SLE (4, 6, 7, 9-11). Vasculitis at the bleeding site was pathologically documented in three of the six active cases (4, 10, 11). On the other hand, Satoh described the a case of a patient who developed a spinal epidural hematoma when the SLE disease activity was low and stable, without vasculitis or vascular malformation at the site of bleeding (12).

In the present case, the hematoma appeared on the ninth day of the initial treatment for lupus nephritis and pericarditis. The serum complement value and the CRP level had returned to normal, whereas the anti-ds DNA antibody level was still high. The SLEDAI score at the onset of bleeding...
was 18. Although it had decreased from a score of 23 at the initiation of treatment for SLE, it continued to indicate a high disease activity. There is a possibility that local vasculitis caused by the SLE activity led to small vascular rupture.

Thrombosis in the spinal vessels can trigger local vasculitis, resulting in the formation of a hematoma. The possibility of APS was considered due to the patient’s prolonged aPTT and slightly elevated D dimer level on admission. However, other coagulation parameters were normal, and tests for both anti-CL antibodies and lupus anticoagulant were negative. The patient also had no history of vasculitis, associated with SLE and APS (8, 14), whereas no bleeding risk factors or bleeding complications have been reported in a few patients with SSDH. Regarding SLE, anticoagulant therapy is usually administered in cases complicated with APS. Bleeding complications have been reported in a few patients with SLE and APS (8, 14), whereas no bleeding risk factors or unique bleeding sites associated with SLE have been reported.

Most patients with SSDH exhibit neurological disorders that require emergent decompression of the spinal cord to prevent irreversible neurological damage (2, 3). Even if prompt surgical evacuation of the hematoma is applied, the outcomes are rather unsuccessful in patients with moderate to severe neurological deficits (2, 3). The present patient did not display any neurological disorders, other than severe

![Figure 2](image)

**Figure 2.** A sagittal magnetic resonance imaging scan of the spinal cord on the day after onset shows a small, round lesion at the seventh thoracic vertebral level with an isointense signal on a T1-weighted image (a) and a highly intense signal on a T2-weighted image (b). An axial T2-weighted image of the spinal cord demonstrating a highly intense signal and round lesion compressing the spinal cord at the seventh thoracic vertebral level (c).

<table>
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<tr>
<th>Age</th>
<th>Sex</th>
<th>Type of bleeding</th>
<th>Type of lupus</th>
<th>Status of lupus</th>
<th>APS</th>
<th>Anticoagulant use</th>
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<td>No</td>
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<td>heparin sodium</td>
<td>Present case</td>
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SSDH: spinal subdural hematoma, SSAH: spinal subarachnoid hemorrhage, SEDH: spinal epidural hematoma, APS: anti-phospholipid antibody syndrome, N/A: not addressed
back and chest pain, despite the administration of spinal compression at the site of the hematoma, and recovered well with conservative treatment. The administration of steroids, with or without surgical evacuation, has resulted in a good neurological recovery in some SSDH case reports (15, 16). The size of the hematoma in reported lupus patients with good neurological improvement remained within three vertebrae and was rather smaller than that observed in patients with worse outcomes. Tang reported a case of cerebral and spinal subarachnoid hemorrhage in a patient with active lupus (4). Despite having a massive spinal hemorrhage, the patient recovered well under prompt aggressive immune therapy and ventricular drainage. In the present case, the size of the hematoma was rather small and remained within the width of one vertebra; therefore, the spinal cord damage may not be irreversibly serious. Furthermore, she received high-dose corticosteroid treatment for lupus nephritis and pericarditis at the onset of the spinal hematoma. We surmise that the administration of high-dose prednisolone relieves spinal cord damage.

In conclusion, acute SSDH is a neurological emergency that is primarily followed by the development of severe neurological disorders. Although SSDH is a rare complication, it should be considered a possible cause of neurological manifestations in patients with active SLE.

The authors state that they have no Conflict of Interest (COI).

References