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Spontaneous Spinal Epidural Hematoma: Report of Two Cases and Literature Review

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Abstract
Spontaneous spinal epidural hematoma (SSEH) is a rare disease but may lead to life-threatening consequences if not timely diagnosed and managed. Emergent hematoma evacuation is indicated before neurological deficits become irreversible. We report two cases. The first case was a 45-year-old man brought to hospital because of an acute onset of quadriparesis and urinary incontinence. His cervical magnetic resonance imaging (MRI) showed an epidural hematoma at the C5-C6 level with severe spinal cord compression. He underwent an emergency C5-C6 right hemi-laminotomy to remove the clot and decompress the cord. Postoperatively, his left-sided deficits immediately resolved. His urinary function returned to normal two weeks after the surgery. He could independently walk two months later. The second case was a 57-year-old man admitted to the hospital because of severe neck pain and paresthesia in both arms. He had been using an antiplatelet for two months. His MRI revealed an epidural hematoma from C2 to C4 with spinal cord compression on the right. This patient was successfully treated with conservative treatment. If SSEH is left undiagnosed and untreated, the neurological deficits may be permanent. Early emergent hematoma evacuation contributes to a favorable outcome. Conservative management is reasonable if neurological deficits are not severe.
Keywords: Spinal epidural hematoma; Neurologic deficits; Myelopathy; Laminotomy

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Introduction

Spinal epidural hematoma is a rare clinical entity, occupying less than 1% of symptomatic epidural injuries.[1,2] Spontaneous spinal epidural hematoma (SSEH) is defined as blood accumulation within the epidural space, not due to trauma or surgery. Its incidence is estimated to be 0.1% per 100 000 people per year.[3] SSEH may be associated with coagulopathy, vascular malformation, tumors, infections, and minor postoperative vertebral wounds.[1,4,5] Common clinical manifestations are sudden cervical pain or back pain with a progressive para- or quadriplegia, depending on the hemorrhage location.[4] Many patients present with complete sensory and motor loss below the lesion level; others have not shown any symptoms until the disease's late stage. Determining clinical-stage patients has a significant prognostic value because those who have at least partial function may make a full recovery.[6,7] Proper imaging and surgical intervention should be indicated as soon as patients show clinical signs suspicious for SSEH.[7] Until now, research on SSEH has not been sufficiently established. Herein, we report two cases of spontaneous epidural hematoma in the cervical spine, causing cord compression. The clinical manifestations, imaging studies, pathogenesis, and treatment of this rare disorder are discussed.

Case Presentation

Two patients agreed to participate in the report after being explained the purpose of it.

Patient 1

A 45-year-old man presented initially with mild neck pain and numbness in both hands. His paresthesia then radiated to both feet, and he developed a gradual weakness in all extremities over the next 36 hours. At the time of hospital presentation, he demonstrated minor neck pain, paresthesia in all limbs, the right-sided weakness that progressed to quadriparesis, then inability to stand or walk. He denied a history of trauma, anticoagulant, or antiplatelet use. On the examination,

power (Medical Research Council Grading) in left upper extremity was 3-4/5 at C5 and C6, and 2/5 at the C7-T1 levels, left lower extremity was 1-2/5 throughout, right upper extremity was 2/5 at C5 and C6, and 0/5 at the C7-T1 levels, right lower extremity was 0-1/5 throughout. Sensory examination showed bilateral loss of crude touch sensation below T4 level. Pain and proprioception sense were absent in both feet but intact in the hands. His lower limb deep tendon reflexes were diminished; Babinski's sign was negative. One day after admission, he developed urinary incontinence.

Magnetic resonance imaging (MRI) of the cervical spine revealed a biconvex mass in the epidural space that severely compressed the cord at the C5-C6 level and appeared isointense on T1 weighted (T1W) image (Figure 1C) and hyperintense on T2 weighted (T2W) image (Figure 1A and 1D). The mass was non-enhancing on T1W with contrast (Figure 1B). His laboratory workup was unremarkable, including the coagulation studies.

In light of the acute clinical manifestations and the MRI findings, the patient was diagnosed with SSEH at the C5-C6 level, causing cord compression. An emergency C5-C6 right hemi-laminotomy was performed to remove the blood clot (Figure 2B). A well-defined hematoma located within the epidural space was removed (Figure 2A). Histological findings suggested the lesion had some features of an organized hematoma (Figure 3).

Immediately after surgery, the left-sided extremity power almost fully recovered; the right upper extremity improved to 4/5 at C5 and C6 level, 3/5 at C7 level, and 2/5 at C8 and T1 level; the right lower extremity was 1/5 at L1-S1 levels. Both crude touch and pain sensation returned to normal. The patient was discharged one week after surgery and continued his rehabilitation program. At his two-week follow-up, he regained regular urinary function; the right upper extremity power was 4-5/5; his complex hand movements (e.g., holding a spoon, fastening buttons) had improved. However, his right lower extremity weakness had not yet improved significantly. After two months of follow-up, he has since been able to

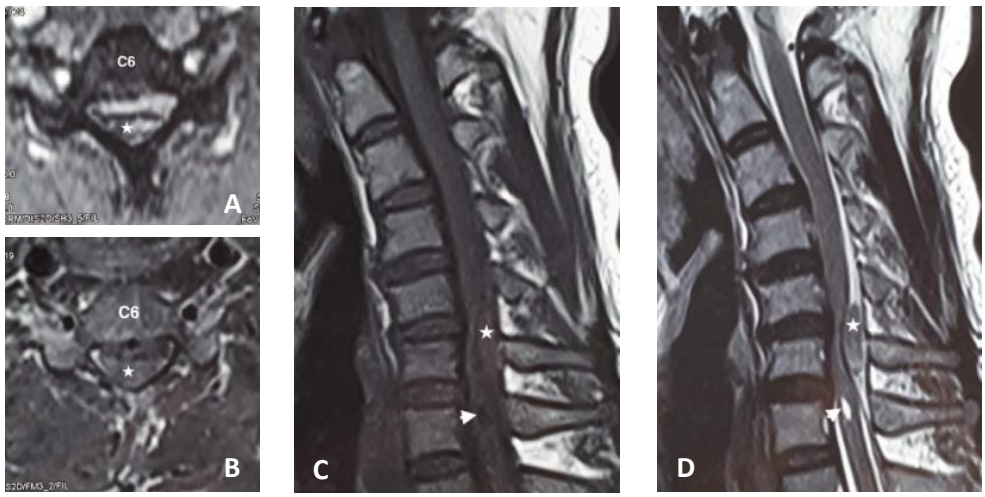

Figure 1. MRI of SSEH showed epidural hematoma at the C5-C6 level (star) and syringomyelia at C7 (head arrow), and revealed heterogeneous hyperintense on T2W image (A, D), homogeneous isointense on T1W image (C), and no enhancement of the SSEH on T1W after Gd administration (B).

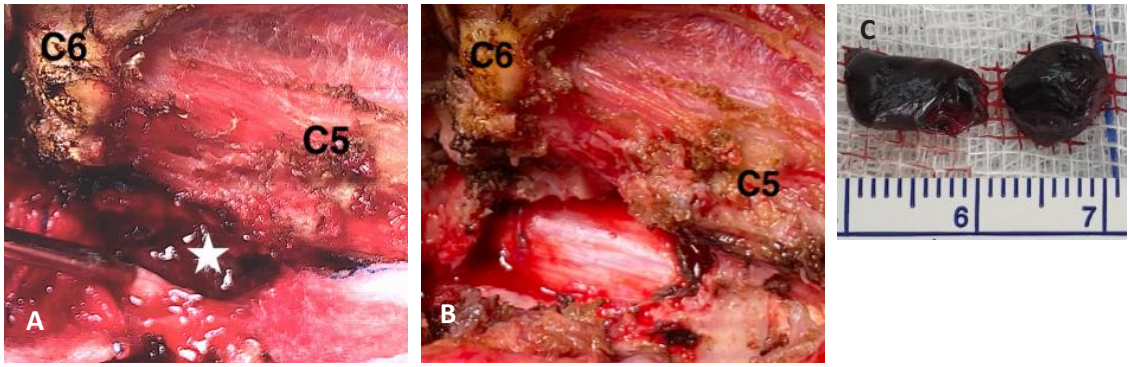

Figure 2. Intra-operation images showed blood clots (star) at epidural space of the C5-C6 level (A) and spinal dural after removing hematoma (B). Hematomas were taken out and measured (C).

walk without any supports. He returned to his job fourth months after operation. Postoperative MRI showed an adequate cord decompression (Figure 4).

Patient 2

A 57-year-old man presented with sudden severe neck pain with numbness in both arms for about one day. He still could walk and urinate normally at the time of hospital admission. After being treated with pain medications, he felt relief of neck pain and numbness in both arms. On cervical MRI, there was a biconvex mass in the epidural space at the C2-C4 level on cervical MRI, which slightly compressed the spinal cord on the right. The features on MRI were similar to those of the first patient (Figure 5). His laboratory workups were almost normal. He was being treated for his diabetes mellitus and

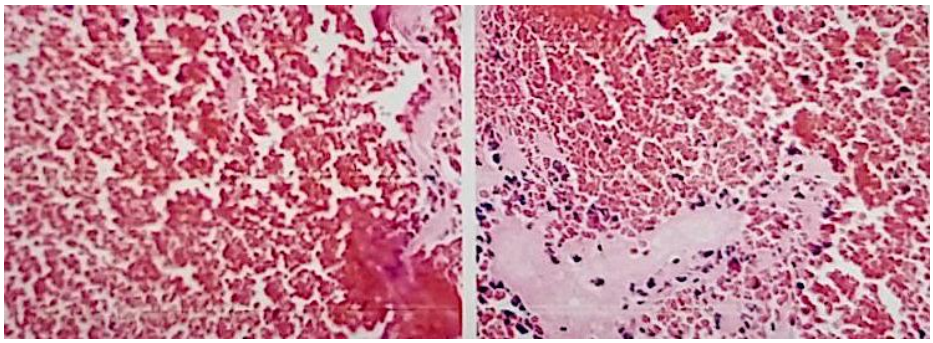

Figure 3. Histological result showing an organized hematoma.

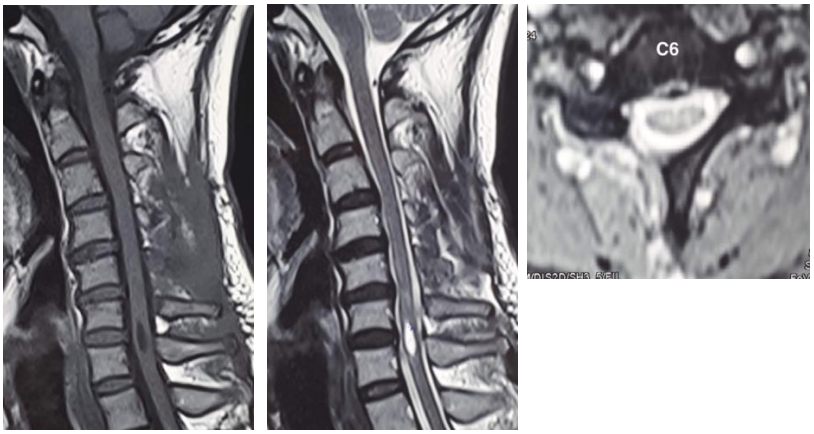

Figure 4. Post-operation MRI showed complete spinal cord decompression but remaining syringomyelia after taking the blood clot out.

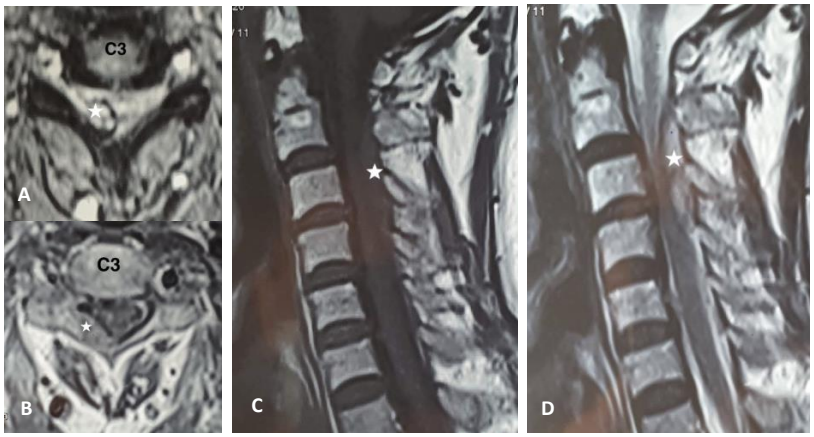

Figure 5. MRI of SSEH appeared epidural hematoma at the C2-C4 level (star), and revealed heterogeneous hyperintense on T2W image (A, D), homogeneous isointense on T1W image (C), and non-enhancing of the SSEH on T1W with contrast (B).

Table 1. Summary of Clinical and Imaging Characteristics and Management Strategies for the Two Patients

Patient	Gender	Age	Clinical Signs	Imaging Signs	Treatment	Outcome
1	Male	45	Sudden neck pain and quadriparesis, No history of trauma, anticoagulant or antiplatelet use	A biconvex mass in the epidural space that severely compressed the cord at the C5-C6 level and appeared isointense on T1 weighted (T1W) image and hyperintense on T2 weighted (T2W) image	Hemi-laminotomy	Walk by himself, Write and hold chopsticks
2	Male	57	Sudden neck pain with numbness in both arms for about one day without quadriparesis	A biconvex mass in the epidural space at the C2-C4 level on cervical MRI with the same feature	Conservative treatment	Complete neck pain relief and denied motor and sensory disorders

taking an antiplatelet (Clopidogrel 75 mg once a day). Our final decision was conservative treatment and to closely follow his neurological function. After five days, he had no neck pain or numbness in his arms. Cervical MRI was obtained ten days afterwards, which revealed almost the blood clots' entire resolution (Figure 6). After consulting the cardiologist about the risks of myocardial infarction unless using antiplatelet, we did not let him restart it. At the time of discharge, eleven days after onset, he demonstrated complete neck pain relief and denied motor and sensory disorders. His symptoms still remained unchanged at two months of follow-up. The summary of the cases are shown in Table 1.

Discussion

Epidemiology

Spinal epidural hematoma is an uncommon disorder, possibly causing death if it is late-diagnosed or left untreated.[8-10] Spinal epidural hematoma was first described by Jackson in 1869[11] and was first operated by Bain in 1897.[12] The incidence is estimated to be 0.1% per 100 000 individuals per year. The invention of MRI has increased the number of newly diagnosed cases as well as its incidence.[7,13,14] This disease is most common in patients in their 1920s and 1970s.[3,14] Halim and colleagues showed that there was no significant correlation with sex and race.[15] SSHE occurs most frequently at C6 and T12 levels. The hematoma often extends from 2 to 4 (3.6 on average) vertebrae and is located posterior to the cord.[8]

Risk Factors

Trauma and surgery are two well-known predisposing factors of spinal epidural hematoma.[1] Nevertheless, there has been limited data about other risk factors of this entity. Many authors have suggested significant associations between SSHE with arteriovenous malformation, anticoagulants used, coagulopathy, vertebral hemangioma, and even hypertension.[1-3,16,17] However, a large study showed that the link between SSHE and hypertension was negligible.[3] Some authors reported up to 17%-30% of cases were related to anticoagulant use.[16-18] Others have listed minor trauma, pregnancy, hemophilia, and leukemia as potential etiologies of SSHE.[16,17] Nevertheless, up to 40-60% of cases are idiopathic. Our first case is of clinical scenarios that could not identify any known cause or risk factor. In contrast, the second case has a history of using antiplatelet.

Pathophysiology

Cervical epidural hematoma often presents in a spontaneous and acute deficit, while hematomas occurring at lower spinal levels tend to progress in a subacute or chronic fashion.[8] The bleeding sources, according to current literature, are from the venous and arterial systems.[3,4,8] However, the former is more widely considered as the origin because the epidural veins lack elasticity, making them more vulnerable to the blood pressure changes. The spinal epidural plexus that drains the abdomen and thorax is low pressure and valveless.[17]

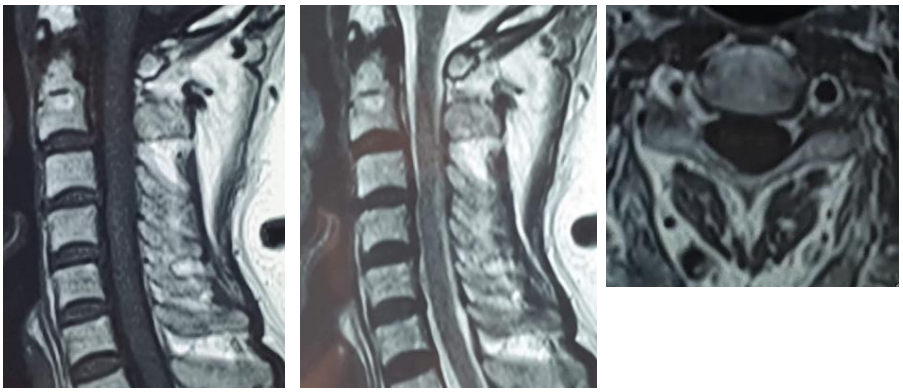

Figure 6. The second MRI showed that the hematoma at the C2-C4 level was almost entirely absorbed ten days afterward. Only thin layer of blood clot was withnessed on these images (arrow).

Thus, some authors have theorized that the increased thoracic and abdominal pressure is transmitted to these valveless veins and thin-walled layer veins in the epidural space, resulting in the rupture. This may explain why SSEH often occurs after stretching, coughing, and sneezing.[4] However, this theory seems not to apply to the cervical area because of its low-pressure venous system. Cervical epidural hematoma originates from free anastomotic arteries connecting with radicular arteries, which run within the epidural space.[3] SSEH has been seen more commonly at the dorsum of the epidural space than the ventral, which has a lower risk of rupture because of its smaller size and the support of the posterior longitudinal ligament.[2,5,17] The higher prevalence of SSEHs in the middle-age population could be explained not only by a change in the vascular component but also by the cumulative effect of gravity, which dilates the vessels.[3] Other studies have shown that flexible spinal segments apply a higher mechanical stretch pressure onto the epidural veins,[17] explaining the higher occurrence rate of SSEHs in cervicothoracic and thoracolumbar regions.[1-3,17]

Clinical Presentation

SSEH has a clinical presentation similar to transverse myelitis, carotid aneurysm rupture, ruptured cervical herniated disk, epidural neoplasia, or infection (e.g., epidural abscess). The signs and symptoms, which consist of severe pain and neurological deficits. Most patients present with back and neck pain with radiculopathy.[1-3,14,17] The clinical symptoms of SSEH depend on the hematoma location by which patients could present with hemiparesis, hemiplegia, quadriparesis, quadriplegia, and other presentations.[3,14] The typical course usually consists of sudden and abrupt pain, followed by motor and sensory deficits. The severity and the progression of sensory and motor deficit depend on the bleeding speed and extent of the lesion. Severity has a prognostic value. Patients whose partial neurological functions after the onset are more likely to make a full recovery than those with no motor function.[6-8] It has been reported that 37% of all SSEH cases showed a complete loss of sensory and motor function, while the remaining cases preserved some sensory or motor function. Both cases had neck pain and neurological deficits at different levels. The patient who had a more extensive hematoma had worse spinal cord disorder, and his clinical presentations were also more severe.

Imaging Study

The age of the hematoma is determined by computed tomography (CT) scan findings. The hematoma is biconvex and hyper-condensed in the acute phase.[3] Sometimes, it is challenging to differentiate lesions from lymphoma or epidural abscess without using contrast. In those cases, MRI is the appropriate next step in the diagnosis. Within the first 24 hours of the onset, an epidural hematoma appears isointense to the cord on T1W images and usually hyperintense or isointense homogeneous on T2W images. Within the first 48 hours, the hematoma shows hyperintense on T1W and T2W images.[1,14,16,17] Chronic hematoma is hypointense on both T1W and T2W images.[1] Short tau inversion recovery sequences may be used to distinguish between the hematoma and epidural fat.[17] Occasionally, actively bleeding hematoma may appear enhancing in the center of the lesion with contrast.[17] Some authors have noted that the hematoma, in several cases, has some contrast-enhancing characteristics, which is probably related to the hyperemia of the dura mater or hypertrophic meninges.[17] In our cases, MRIs showed isointense on T1W and hyperintense on T2W even though these images had been taken more than 24 hours since the onset of neurological symptoms.

Treatment

Conservative Management

According to several studies, some SSEH cases have been successfully treated with conservative management. All these patients only presented with minimal neurological deficits on the initial examination and made significant improvement afterward.[6,15] Based on one report,[19] 84% of all patients in this study had completely recovered. However, this conclusion cannot hold for all SSEH cases because most of this study's patients had modest symptoms and could be diagnosed via imaging studies alone. Their lesions were extremely small in size resulting in bias. Mild cases without evidence of a mass could be managed conservatively; nonetheless, neurosurgeons always have to keep in mind that SSEH could progress unexpectedly whenever there is a mass effect on the spinal cord. In several cases, the neurological symptoms have been worsened in the recovery phase and should be treated surgically. A spontaneous self-regression in SSEH happens when a hematoma spreads throughout the epidural space to the point that is alleviated.[8] Conservative management would be an appropriate choice for young patients with mild neurological signs or in those whose symptoms have already resolved spontaneously before examination. If conservative treatment has been selected, patients should undergo a close observation followed by serial neurological examinations and a mandatory early MRI. Patients who present with any worsening clinical condition or new neurological signs should be immediately managed with a surgical intervention.[3,4,8] Our second patient was managed conservatively because he only had slight dysesthesia without motor function disturbance. Simultaneously, we also clearly explained to this patient the sequence of this disease and closely followed his clinical presentations.

Surgical Management

Surgical options for SSEH patients are usually hemi-laminotomy or laminectomy with evacuation and removal of a hematoma. Coagulopathy, if present, should be well-controlled before undergoing any surgical procedures.[4,17] There are three studies assessing in detail the surgical outcomes of SSEH.[9,10,20] Factors significantly affecting the postoperative recovery process include the anatomic level of a lesion, the severity of neurological deficits, and the operating time. The results are more desirable in cases with partial damage than those with a complete neurological lesion.

According to Liao and colleagues,[10] the rate of full recovery after one year was 88.9% in patients with partial deficits but only 37.5% in those whose neurological function was lost entirely. A similar conclusion has also been found in other studies.[20]

As soon as the diagnosis of SSEH has been established, emergent surgery is indicated.[2,4,21] Some authors argue that patients should be surgically decompressed within 12-48 hours of the onset of clinical symptoms to maximize the neurological outcome.[2,7,17] However, several studies have failed to provide a statistically significant difference in the end result when the reports depend solely on the time from symptom onset to operating room.[21] Many authors suggest that the decompression procedure should be done within 24-36 hours for complete neurological deficit and within 48h for incomplete deficit.[5,17] Despite these controversies, the long-term outcome relies on preoperative neurological deficits. This is the most crucial prognostic tool.[2,3,16,21] Our first patient had a Frankel B neurological lesion on the right and a Frankel C on the left; he underwent surgery 36 hours after the onset of symptoms. There was an outcome difference between these two sides. The side classified as more severe damage (Frankel C) had a longer and more inefficient recovery time. This is consistent with the literature.

Patients presenting with symptoms that rapidly worsen over a shorter period of time usually have a larger hematoma size; they achieve related to a more inferior result, especially with lesions involving over four vertebrae.[3,15] In addition, patients with a little residual sensory function show a worse outcome when compared with those with a mild sensory loss.[3] Spinal regions with a limited room for the cord (e.g., thoracic region) restrict expansile capacity of a hematoma and, in turn, worsen the prognosis.[3]

The etiology of neurological deficits may not only be associated with compression from hematoma but also secondary to inflammatory reactions.[3] Therefore, even if patients are classified as American Spinal Injury Association (ASIA) score A or admitted to a hospital late have to undergo surgery.[4,21] Many studies have indicated that some patients with ASIA A could even improve their status up to ASIA E after surgical intervention.[4,16] The recovery tends to be more remarkable in pediatric patients.[4]

Complication and Mortality

Postoperative mortality depends on whether patients suffer a complete or partial loss of neurologic function. Those with a complete lesion have a higher mortality rate.[8] In a patient group of Lawton, 3% of patients died because of surgical interventions.[9] In a group of 35 patients, the mortality rate related to SSEHs was 5.7%, the adverse events rate was 2.9%, and surgery-related deaths was 0%.[10]

Conclusion

In summary, we can conclude that SSEH is a surgical emergency because of its rapid progression and potential for causing permanent neurological sequelae. Therefore, early surgical management leads to a better outcome. The preoperative location of an injury, the severity, and the operating time crucially determine the result of surgery. Controversial treatment would be reasonable if the patients have a mild clinical presentation. Although some risk factors have been suggested to serve as potential etiologies of SSEH, the actual underlying cause remains unclear. Our first case, as well as other reports, cannot ascertain the causation of SSEH. Because of its rare occurrence, research to uncover the root cause encounters several challenges. Thus, more studies with large numbers of patients are required to elucidate the disease.

Conflict of Interest Disclosures

The authors declare that they have no conflict of interests.

Ethical Statement

Informed consent was obtained from patients and their families for the publication of this report.

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