ABSTRACT

Spontaneous spinal epidural hematoma (SSEH) is a rare emergency with potentially serious sequel. We report a case of SSEH to emphasize the importance of early recognition and urgent surgery for this challenging emergency. A 10-year-old boy presented with gradual onset of neck and back pain, vomiting, and nuchal rigidity followed by rapidly progressive paraparesis, abdomen and lower extremities hyperesthesia and sphincter dysfunction. MR imaging demonstrated epidural hematoma of cervico-thoracic spinal segments (C7-T3). Emergent decompressive laminectomy with hematoma evacuation was performed the third day after paraplegia onset. Neurological improvement was obvious but slow, and the patient was discharged walking with aid 10 days after surgery. Two months later he got full functional recovery. We conclude that SSEH is a rare but disabling or even fatal clinical challenge. Early diagnosis and prompt surgery improve the neurological and functional outcome. Initial nonspecific symptoms can lead to a delay in diagnosis, especially in younger children. Relevant physicians should pay attention to the symptoms of this rare entity and SSEH should be one of differential diagnoses of rapidly evolving spinal cord syndrome.
INTRODUCTION

After Jackson reported the first case of spontaneous spinal epidural hematoma SSEH in 1869,1 SSEH became a well-known though rare cause of spinal cord compression in the emergency department. The estimated incidence is about 1 patient per million individuals and represents less than 1% of spinal space-occupying lesions.2 The underlying cause cannot be identified in more than 40% of the cases.3,4 However, many underlying diseases, such as coagulation disorders, vascular malformations may be the cause.5-8 Association with spine diseases such as ankylosing spondylitis6,10 and Paget’s disease11 has also been reported. SSEH usually presents with neck or back pain followed by acute spinal cord compression syndrome. Only about 30 pediatric cases of SSEH have been documented in the medical literature.8,12 We report a pediatric case of SSEH and review the relevant literatures.

CASE PRESENTATION

A previously healthy 10-year-old boy was referred to the emergency department (ED) with rapid onset of upper back pain, occipital headache and sharp pain radiating to the right upper limb. There was no recent history of trauma or infection. He was so agitated and had misleading manifestations such as abdominal pain, vomiting and noticeable nuchal rigidity. Although he had difficulty with walking but he looked rather lethargic for lower limbs weakness was not as prominent. After admission to the pediatric department, investigations included LP and CSF analysis was normal except for mildly high protein level (82 mg/dl). Laboratory investigation included complete blood count, chemistry panel, and coagulation profile were all within normal limits. Next day he started to experience increasing lower limbs weakness and burning sensation and was unable to walk. Spinal MRI and neurosurgical consult was ordered.

On examination the patient was orientated, but looked irritated and disturbed with pain to a degree that made communication difficult. Abdominal examination revealed soft but dysesthetic abdomen, with distended bladder. He was afebrile and had unremarkable cardiovascular and chest examination.

Neurological examination revealed almost complete paraplegia, severe lower limbs hyperesthesia, and prominent nuchal rigidity. Impaired sensation below T1 level was noted. His reflexes were hypertonic and Babinski sign was positive bilaterally.

MR imaging demonstrated acute spinal epidural hematoma from C7 through T3 causing compression of the spinal cord. The hematoma was relatively isointense on T1-weighted images and hypointense on T2-weighted images (Figure1).

Emergent decompressive laminectomy was performed to remove relatively large epidural hematoma about 4 days after admission and 3 days after the onset of paraplegia. Intraoperatively there was
an abnormal artery on the dorsal surface of dural sac that caused problematic intraoperative bleeding which was controlled with isolation and bipolar coagulation. The hematoma was evacuated except for small anterior residue. Two days after operation sensation and urinary continence regained, but muscle strength recovery was very slow. The patient was discharged 10 days later walking with aid. Two months later he got full recovery of neurological function.

**DISCUSSION**

Spontaneous Spinal Epidural Hematoma (SSEH) is an accumulation of blood in the vertebral epidural space in the absence of trauma or iatrogenic procedure such as lumbar puncture. The location and onset age of SSEH have a bimodal distribution with the location peaks at C6 and T12 and onset age peaks at 15-20 and 65-70 years respectively.\(^{13,14}\) The male/female ratio is 1.4:1.\(^{3,15}\) Pediatric SSEH is extremely rare, with only about 30 cases of SSEH documented in the medical literature.\(^{8,12}\) Idiopathic cases account for at least 40% of all cases.\(^{6}\) However, certain precipitating factors are suggested to be correlated with SSEH. This could be divided into two categories. First; medications or disorders that alter normal coagulation such as anticoagulant therapy for prosthetic cardiac valve,\(^{5,7}\) thrombolysis therapy for acute myocardial infarction,\(^{7}\) uncontrolled hypertension,\(^{16}\) aortic coarctation,\(^{17}\) end-stage renal disease receiving hemodialysis,\(^{18}\) long-term antiplatelet usage\(^{19}\) and congenital diseases with factor XI deficiency or hemophilia B.\(^{20-23}\) Second; spinal diseases such as ankylosing spondylitis\(^{9,10}\) and Paget’s disease.\(^{11}\)

The origin of these hematomas is still debatable. Many authors accept the venous etiology hypothesis due to lacks of venous valves in epidural venous plexus.\(^{24}\) However, several authors have proposed the spinal epidural arteries as a source of hemorrhage,\(^{25}\) due to the fact that pressure from arterial bleeding is more likely to compresses the spinal cord, as intra-thecal pressure is higher than the venous pressure.\(^{26,27}\) Only rarely, SSEH associated with an identifiable underlying vascular lesion\(^{28}\) such as hemangioma\(^{29}\) or vascular malformation.\(^{30-32}\) In our case an abnormally tortuous dural artery was evident at surgery. Anyway, further studies are needed to clarify the precise pathogenesis of SSEH.

The usual clinical presentation of SSEH is sudden onset of severe neck and/or back pain. Motor and/or sensory deficits caused by compression of nerve roots and spinal cord follow and progress rapidly with sphincter incontinence. Despite the characteristic syndrome of SSEH, initial nonspecific symptoms can lead to a delay in diagnosis, especially in younger children. Unfortunately, children often present with nonspecific presentation and may suffer from additional symptoms of irritability, nuchal rigidity, and occasionally urinary retention.\(^{12}\) Furthermore, a recent case of SSEH in a 10-year-old girl is interesting in that she presented with what initially appeared to be a pure brachial plexopathy.\(^{33}\) In fact, in our case the abdominal pain, right shoulder pain, and vomiting were so deceiving so that hepatitis, cholecystitis and meningitis were all on top differential diagnosis.

Currently the introduction of MR imaging help making the diagnosis early and accurate, and define the location and size of hematoma. According to the age of hematoma the MR images of acute SEH reveal variable findings from isointense to hypointense to hyperintense on T1-weighted images and from hyperintense to hypointense on T2-weighted images. Beside specific signal changes, contrast enhancement pattern and morphological findings on MR images can differentiate acute SSEH from spinal epidural neoplastic mass or abscess.\(^{34}\) In addition, MRI may rarely demonstrate the underlying lesions, such as spinal vascular malformations.\(^{31,35}\) The differential diagnosis of spontaneous spinal epidural hematoma includes an acute herniated intervertebral disc, acute ischemia of the spinal cord, epidural tumor or abscess, spondylitis, transverse myelitis, or even a dissecting aortic aneurysm and acute myocardial infarction.

SSEH is generally surgical emergency with rapid hematoma evacuation being the most effective treatment.\(^{36,37}\) Conservative treatment is still an important option of treatment in some selected patients.
with mild and rapid spontaneous recovery or in cases that is highly risky for surgery.\textsuperscript{38} It is interesting in this regard to note that some non-surgical SSEH patients have been noted to improve following lumbar puncture; that may be due to reduction of intraspinal pressure via thecal sac puncture and cerebrospinal fluid egress which leads to partial cord decompression.\textsuperscript{39}

The prognosis of SSEH correlates with the size and level of hematoma, severity of preoperative neurological deficits and time interval between symptoms onset and surgery. Of these, the interval from symptoms onset to surgery appears to be the single most important factor.\textsuperscript{40} Recurrence is extremely rare and there is a case that documents recurrence of a cervical epidural haematoma 3 years after the first incidence.\textsuperscript{41,42}

\textbf{CONCLUSIONS}

Still remain a clinical challenge, SSEH is a rare and serious condition that can be managed with good outcome. Early diagnosis with MRI and urgent surgical decompression is the key. Relevant physicians should keep in mind SSEH as one of differential diagnoses of rapidly evolving spinal cord syndrome.

\textbf{REFERENCES}