Case reports

Spontaneous cervical epidural hematoma of idiopathic etiology: Case report and review of literature

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Context: Spontaneous spinal epidural hematoma (SSEH) is a rare idiopathic condition that leads to acute onset of neurological deficits, which if not recognized early can have catastrophic consequences. The definition and pathophysiology of this condition remain controversial. High index of suspicion followed by T2-weighted gradient echo sequences are particularly useful in early diagnosis. Management consists of prompt surgical decompression of the hematoma though a recent trend is toward non-surgical treatment.

Findings: A 70-year-old man presented with acute onset neck pain with a radicular component and rapidly progressive quadriparesis. Magnetic resonance imaging revealed a posteriorly located cervical extradural hematoma with cord compression that was promptly evacuated. Functional recovery to near normal function occurred within 24 hours of surgery.

Conclusion: SSEH in its true idiopathic form is a rare pathologic entity. Because of the high risk of poor outcome without treatment, SSEH should be a diagnostic possibility when presentation is even slightly suggestive. Prompt surgical evacuation of the hematoma leads to a favorable neurological outcome, whereas delay in treatment can be disastrous. The role of conservative management needs to be proven and should be tailored on an individual basis.

Keywords: Tetraparesis, Tetraplegia, Paralysis, Myelopathy, Laminectomy, Neurologic manifestations, Progressive, Spinal cord compression, Cervical, Hematoma, Idiopathic spontaneous spinal epidural hematoma

Introduction

Spontaneous spinal epidural hematoma (SSEH) is an uncommon condition that requires early recognition and treatment to ensure complete recovery of function. The exceedingly high morbidity of untreated SSEH, however, warrants its inclusion in the differential diagnosis of presentations suggesting spinal cord involvement. This condition is being increasingly recognized now that patients presenting with rapid progressive neurological deficits of spinal cord origin are undergoing early evaluation with magnetic resonance imaging (MRI). The definition of SSEH, however, still remains debated as different authors have used various criteria for inclusion of patients under this heading. Few authors have suggested a conservative approach in the management of these cases with subsequent

neurological recovery. We report a case of spontaneous cervical epidural hematoma in a patient without any previous predisposing factors and discuss the clinical presentation, pathogenesis, and treatment of this rare entity.

Case report

A 70-year-old male physician presented with acute onset neck pain with progressive tetraparesis of 36 hours duration. His symptoms started as severe pain in the nape of neck associated with radiation of pain along his left upper limb and difficulty in neck movements. Within a few hours he noticed mild left-sided grip weakness and also mild difficulty in walking. The next day the symptoms progressed and he developed tetraparesis with inability to walk and hesitancy of micturition.

His past medical history was unremarkable. He had no history of neck trauma, smoking, drinking, or drug use. The patient was neither on anticoagulation nor

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antiplatelets at the time of presentation. On neurological examination, power in both upper limbs was 4/5 (Medical Research Council grading) with significant left-sided grip weakness. Motor power in the lower limbs was 2/5 at the hip and knee joints and 4/5 at the ankle. There was bilateral lower-limb hypotonia with absent reflexes and Babinski sign was negative. Sensory examination showed absent vibration sense below C6 level and impairment of joint position sense in both feet.

An urgent MRI of the cervical spine revealed a well-defined dorsal extradural lesion at C5–C7 levels which was iso/hyperintense to the cord on T1 weighted (T1W) images (Figs 1A and D) and hyperintense with a band of hypointensity on T2 weighted (T2W) images (Figs 1B, C and E). Poorly delineated hyperintensities were noticed within the spinal cord at this level. The lesion showed blooming on T2W gradient echo sequence (Fig. 1F) displacing the spinal cord anterolaterally toward the right side. There was straightening of the cervical spine with posterior marginal osteophytes at multiple levels. His initial laboratory workup failed to show any significant abnormalities and coagulation profile was normal.

Given the acute presentation and the MRI findings, a diagnosis of spontaneous cervical epidural hematoma causing cord compression was suspected. The patient underwent an emergency C5–C7 laminectomy in the

prone position. A well-circumscribed clot was seen in the corresponding epidural space which was removed and the pressure on the cord was relieved. No other pathology was detected intraoperatively. The histopathology of the clot was suggestive of an organized hematoma and there was no evidence of any abnormal vessels on elastin Van-Gieson stain. Postoperatively, the power in all the limbs improved to grade 4/5 within 24 hours. At 1-month follow-up the patient was ambulatory with mild residual left grip weakness. Postoperative films revealed adequate cord decompression (Fig. 2), which confirmed our clinical findings.

Discussion

Epidemiology

Spinal epidural hematomas were first described by Jackson in 1869¹ and first treated surgically by Bain in 1897.² The incidence of SSEH is estimated at 0.1/100 000/year.³ Following the introduction of MRI into standard medical practice the mean incidence of SSEH cases reported in the literature has further increased.⁴ But whether all cases reported in the literature qualify the diagnosis of SSEH is questionable since there is controversy regarding the accurate definition of SSEH. Spontaneous hematoma is most often defined as a hematoma occurring in the absence of any trauma or iatrogenic procedure.⁵ It does not

Figure 1 Axial T1W (A) and T2W (B) MR images showing the extradural hematoma compressing and displacing the spinal cord. Sagittal T1W (D) and T2W (C,E) images showing the clot situated dorsally at the level of C5–C7 vertebral bodies. Sagittal T2W gradient echo sequence (F) demonstrating blooming within the lesion.

Figure 2 Axial (A) and sagittal (B) T2W MRI showing postoperative changes in the form of laminectomy at C5–C7 levels. There is no evidence of any residual hematoma or cord displacement. On contrast study (C) there is no evidence of any pathological enhancement.

exclude causes such as coagulopathy, vascular malformation, cavernous angioma, and tumor. But few authors describe a hematoma as spontaneous when it is of idiopathic origin⁶ and only these cases should be considered under the heading of SSEH. In 40–50% of cases of spinal epidural hematoma no cause is found and a diagnosis of SSEH is appropriate.⁷

The location of SSEH appears to have a bimodal distribution, with peaks at C6 and T12 levels.⁸ Most hematomas measure approximately 3.6 vertebral levels in length, and are located dorsal to the spinal cord. This is supposedly due to the presence of Hoffmann ligaments connecting the ventral dura to the posterior longitudinal ligament.⁹ Age also shows a bimodal distribution, with peaks at second and seventh decade.¹⁰ There is no significant correlation with race or gender.¹¹

Pathophysiology

Spinal epidural hematoma of the cervical spine is usually spontaneous and of acute onset while hematomas occurring at the lower levels tend to have a subacute or chronic course. In the latter, the cause of bleeding is more likely to be defined. Current literature supports both venous¹² and arterial origins as to the source of spontaneous epidural hematomas. The most widely accepted hypothesis for source of bleeding is venous, owing to the fact that spinal epidural veins have no valves and are thus unprotected from changes in abdominal or thoracic pressure.³ According to the hypothesis proposed by Bruyn and Bosma¹³ increased intrathoracic and intra-abdominal pressure leads to brief increase in intravenous pressure in valveless and thin-walled epidural veins subsequently leading to

their rupture. This accounts for the cases reported to have occurred with activities such as straining, bending, coitus, coughing, sneezing etc. In a recent case series by Liao *et al.*,¹⁴ 54% of patients reported a straining-associated event during the initial attack, further supporting the venous etiology theory. However, in the cervical region, owing to low venous epidural pressure (even lower than intrathecal pressure), this venous theory does not seem valid. Also, the rapidity of development of a cervical epidural hematoma points toward its arterial origin. According to Beatty and Winston's hypothesis,⁶ the source of arterial bleeding in cervical epidural hematomas is free anastomotic arteries running in the epidural space that also connect with radicular arteries. The high mobility of the C6–C7 cervical spine accounts for this common location of cervical epidural hematoma.

Clinical presentation

SSEH can have a wide spectrum of presentation ranging from simple cervical radiculopathy to acute quadriplegia depending upon severity and rapidity of compression. Most patients present with severe back and/or neck pain, often with a radicular component.¹³ The pain usually mimics the pain of a disc prolapse. The pain is usually followed by progressive motor or sensory deficits depending on severity and rapidity of bleed. Approximately 37% of patients present with complete sensorimotor deficits, while the remainder have some sensory or motor function intact. This has prognostic significance, as those with some residual function are more likely to show complete recovery than those with no motor function.^{8,13–17}

Diagnosis

The investigation of choice is MRI. In the first 24 hours, an epidural hematoma is isointense to the cord on T1W images and is usually hyperintense, although it may be heterogeneous, on T2W images. By 48 hours the hematoma appears hyperintense on both T1W and T2W sequences.^{3,9} A negative scan once the blood has resolved negates the need for a spinal angiogram although small, low flow vascular malformations could escape detection on MRI. The radiological differential diagnosis includes epidural abscess and spinal epidural lymphoma.

Treatment

Conservative management

There are an increasing number of reports of individual cases managed conservatively.¹⁸ According to Groen⁸ 84% of patients treated non-operatively recovered completely. However, this conclusion cannot be extrapolated to all cases of SSEH, as many of his patients had less severe signs and symptoms and were more likely to be diagnosed based on imaging alone. Their lesions were significantly smaller in size thereby biasing the results. Holtas *et al.*³ reported 11 of 13 patients treated conservatively with good outcomes. Although some patients with minimal symptoms and no mass effect on spinal cord may be managed successfully with non-operative treatment, the treating surgeon should always remember that the outcome of SSEH can be very poor when mass effect is exerted on the spinal cord. Conservative treatment usually results in a poor outcome, particularly when the hematoma compresses the cervical cord. Cases have been reported where recovery was followed by deterioration requiring surgery.¹⁸ Observation in a hospital without neurosurgical services is therefore inappropriate.

The reason for spontaneous recovery in some patients with SSEH is spread of hematoma throughout the epidural space, thereby decompressing the spinal cord causing decrease in the mass effect.^{8,19} The decision of conservative management is always fraught with risk of sudden increase in the size of epidural hematoma and deterioration of symptoms thereby making the ultimate outcome worse. Non-operative treatment and masterly inactivity can be a reasonable option in younger patients presenting late with minimal deficits on neurological examination or in those patients whom spontaneous recovery has already started at or before medical evaluation. Conservative treatment protocol should involve close observation in a neurosurgical unit with early repeat MRI. Surgical intervention may still be necessary if symptomatic recovery with hematoma resolution does not continue.

Surgical management

There are three major case series giving a detailed review of outcome of surgical management of SSEH.^{14–16} The factors significantly affecting postoperative neurological recovery are the level of preoperative neurological deficit, severity of the neurological deficit, and operative interval. The outcome is better with incomplete deficits, as compared to complete deficits. According to Liao *et al.*,¹⁴ 1 year of complete recovery was 88.9% for incomplete deficits but only 37.5% for complete deficits. Similar results have been found by other authors.^{8,16}

As far as operative interval, earlier the intervention better is the outcome. In a series of 35 patients by Liao *et al.*,¹⁴ chances of complete recovery were significantly improved when patients with complete and incomplete deficits were operated on within 36–48 hours, respectively. Many other authors have also emphasized on a significant relationship between surgical timing and neurological outcome. The latter is inversely related to the time interval between symptom onset and surgery.^{7,8,15,16} In a series of 14 patients by Shin *et al.*,¹⁶ the patients who were operated on less than 12 hours after onset of symptoms scored significantly higher (84%) on the recovery scale of the Japanese Orthopedic Association than those who underwent surgery from 12 to 24 hours postsymptom onset (63.6%) and those who underwent surgery after 24 hours (46.7%).

Complications and mortality

The postoperative mortality depended on whether or not the deficit was complete or incomplete; patients with complete deficits having a higher mortality.⁷ In Liao's series of 35 patients, the disease-related mortality rate was 5.7%, and the complication rate was 2.9%.¹⁴ In Lawton's series, the overall mortality of surgical treatment was 3%.¹⁵

Conclusion

The diagnosis of SSEH should be made only if the etiology is truly idiopathic without any predisposing factors. Keeping this criterion, many of the earlier-reported cases in the literature may have been wrongly misdiagnosed as SSEH. This entity in its true form is a rare pathologic entity usually presenting with non-specific pain, progressing rapidly to sensory and motor deficits. High index of suspicion followed by T2W gradient echo sequences are particularly useful in the early stages due to their sensitivity for the detection of deoxy-hemoglobin. It should be treated with urgent laminectomy and evacuation of hematoma as any delay in surgical treatment can be catastrophic. Outcome

depends on time to operation and prognosis is affected by the severity and level of preoperative deficit. Conservative management should be tried in patients presenting with axial pain and radicular symptoms but without neurological deficits. Rapid, appropriate treatment of these patients can often lead to complete recovery with excellent neurological outcome.

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