

Acute Quadriplegia Caused by Idiopathic Spontaneous Epidural Haematoma in Young Adult-Case Report and Review of Literature

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Abstract

Introduction: Authors tried to emphasize the importance of diagnosing and rapidly treating spontaneous spinal epidural hematoma (SSEH), a rare idiopathic condition which if not recognized early can have catastrophic consequences. The definition and pathophysiology of this condition remains controversial. Usual management consists of urgent MRI imaging followed by immediate surgical decompression.

Clinical presentation: A 22 year young adult presented with the symptoms of sudden severe chest pain, followed by weakness and numbness over the all four extremities, progressing to intense paraparesis, anesthesia and consecutively quadriplegia and sphincter control loss. MR imaging demonstrated acute epidural hematoma of cervico-thoracic spinal segments (C7-Th2) with cord compression. Idiopathic SSEH was diagnosed and prompt decompressive laminectomy with hematoma evacuation was performed. During the operation cause of bleeding was not identified. Postoperatively CTA and DSA were performed which came back negative. After 2 weeks, there was no residual weakness/parasthesias, sphincter control was fully obtained and full motor power was required.

Discussion: SSEH in its true idiopathic form is a rare pathologic entity which can be disabling or fatal. According to the literature, most patients present with severe back and/or neck pain, often with a radicular component, followed by motor and/or sensory deficits which are symptoms to be aware of. Prompt diagnosis should be made with MRI, and evacuation of the hematoma should be immediate, ideally before the onset of neurologic signs or symptoms. As evidenced outcome depends on prompt operation timing and prognosis is impacted by age and preoperative deficit. Because of the high risk of poor outcome without treatment, SSEH should always be a diagnostic consideration in patients whose presentation is even slightly suggestive. Rapid, appropriate surgical treatment can often lead to complete recovery of function, whereas any delay in appropriate treatment can be disastrous. The role of conservative management needs to be proven and should be tailored on an individual basis.

Conclusion: SSEH is a rare and potentially fatal disease. Prompt diagnosis and surgical treatment is required to obtain good outcome results. Physicians should be aware of the early symptoms so the best treatment can be achieved.

Keywords: Quadriplegia; Paralysis; Laminectomy; Neurologic manifestations; Spinal cord compression; Spontaneous spinal epidural hematoma; Magnetic resonance imaging

Introduction

The first case of spontaneous spinal epidural hematoma (SSEH) dates back to the year of 1869 and the first operation of SSEH to 1897 [1]. It is still considered to be a rare, dangerous and very important cause of spinal cord compression [2]. The incidence of SSEH stands at 0.1 patients per 100,000 patients currently accounts for less than 1% of spinal space lesions [3]. The common symptoms of SSEH are: sudden neck/chest pain which is rapidly followed by nerve root and spinal cord compression signs (paresis and paralysis) [4]. Positive outcome is closely related to the quick diagnosis and treatment, usually surgical decompression [5]. The authors present a rare case of idiopathic SSEH that occurred in the cervical-thoracic spinal segments (C7-Th2) and review the related literature.

Case Report

A 21-year-old male experienced sudden severe sharp chest pain and limb paralysis that followed shortly after so he was brought to the emergency room (ER). The symptoms occurred during sleep and developed rapidly. There was no history of spine related injuries or positions that are known to be prone to put stress on the spine. No positive past history of head and spinal trauma, smoking, drinking or history of vascular problems (personal or familiar) was obtained. He had no diseases and did not take any antiplatelet or anticoagulant medication. On the admission, the patient was conscious, orientated, and communicative. The vital signs were within normal range, Glasgow Coma Score (GCS) was 15. The examination of head, neck, chest, cardiovascular and abdomen was unremarkable. Neurological status showed that the quadriplegia in all four limbs was present. Acute urinary retention developed after the admission and Foley catheter was placed. Laboratory investigation including complete blood count, chemistry panel, and coagulation profile was within normal limits. Computer tomography (CT) and Magnetic resonance imaging (MRI)

showed acute compressive posterior spinal epidural hematoma from C7 to Th2 vertebra causing compression of the spinal cord and nerve roots. MRI imaging showed relative iso-intense lesion on T1 sequence (Figure 1) which was hyperintense on T2-weighted images (Figures 2 and 3). In the first 12 hours since the symptoms onset, urgent operation was performed-decompressive laminectomy (from C7 to Th2) with the removal of the epidural hematoma. No pathological blood vessels or hemorrhagic tumors have been noted during the operation. Postoperatively, muscle strength recovered well (4/5 grade) in all four limbs with return of the bladder sphincter control. After physical rehabilitation full muscle power was recovered in every limb. Post-operative follow-up CT of cervical and thoracic spine revealed adequate cord decompression. Intra-hospitally both CTA and DSA were performed and both came back negative.



Figure 1: T1-weighted sagittal magnetic resonance image: Iso-intense mass compressing the posterior aspect of the spinal cord from C7 to Th2.



Figure 2: T2-weighted sagittal image shows a longitudinal hyperintense epidural haematoma ranging from C7 to Th2.

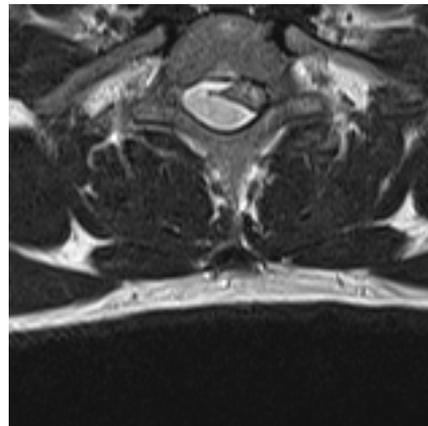


Figure 3: T2-weighted axial image shows an ovoid high-signal intensity epidural haematoma in the right postero-lateral side with spinal cord compression.

Discussion

No strict definition exists about spontaneous spinal epidural hematoma (SSEH). It can be summarized as the accumulation of blood in the vertebral epidural space without trauma or iatrogenic procedure performed (e.g., lumbar puncture) [6]. Coagulopathy [7,8], vascular malformations [9,10] and hemorrhagic tumors are considered to cause SSEH by some authors [11]. The others represent the stand that only those SSEH that are purely idiopathic can be labeled as spontaneous [12,13].

Around 50% (40-60%) of SSEH account for idiopathic according to some authors [14]. Most of hematomas occur between C6-Th12 levels of spinal canal while the age of onset has a bimodal distribution, peaking at 15-20 and 65-70 respectively [1]. Females are more prone to SSEH and the gender ratio is (Male-Female) is 1.4:1 [3].

Risk factors associated with SSEH are proven to be are: anticoagulant therapy and thrombolysis therapy for cardiac related diseases [15-17], non-regulated hypertension [18], hemodialysis dependent renal disease [19], chronic antiplatelet drugs usage [2,15,17] and some congenital diseases with factor XI deficiency like hemophilia B [7].

Sudden, intense neck/back pain followed by motor and sensory deficits in the subsequent hours are the usual symptoms of SSEH [1,3,20,21]. In some cases sphincter disorders may develop [22]. Despite the abovementioned signs, fast and precise diagnosis still represents a challenge for almost every physician [23].

The introduction of CT/MRI diagnostic devices greatly improved swift and accurate diagnosis. The location, extent, edema and severity of spinal cord compression of the hematoma were easier to diagnose and so was to determine the treatment plan [24]. SSEH is usually seen as isointense or hypointense lesions on T1-weighted images and hyperintense on T2-weighted images in the early MRI imaging. MRI is also a valuable tool in differentiating acute SSEH from epidural neoplastic mass or abscess based on specific signal changes, contrast enhancement and anatomical/pathological findings [2,24,25].

Authors support the theory that pathogenesis of SSEH is closely connected to both arterial and venous blood vessels [26]. Lack of venous valves in epidural veins can be one of the explanations for the hematoma formation, as the acute increase of intra-thoracic or intra-abdominal pressure can result in vessel rupture and bleeding [26,27].

A history of straining shortly before the first symptoms was reported in more than 50% of the patients [28]. The pressure in venous plexus being lower than epidural space could be the reason for epidural artery rupture according to some scientists. In order to clarify the etiology of SSEH further clinical research is needed [1,3,26,28].

Best treatment of SSEH at this moment is urgent surgical decompression-laminectomy with hematoma evacuation [29,30]. Patients with elevated surgical risk, heavy cardiovascular disease, irreversible spinal cord lesion or severe systemic disease should be treated conservatively at first [15,31,32]. Contrary to the previous sentence recent paper claims that the outcomes of conservative approach are very poor, especially in the cervical leveled lesions [30,33]. Post-OP mortality of SSEH ranges from 3% to 6% [1-3]. Size and location of hematoma, pre-operative neurological deficits and timing of the surgical decompression is directly connected to the prognosis [23,30,33].

SSEH usually ranges between 2 to 10 vertebral segments and longer hematomas have the worse outcome [1,6,14]. A group of authors report that the larger size of SSEH is more connected to the poor post-operative recovery [11,26].

As mentioned, surgical outcome is without a doubt closely related to the prompt and rightly timed surgery. If the operation is performed within first 48 hours from the onset of neurological deficits, prognosis of good recovery is greatly improved [11,30]. A group of Japanese authors published a scale which mentions the percentages of 83/63.6/46.7% for the operated patients in less than 12/24/48 hours from the first symptoms [25]. Lawton argued that surgery in the first 12 hours should be the benchmark for the good neurological outcome [34]. Another set of authors identified a close relation to permanent spinal cord neurological deficit in patients who were operated later than 12 hours from the symptoms onset [33]. Therefore, the stand is that surgery in the first 12 hours is essential for positive functional recovery in operatively treated patients with SSEH [5,33].

Conclusion

SSEH despite being very rare can be dangerous and sometimes fatal disease. Prompt diagnosis and quick management are essential to the outcome and therefore still represent a clinical challenge. Typical symptoms and signs of SSEH should be understood and recognized by every physician in order to preserve time, which is crucial for the treatment of these lesions.

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