Spontaneous Spinal Epidural Hematoma: a descriptive systematic review

Hematoma Epidural Espinhal Espontâneo: uma revisão sistemática descritiva

João Marcos Alcântara de Souza
Sávio Batista
Stefan Koester
Ghaspar Francisco
Mateus Barbosa
Rafaela de Souza
Roberth Fernandes
Raphael Bertani
Caio Moreno Perret
Othavio Lopes
Gustavo Pina dos Santos
Dan Zimelewicz Oberman

ABSTRACT
Spontaneous Spinal Epidural Hematoma (SSEH) is rare, with unknown etiology and devastating potential. Also, it is a pathology of difficult diagnosis, the exact knowledge of signs and symptoms is fundamental to allow a good prognosis. A systematized review of the literature was performed selecting articles published until 2021 in Pubmed, Embase, Web of Science, and Scopus databases about SSEH resulting in 2325 clinical studies related to the main diagnosis, and its etiologies, treatments and outcomes were described. A thorough search of these articles suggests that vascular malformations, anticoagulant therapy, neoplasms, and previous spinal surgery are the most common causes of SSEH and the main diagnostic method of SSEH is a thorough patient history and imaging studies, mainly MRI. Moreover, surgical treatment is indicated for most of cases, consisting of surgical decompression and hematoma evacuation, with conservative treatment recommended for fewer, selected cases with pieces of evidence suggesting that surgical timing influences the prognosis.

Keywords: Spinal epidural hematoma; Spinal hematoma; Spontaneous hematoma; Spontaneous spinal hematoma; Spontaneous spinal epidural hematoma

RESUMO
O hematoma epidural espinhal espontâneo (HEEE) é raro, com etiologia desconhecida e potencial devastador. Além disso, é uma patologia de difícil diagnóstico, sendo fundamental o conhecimento exato dos sinais e sintomas para permitir um bom prognóstico. Realizamos uma revisão sistemática da literatura selecionando artigos publicados até 2021 nas bases de dados da Pubmed, Embase, Web of Science e Scopus sobre HEEE resultando em 2325 estudos clínicos relacionados ao diagnóstico principal, em que foram descritas etiologias, tratamentos e desfechos. Uma busca minuciosa desses artigos sugere que malformações vasculares, terapia anticoagulante, neoplasias e cirurgia espinhal prévia são as causas mais comuns de HEEE. O principal método diagnóstico de HEEE é uma história completa do paciente e exames de imagem, principalmente a ressonância magnética. Além disso, o tratamento cirúrgico é indicado para a maioria dos casos, consistindo em descompressão cirúgica e evacuação do hematoma, com tratamento conservador recomendado para poucos casos selecionados com evidências, sugerindo que o tempo cirúrgico influencia o prognóstico.

Palavras-Chave: Hematoma epidural espinhal; Hematoma espinhal; Hematoma espontâneo; Hematoma espinhal espontâneo; Hematoma epidural espinhal espontâneo

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INTRODUCTION

Spontaneous spinal epidural hematoma (SSEH) is a rare, acute condition, with an incidence of 0.1 per 100,000/year. SSEH is caused by the accumulation of blood in the epidural space, which can mechanically compress the spinal cord. Since the first case of SSEH in the 19th century, more than 600 cases with various etiologies have been reported in the literature and, in cases where progressive neurological deficits appear, urgent neurosurgical evacuation of the hematoma and spinal cord decompression may be required. Comprehensive and broad reviews of this diagnosis are relatively lacking in the literature. Our aim is to conduct a thorough review of the literature in order to address the main scientific evidence on SSEH and summarize the main diagnosis, treatments, and outcomes of this disease.

MATERIALS AND METHODS

Our review was based on the guiding question: “what are the most relevant aspects in pathophysiology, diagnostics, treatment, and outcomes in relation to the spontaneous spinal epidural hematomas in the general population?” A comprehensive literature review was performed using Pubmed, Embase, Web of Science, and Scopus databases using as keywords: “spontaneous spinal hematoma” OR “spontaneous spinal epidural hematoma.” Information collecting included etiology, pathophysiology, clinical signs and symptoms, diagnosis, treatments, and outcomes.

Eligibility criteria considered involved all articles in English from current literature. Letters to the editor and commentaries were excluded from the review. Studies involving hematomas occurring after trauma, epidural anesthesia, lumbar puncture, or surgery, SSEH in association with subdural and/or spinal subarachnoid hemorrhage, or vertebrae or spinal canal tumors were excluded.

The search strategy revealed a total of 5551 initial articles, with 1612 being excluded due to duplications. A total of 3939 articles were selected by a reviewer (SB), excluding 2325. Two reviewers (SB and JM) performed the analysis and extraction of data following abstract and full-text of 1614 remaining articles, with disagreements of inclusion settled by a senior author (DZ).

RESULTS

Although etiology of SSEH has not been completely elucidated, a number of factors such as hypertension, anticoagulation therapy, straining, sneezing, lifting, coagulation disorders, cavernous angiomas, and spinal vascular anomalies have been hypothesized to predispose to SSEH. In a meta-analysis of 613 patients with spinal hematoma it was verified that in approximately a third of the cases, no etiological factor can be identified as the cause of the bleeding. Bleeding diathesis and arterial hypertension are conditions often associated with SSEH, however, the pathogenesis of those conditions remains obscure. Coagulation disorders, arteriovenous malformations, antiplatelet therapy, surgery trauma, and conditions causing increased intrathoracic-intra-abdominal pressure (coughing, Valsalva maneuver) may act as predisposing factors.

The pathogenesis and bleeding mechanism of SSEH are still unclear. However, two mechanisms for the development of SSEH have been considered in the literature: the rupture of epidural vessels, and hemorrhage from vascular anomalies. The most widely accepted hypothesis is that spinal hematoma is caused by rupture of epidural veins (venous bleeding). The pathophysiological mechanism involves a locus minoris resistentiae that ruptures upon transmission of increased intra-abdominal or intrathoracic pressure due to its connection with the valveless veins of epidural venous plexus. Since epidural veins are valveless, they have no protection against the variations of abdominal or thoracic veins’ pressure. Some researchers see the epidural venous plexus as an alternative path for blood amid the inferior and superior vena cava, which explains why there is a vascular bed that is much larger than would be necessary for the perfusion of the meninges alone. In cases of increased intrathoracic or intra-abdominal pressure, this alternate route is utilized. If such pressure variations operate on acquired or congenital abnormalities of vascular walls, a place of less resistance (locus minoris resistentiae), rupture can occur. Epidural veins’ rupture could happen during coughing, sneezing, defecation, coitus, straining (e.g. lifting of weights), pregnancy, and intrapartum in newborns. However, these insignificant physiological processes are numerous in daily life.
and the relative development of SSEH in these straining-associated activities is rare5,7.

Alternatively, another hypothesis is that SSEH may be originated from an arterial hemorrhage, since the venous plexus pressure is lower in the epidural space, theoretically insufficient to cause a hematoma, as proposed by Beatty and Winston, mainly at the cervical level12. Most SSEH are localized in the dorsal portion of epidural space and this supports the hypothesis of venous bleeding because the internal epidural venous network consists of a more prominent dorsal and a smaller ventral part, whereby the ventral portion is partially covered by the posterior vertebral longitudinal ligament and separated from the epidural space5.

However, the “venous hypothesis” will be unlikely to be applicable in the cervical region. Firstly, venous pressure is lower than intrathecal pressure13. Secondly, the cervical epidural hematoma usually produces rapid deterioration11. Anastomotic arteries that run in the epidural space and are also connected with radicular arteries are a possible origin for arterial bleeding13. The pressure of arterial bleeding is sufficient to compress the spinal cord, and such bleeding would likely not be tamponade by the counter-pressure of the thecal sac14. Prior literature has shown that the more mobile segments of the cervical spine (C6-C7) were the most common sites of hemorrhage, in which movement at this level might stretch the arteries beyond their limits, leading to rupture, thus making this a common location of spinal epidural hematomas13.

Spinal compression involving SSEH occurs due to the limitation of the hemorrhage by epidural fat tissue and by coagulation of the blood so that additional bleeding does not spread into the relatively large epidural space that measures 3 to 6 mm on average5. The interval between the initial lesion and symptom onset could correspond to the gradual collection of blood in the epidural space that will later cause direct compression of the spinal cord or secondary ischemic injury of the spinal cord due to compression of spinal blood vessels5.

Venous epidural hemorrhages are believed to be self-limited due to compression before they become mass lesions, which has led some authors to believe that there should be an arterial source for SSEH1. Moreover, most hematomas are located dorsolaterally and often present with radicular pain, which suggests a lesion in the radicular arteries, which are also located laterally and run along with the nerve roots5. The radicular arteries accompany the nerve roots into the spinal canal and may form longitudinal bridging arteries that could be easily injured by mechanical forces5. Since most hematomas are localized laterally (at the nerve root) and, similarly to epidural arteries, surrounded by a venous network, intraoperative observations would possibly support the hypothesis of arterial bleeding5. Beatty and Winston13 verified that their patients’ hematomas were predominantly lateral and surrounded by a net of epidural veins, where epidural arteries would be found.

Clinical presentation of SSEH is usually an acute and severe onset of pain in the location of hemorrhage that may be radiated to limbs, and sometimes there is radicular paraesthesia, similarly to vascular lesions13,14. Radicular pain may precede spinal pain, and subsequently, signs of the spinal cord or root compression ensue5,13,17. Symptom onset may vary from several minutes to several days5. Rarely, patients may present with slowly progressive, chronic, or relapsing symptoms or with neurological signs and symptoms that mimic an acute intervertebral disc herniation15. In most cases of SSEH, severe neck and back pain at the segment of bleeding is more prominent, but pain with radiation into the extremities is verified in many cases5. Hematomas in the superior cervical region are marked by onset with nuchal pain, while the hematomas in the inferior cervical spine present with interscapular pain5. The severity and progressiveness of sensory and motor deficit depend on the severity and rapidity of bleeding5.

In high cervical regions, SSEH could cause spinal shock, leading to fatal conditions. Hematomas occurring at the lower levels (thoracic and lumbar spine) tend to have a subacute or chronic course13,17.

Diagnosis of SSEH is made by prompt imaging exams and clinical suspicion1,18. Patients with back pain and acute or subacute neurological deficits with no apparent cause or secondary to minor trauma or a physiological effort should be suspected and an investigation for underlying coagulopathies and arteriovenous malformations (AVMs) should be performed. Differential diagnoses should include neoplasia, epidural abscesses, and lipomas18. SSEH on MRI scans may be seen as isointense lesions that progress to hyperintense on T1-weighted or heterogeneous hyperintensities on T2-weighted images with focal hypointense areas, seemingly associated with deoxyhemoglobin or too fibrous septa that attach to the dura to the walls of the spinal canal.
The latter finding can help to differentiate between SSEH and malignant lesions of the epidural space⁴.

Despite of common characteristics of T1-weighted images, some cases in the literature show atypical presentations on MRI, such as hypointensity and persistent isointensity for more than 48 hours. In these cases, clinical history is essential for differential diagnosis, and T2-weighted imaging seems to be of utmost importance for differential diagnosis, especially when patient history is confusing. Similarly, T1 contrast-enhanced images show peripheral enhancement in most cases, in sharp contrast with neoplastic alterations that usually show central enhancement⁸.

According to reports from the literature, surgical decompression and evacuation of hematoma will be necessary in the majority of patients⁷,¹⁰,¹⁴. Although in some cases, conservative treatment could be a feasible alternative⁹. As an acute or subacute condition with severe potential complications, SSEH with neurological deficits should be immediately surgically treated. It is worth noting that patients treated within 48 hours (if incomplete deficits) or 36 hours (if total deficits) have been reported to have better outcomes⁹. Some studies suggest the ideal timing for treatment to be within 12 hours of onset. This may suggest a narrower therapeutic window and increased necessity of celerity in diagnosis and management⁴,¹². As shown by Liao et al.⁷, operated patients had a complication rate of 2.9% and no deaths were reported in their series, compared to the disease-related mortality rate of 5.7%.

Interestingly, according to Groen et al.¹⁴, in 1996, the mortality seems to be higher in patients with cervical hematomas when compared to other segments (thoracic and lumbar segments) (p <0.05) and it was observed that a possible major cause of mortality in this series was thromboembolic events, such as pulmonary embolism and myocardial infarction, which may be secondary to the perioperative interruption of previous anticoagulant therapy.

Even so, there seems to be a growing trend in literature to adopt conservative treatment for SSEH. According to Groen et al.¹⁵, this effectiveness may correlate with increased use of MRI imaging for the diagnosis of SSEH, which may have led to higher diagnostic rates and patients with milder symptoms being more frequently diagnosed and treated conservatively. Another interesting point observed by Groen¹⁵, in 2003, is that patients with conservative treatment usually have longer-lasting symptoms than patients treated with operative treatment. This may be correlated to the “spreading theory” of spontaneous cure of SSEH, which hypothesizes that the hematoma spreads on the epidural space before being absorbed, which leads to spinal decompression and neurological amelioration¹⁵.

Surgical approaches remain the primary therapeutic intervention, with high rates of neurological recovery (88.9% complete recovery in patients with incomplete neurological deficit and 37.5% in those with a complete deficit¹⁵), being possibly replaced with conservative treatment in cases of a benign course with mild symptomatology and earlier improvement of sensorimotor symptoms¹⁵.

In patients who underwent surgery, the main factor related to the postoperative outcomes was the time since ictus to surgery, average Frankel grade, initial neurological deficit, size of the hematoma, and level of spinal cord compression. The average Frankel grade in neurological equivalents (5 as grade E, 4 as grade D, etc.) was 4.7 in patients treated before 6 hours, compared to an average grade of 3.7 in patients treated after 24 hours, and a decrease from 67% to 12% of complete recovery between these intervals¹. Complete recovery was seen in 65.9% of patients treated before 12 hours of onset and 64% of deaths, severe neurological outcome, or no improvement in symptoms in patients treated between 13-24 hours⁸. Approximately 25% of patients who had total recovery were Frankel Grade A and 83% of these patients were Frankel grade of D¹. Lower affected levels (thoracic and lumbar) and smaller hematomas were associated with better recovery of neurologic function¹ and cervical hematomas were associated with higher mortality rates¹⁰. Additionally, the use of anticoagulants was associated with a worse prognosis².

SSEH has an extremely low incidence in the population. Although studies suggest that its main etiologies may be vascular malformations, anticoagulant therapy, neoplasms, and trauma, remains unclear. There is evidence that cervical hematomas may carry higher mortality rates than hematomas in other spine segments. Regarding prevalence, there is no significant difference between genders and some studies suggest that it may be more common in people over 50 years of age. The primary diagnostic method is the combination of a well-documented medical...
history and magnetic resonance imaging. Furthermore, in most cases, surgical treatment is indicated. In a few selected cases conservative treatment might be an option. Future prospective studies are necessary to better understand the natural history, potential outcomes of this disease, and how treatment could be tailored for each case.

REFERENCES


CORRESPONDING AUTHOR

Sávio Batista
Medical student
Federal University of Rio de Janeiro
Faculty of Medicine
Rio de Janeiro, Rio de Janeiro, Brazil
E-mail: saviobatista360@gmail.com

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