Chapter

Spontaneous Spinal Epidural Haematomas

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Abstract

Spinal epidural haematomas (SEH) occur from extra-dural bleeding, most commonly after trauma, epidural anaesthesia or operative intervention. However, these also occur in arterio-vascular malformations or spontaneously without an obvious inciting event. Spontaneous spinal epidural haematomas (SSEH) are rare, with a quoted incidence of 0.1 cases per 100,000 population annually. However, these haematomas carry a significant risk of spinal cord or nerve root compression potentially resulting in permanent neurological dysfunction or death. This chapter reviews the presentation, diagnosis and treatment of SSEH in adults, pregnancy and children in order to provide clinicians with an understanding of their typical presentation, required investigations and treatment algorithms.

Keywords: epidural, haematoma, spine

1. Introduction

Spinal epidural haematomas (SEH) occur from extra-dural bleeding, most commonly after trauma, epidural anaesthesia or operative intervention [1, 2]. These haematomas are rare but carry a significant risk of spinal cord or nerve root compression potentially resulting in permanent neurological dysfunction and death [3, 4].

Spontaneous spinal epidural haematomas (SSEH) occur without an obvious inciting event, although they may be associated with vascular malformations of the spinal canal. These haematomas are rare with a quoted incidence of approximately 0.1 cases per 100,000 population annually [5–7]. When they occur they often mimic other conditions such as cerebro-vascular accidents [8, 9], which may make them a difficult condition to diagnose, resulting in a delayed diagnosis and potentially mismanagement with anticoagulation therapy [9, 7, 10]. However, the consequences of SSEH can be devastating with progressive neurological decline, permanent cord injury and even death [3, 11, 12]. Thus, early and accurate diagnosis and management is essential to preserve neurological function and optimise the chance of recovery [13].

The aims of treatment in SSEH are to restore normal neurological function with minimal risk to the patient, however the method of achieving this remains uncertain [13]. Surgical decompression, typically through a partial or complete laminectomy or laminoplasty, allows direct decompression of the neural elements and evacuation of the haematoma but carries both anaesthetic and surgical risk. Less invasive options include radiologically guided aspiration of the haematoma, however this also carries

risks of neural injury and further bleeding, often without complete evacuation of the haematoma. Medical therapy with steroids and/or pro-coagulation therapy is aimed at reducing local swelling and preventing further bleeding but fails to remove the compressive effects of the haematoma. Similarly, embolisation of the bleeding vessel may prevent further bleeding, but fails to remove the haematoma. The treatment approach is therefore complex and depends on the patient's presentation. Three defined groups of patients where the management varies are adults, pregnant women and children.

2. Adults

2.1 Demographics and risk factors

SSEH can affect all adult age groups, but most reported cases are aged between 50 and 80 years [14]. There is no obvious gender association, although pregnancy is an independent risk factor [14]. In most adult patients no known risk factors are identified, but according to the most recent review 19% had hypertension, 14% had cardiovascular disease, 3% had bleeding disorders and 6% had diabetes or obesity [14]. Interestingly, bleeding disorders did not specifically affect a younger population, but as expected by their cumulative medical comorbidities, patients on anticoagulation therapy were generally older. The most common anticoagulants identified in adults with SSEH were Warfarin (43.9%), Aspirin (36.8%) or Clopidogrel (10.5%).

2.2 Clinical presentation

At presentation, most adults (98%) complain of back and radicular pain [14]. Most haematomas reported in the literature cause spinal canal compression with severe caudal motor (58% Medical Research Council grade 0) and sensory dysfunction (70% American Spinal Injury Association (ASIA) A-C) [14]. In contrast, relatively few patients presented with compression of a nerve root in the neuroforamen, and in these cases most patients (63%) maintain normal or near normal motor (70% MRC grade 4 or 5) and sensory function (63%) [14].

Of concern, up to 15% of adults with SSEH are suspected of having an alternative diagnosis, half of which are thought to be affected by an ischaemic incident for which anticoagulation therapy is considered or administered. The most common diagnoses that SSEH mimic are cerebro-vascular accidents (CVA), myocardial infarctions, spinal tumours and disc prolapses. Thus, clinicians should be aware of this condition and its ability to mimic alternative diagnoses, prior to considering anticoagulation therapy.

2.3 Investigations

The diagnosis is easily confirmed on cross-sectional imaging, most commonly an MRI scan or CT myelogram. An MRI is preferential as it offers a clear localisation and cause of compression (**Figure 1**). In contrast to a CT which accurately localises compression but is less accurate in determining the underlying cause (**Figure 2**).

In adults most SSEH affect the cervico-thoracic region between C3 and T3. This is attributed to this region being the only area of the spinal canal with a compact continuous epidural venous network [15]. These haematomas can be large and affect up to 28 spinal levels, with the average number of levels affected being 5.4 vertebral levels. Furthermore, although rare, multi-site haematomas can occur, therefore whole spine imaging is advocated.

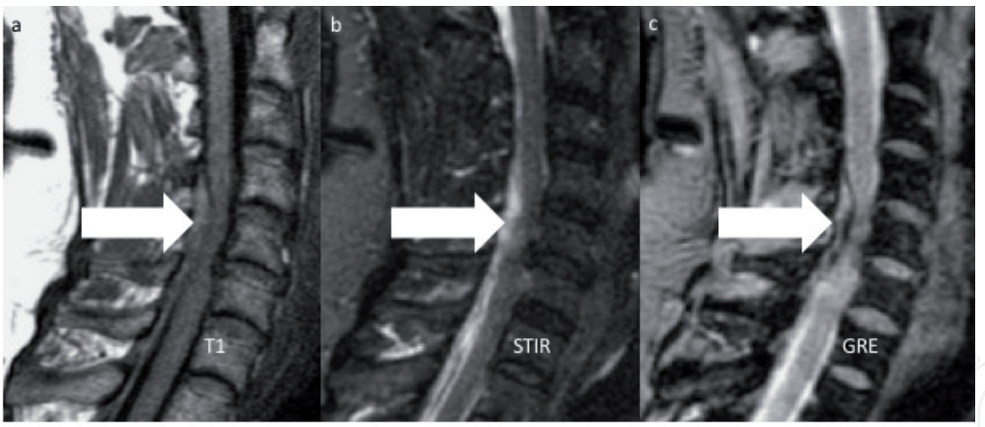

Figure 1.
A T1 (a), STIR (b) and GRE (c) sagittal MRI sequence of a C4–6 SSEH compressing the spinal cord.

Figure 2.
A sagittal non-contrast CT scan of a C2–6 SSEH compressing the spinal cord.

2.4 Treatment

The gold standard of treatment for patients with SSEH and neurological dysfunction is expeditious surgical decompression. However, in patients with no neurology or with improving neurology, non-operative treatment with close neurological monitoring is the treatment of choice. There is no evidence to support operative intervention based on the presence of back pain or the size of the haematoma, rather, neurological dysfunction guides the decision to operatively intervene. In patients with a bleeding tendency, systemic pro-coagulant therapy should also be used.

With this approach to treatment, 85% of patients can be expected to make a complete or partial recovery [14]. This recovery is usually rapid, but in some patients this may take a number of years, which results in a reported average time to

recovery of 52.4 days [14]. These outcomes are unrelated to patient age, sex, haematoma size or location.

According to the current literature, 5% of patients worsen their neurological function despite treatment and the mortality rate is 3.5% [14]. The most common cause of death is cardiac disease rather than neurological dysfunction from SSEH.

3. Pregnancy

Unlike in the general population, SSEH carry the additional concern for the well-being of the foetus. Thus, the goal of treatment is to restore normal neurological function to the mother, but with minimal risk to the foetus [13].

Like the general adult population, the treatment options include surgical decompression, but this carries surgical and anaesthetic risks; radiological aspiration with or without embolisation, but this too carries anaesthetic risks, rarely evacuates the haematoma completely, risks neuro-vascular injury and exposes the mother and foetus to radiation; or chemotherapy with steroids and/or coagulation therapy, but this fails to remove the compressive effects of the haematoma and requires systemic administration of the drug. Thus, the treatment approach is complex and requires consideration of both the mother and foetus.

3.1 Demographics and risk factors

In pregnancy, an increased blood volume and pressure predisposes to rupture of vascular walls and therefore SSEH [16]. Thus, it is rare for SSEH to occur in the first trimester but the risk increases with the duration of pregnancy with an average gestational age of 34.3 weeks being reported in the literature [17]. No cases have been reported during labour, although some have been reported in the days and weeks following delivery, which suggests that the pregnancy-induced dilation of epidural vessels, rather than the mechanical straining of labour, is the predominant cause of SSEH in pregnancy.

There does not appear to be a relationship between the gravid status and the risk of SSEH [17]. Furthermore, most patients do not have any known risk factors, although pre-eclampsia has been reported in two cases. No pregnant patients with SSEH have been reported to be on anticoagulation therapy [17].

3.2 Clinical presentation

Like the general population, patients present with back or neck pain, usually with neurological dysfunction [17]. Within the English literature, most patients have severe spinal cord dysfunction with gait disturbance, caudal weakness and sensory disturbance. Only one reported patient has presented without neurological dysfunction [17].

3.3 Investigations

In pregnancy, limited radiation exposure to the foetus is necessary, thus an MRI is the investigation of choice. No patients have been reported to have had a CT scan, although, if an MRI is not possible and the diagnosis is critical then a CT or CT myelogram is necessary with protection of the pelvis [7].

Like the general adult population, the typical location of the SSEH is the cervico-thoracic region [17]. The size of the haematoma has been reported to span two to six spinal levels [17].

3.4 Treatment

In mothers with neurological dysfunction and less than 32 weeks of gestation, surgical decompression with foetal monitoring is advocated. In contrast, in mothers with neurological dysfunction and more than 32 weeks of gestation, a caesarean section followed by surgical decompression is advocated. Like in the general population, neurological dysfunction guides surgical intervention, not the presence of back pain or the size of the haematoma. Thus, in mothers without neurological dysfunction, close monitoring without surgical intervention is appropriate [17]. With such treatment most mothers are expected to make a complete (56%) or partial (38%) recovery.

4. Children

SSEH can occur in children. However, in contrast to adults the difficulty conversing with children and attaining investigations, such as cross-sectional imaging, makes the diagnosis difficult. In addition, their rarity and occasional atypical presentations mimicking alternative diagnoses compounds the challenges of diagnosis [9, 8]. Yet, the consequences of this condition can be devastating.

4.1 Demographics and risk factors

Children of all ages (range 3 months - 18 years) have been identified with SSEH [18]. There does not appear to be a gender relationship to the risk of developing SSEH, however bleeding tendencies, such as haemophilia, which do construe a risk (8% of reported cases), are clearly more common in males [18]. One case was reported to be on anti-coagulation therapy and two have been reported to have an arterio-venous malformation.

4.2 Clinical presentation

Due to the variance in patient age, young children cannot report the location of pain, however, in those that are conversant most complain of back or neck pain, with radicular pain being less common [18]. Similarly, the neurological dysfunction, particularly sensory disturbance, is harder to define than in an adult population. However, most children are affected by neurological dysfunction (97%), most commonly spinal cord dysfunction, although isolated nerve root dysfunction is also reported [18].

Nearly one third of all reported cases are initially suspected of having an alternative diagnosis, most commonly Guillain-Barre syndrome, meningitis, cerebral palsy, Grisel's syndrome or an acute abdomen [18].

4.3 Investigations

A whole spine MRI scan is the investigation of choice. A CT myelogram or non-contrast CT may be required depending on the patient's compliance and compatibility with an MRI scan, although the ionising radiation risk should be recognised and exposure limited.

As with adults the typical location of these haematomas are the cervico-thoracic region, and the size of the haematoma may span one to 22 spinal levels [18].

4.4 Treatment

Similar to adults, neurological dysfunction determines surgical intervention, not the presence of back pain or the size of the haematoma. Thus, in patients with neurological dysfunction, the gold standard treatment is surgical decompression and evacuation of the haematoma. The surgical approach to achieve this depends on the haematoma size, location, consistency and surgeon familiarity. While partial or complete laminectomies as well as laminoplasties have been reported, the consequences of each approach on the long-term consequences on spinal growth and function in these patients remain unknown. Thus, the least invasive approach to achieve decompression and haematoma evacuation should be utilised, but patients should also be counselled of the longer-term risks that surgical intervention may impose.

In patients without neurological dysfunction or with improving neurology, non-operative care with close neurological monitoring is appropriate. In patients with a bleeding tendency, systemic pro-coagulant therapy should be initiated, and if neurological improvement rapidly occurs, then non-operative treatment should be continued, but if no improvement occurs, then surgical intervention should be undertaken [18].

With this approach most children (83%) are expected to make a complete (69%) or partial (14%) neurological recovery [18]. However, this condition can result in death, with two children having been reported to have died following a SSEH. One child died from respiratory failure after operative intervention and the other without a cause for death being defined, but with evidence of progressive neurological dysfunction after surgical decompression [3, 4].

5. Vascular malformations

While most SSEH are believed to occur in the absence of local vascular malformations, there remains debate as to the number of these haematomas that have a predisposing vascular pathology, most notably arterio-venous fistulas, malformations and aneurysms. This is predominantly due to a poor specificity of initial imaging modalities to identify small vascular pathologies in the presence of a epidural haematoma, the rarity of sending pathological specimens of the local vasculature for histological assessment and the lack of follow-up imaging. However, local vascular pathologies have been identified in spinal epidural haematomas that develop without an inciting event in all age groups, although it is more commonly reported in children [18].

Clearly, an area of vascular fragility, due to an underlying malformation can predispose to spontaneous bleeding. The location of the subsequent haematoma from the bleed depends on the location of the vascular disruption. In the case of an epidural haematoma, the pressure exerted on the neural structures may cause neurological dysfunction for which the treatment is the same as that of a SSEH. If surgical intervention is necessary then coagulation of the vascular anomaly at the time of surgery to prevent further bleeding is necessary. This is often inadvertently performed during the coagulation manoeuvres undertaken to stop bleeding at the time of surgery. Fortuitously, this typically treats the underlying vascular pathology and avoids recurrence.

6. Summary

To date, the major limitations of the literature reporting on SSEH are the low numbers, publication bias, the variance in imaging and reporting of vascular

malformations and the lack of randomised control trials assessing outcome. However, it is clear that all age groups can be affected by SSEH and that three subsets of patients exist, namely adults, pregnant women and children.

In all groups, SSEH can present with variable symptoms and signs but most have neck or back pain with neurological dysfunction. In adults, SSEHs can mimic ischaemic events and lead to inappropriate anticoagulation therapy, which can be avoided by clinical awareness of this condition and whole spine cross-sectional imaging. Similarly, in children the diagnosis can be challenging due to the problems conversing with children and the variable presentations, often mimicking neuromuscular disorders. It is further compounded by the challenges of attaining cross-sectional imaging, which is again diagnostic. In pregnancy, the further difficulties lie in the protection of the unborn foetus, with the avoidance of ionising radiation, but cross-sectional imaging remains diagnostic.

In all groups, most SSEH affect the cervico-thoracic region and span multiple spinal segments. The gold standard of treatment for patients with neurological dysfunction is surgical decompression and haematoma evacuation. However, in patients without neurological dysfunction, close neurological monitoring without surgical intervention is appropriate. In adults with bleeding disorders, pro-coagulant therapy should be used as an adjunct to treatment, however in children with bleeding disorders pro-coagulant therapy may be trialled prior to surgical intervention. In pregnancy, surgical intervention involves surgical decompression with foetal monitoring in those of gestational age less than 32 weeks and a caesarean section followed by decompression in those of gestational age greater than 32 weeks. In children, the long-term effect of surgical intervention on spinal growth and function should be recognised.

Author details

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