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CASE REPORT

Spontaneous spinal epidural hematoma in an infant presenting with Horner syndrome

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Abstract

Background Spontaneous spinal epidural hematoma (SSEH) is a rare neurologic entity, especially in infants, that develops in the absence of underlying coagulopathy, bleeding diathesis, infection, vascular malformation, trauma, iatrogenic, or other identifiable cause. In contrast to adults, diagnosis is frequently delayed or missed in infants due to non-specific symptoms and limited clinical examination.

Case illustration An 11-month-old female demonstrated symptoms of irritability, intermittent diarrhea, lethargy, decreased oral intake, and difficulties crawling before presenting to the emergency room. At time of presentation, she was noted to have minimal spontaneous movement of the lower extremities and anisocoria with ptosis of the right eye. Given her clinical presentation, a magnetic resonance image (MRI) of the spine was obtained which revealed an epidural hematoma with compression extending from C7-T3. She underwent C7-T3 laminoplasty and hematoma evacuation. Following surgical intervention, she demonstrated significant improvements in her lower extremity strength and resolution of Horner syndrome.

Conclusion SSEH in infants is a rare neurologic condition, with diagnosis often delayed due to nonspecific symptomatology. Prompt diagnosis and intervention are essential in the treatment of SSEH to prevent permanent neurologic dysfunction. Physicians should have a high index of suspicion for SSEH in these instances, and investigation with spinal MRI imaging is recommended.

Keywords Spontaneous spinal epidural hematoma · Horner syndrome · Spinal compression

Introduction

Spontaneous spinal epidural hematoma (SSEH) is a rare entity, especially within the pediatric population [1–3], that develops in the absence of coagulopathy, bleeding diathesis, infection, vascular malformation, trauma, or other identifiable cause, and accounts for 40–50% of all spinal epidural hematomas [4]. Diagnosis is frequently delayed in infants due to non-specific symptomatology and limited clinical examination. Further diagnostic clues and high index of suspicion are required to identify this pathology and maximize recovery. If left untreated, SSEHs can lead to serious neurologic deficits. The authors describe a case of an 11-month-old presenting with irritability, lower extremity paresis, and Horner syndrome in the setting of SSEH.

Case illustration

An 11-month-old female presented to her pediatrician multiple times during the months leading up to her hospitalization for evaluation of irritability, diarrhea, and decreased oral intake. Her symptoms appeared to resolve spontaneously, and she was diagnosed with a gastrointestinal viral illness. Three days prior to her hospital presentation, she was noted to be irritable, with difficulties crawling and limited spontaneous movement of her lower extremities. She was taken

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Fig. 1 Photograph demonstrating miosis and ptosis of the right eye, consistent with Horner syndrome

to an urgent care facility and diagnosed with sequela from a viral illness.

The following morning, parents noted no movement of her legs and immediately presented to the emergency department. Upon initial evaluation, the patient was noted to have significant bilateral lower extremity paresis and lethargy. Computed tomography (CT) of the head was unremarkable. Due to concern for Guillain–Barre Syndrome (GBS), a lumbar puncture (LP) was performed which demonstrated elevated protein and normal inflammatory markers. Examination by the consulting pediatric neurologist revealed anisocoria and ptosis of the right eye in addition to lower extremity paresis (Fig. 1). This critical examination finding of Horner syndrome was confirmed by her parents to be present prior to arrival to the emergency room. Magnetic resonance imaging (MRI) of the spine was urgently recommended which revealed a compressive epidural hematoma extending from C7-T3 (Fig. 2a). She underwent emergent C7-T3 laminoplasty and hematoma evacuation. Postoperative imaging demonstrated decompression of the spinal elements with remaining signal change within the spinal cord from C6 to T1 (Fig. 2b). Following surgical decompression, she demonstrated early improvement in her lower extremity function. Further clinical workup remained negative for underlying coagulopathy, hemophilia, or other bleeding disorder. She was subsequently discharged from the hospital. At her 3-month clinic visit, she exhibited significant improvements in her lower extremity strength and resolution of the Horner syndrome.

Discussion

SSEH is a rare entity that can produce varying degrees of neurologic deficits. Symptoms range from pain or radiculopathy to complete paralysis, sensory loss, and incontinence. Without swift diagnosis and treatment, SSEH has the potential to cause permanent neurologic compromise. Identified causes of spinal epidural hematomas include vascular malformations, including hemangiomas or arteriovenous malformations (AVM) [5], trauma [6], bleeding disorders [7], coagulopathy, or use of anticoagulants [8]. Iatrogenic cases have also been described in the setting of lumbar puncture and epidural anesthesia [9, 10]. In other instances, the cause remains unidentifiable. The incidence of SSEH has been reported as 0.1 per 100,000 patients in the general population and is exceedingly rare in infants less than 1 year of

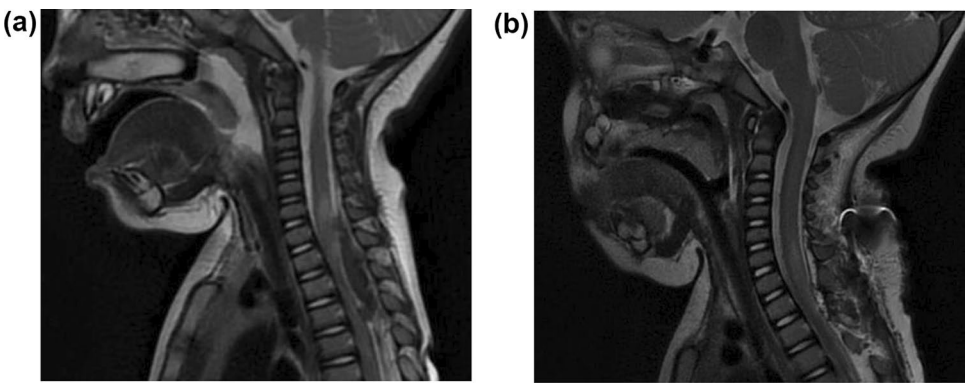

Fig. 2 **a** A sagittal T2-weighted MRI of the cervical and thoracic spine demonstrating the dorsal epidural hematoma and spinal compression from C7-T3. **b** Postoperative T2-weighted MRI of the cervical spine demonstrating decompression of the C7-T3 spinal levels with residual edema within the spinal cord from C6-T1

age [11], with only 7 prior cases reported in the literature [12–18].

Most authors consider SSEHs to be venous in origin. The spinal internal venous plexus is composed of both anterior and posterior portions. The anterior plexus is held adherent to the posterior longitudinal ligament via Hofmann's ligament [19], while the posterior plexus courses among the loose epidural fat making it prone to rupture. The valveless nature of the spinal venous plexus increases its susceptibility to elevations in intra-abdominal and intrathoracic pressure [20]. Sudden increases in pressures induced by crying, voiding, coughing, or straining can result in venous backflow and subsequent rupture. SSEHs are predominantly dorsal which is in agreement with the proposed posterior venous plexus hypothesis, although hematoma development along the ventral aspect of the spinal canal has been described [21]. Pediatric patients are more likely to develop cervico-thoracic SSEHs as compared to the majority of thoracolumbar SSEHs seen in adults [22]. Disproportionate head to weight ratios, increased cervical spine mobility, and underdeveloped neck musculature may account for this difference [13].

Formulating the appropriate diagnosis is challenging in young children. Children under 2 years of age often present with nonspecific complaints of irritability and excessive crying followed by progressive motor weakness days to weeks later [22]. Symptoms may progress rapidly, leading to dense neurologic deficit. The clinical presentation and severity depend on a variety of factors including speed of blood accumulation, hematoma volume, and spinal level involvement [23]. It is therefore important for physicians to have a high index of suspicion to distinguish this entity from other common pediatric pathologies. As shown in other cases and our own, SSEH may be mistaken for GBS as both share similar symptomatology and cerebrospinal fluid (CSF) albuminocytologic dissociation [12, 18, 24]. Diagnostic imaging, specifically MRI of the spine, is essential to diagnose SSEH and minimize morbidity [25].

Spinal epidural hematomas producing cervical cord compression may also generate subtle clinical exam findings that, if detected, can assist practitioners in localizing the pathology. Munakomi et al. [26] described a case of a 16-year-old who developed Horner syndrome in the setting of spinal epidural hematoma following rupture of an AVM. To our knowledge, our case is the first documented presentation of Horner syndrome in an infant with SSEH. Clinical outcome of SSEH is related to the affected spinal level [23], duration of symptoms prior to treatment [23, 27, 28], and severity of neurologic deficit on presentation [12, 23, 27, 28]. Although neurologic recovery has been described with conservative treatment [21, 29, 30], prompt diagnosis and operative intervention remain the recommended management for symptomatic SSEHs. Even in the setting of delayed diagnosis, surgical intervention has been shown to result in more favorable outcomes in children less than 2 years of age as compared to adults [1, 13, 22].

Prompt diagnosis and intervention are essential to prevent permanent neurologic dysfunction in infants with SSEH. This case highlights the importance of a thorough clinical examination, which may result in identification of subtle neurologic clues to aid in diagnosis.

Declarations

Informed consent The legal guardian has consented to the submission of the case report to the journal, including the use of clinical data and photographs.

Conflict of interest No conflicts of interest for each author related to the manuscript or its subject matter.

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