Brain & Development 41 (2019) 392–395

BRAIN & DEVELOPMENT
Official Journal of the Japanese Society of Child Neurology

www.elsevier.com/locate/braindev

Case Report

Spontaneous spinal epidural hematoma mimicking Guillain-Barre Syndrome

Aya Kondo [a], Hiroshi Yamaguchi [a,b,*], Yusuke Ishida [a], Daisaku Toyoshima [a], Mai Azumi [c], Nobuyuki Akutsu [c], Junji Koyama [c], Hiroshi Kurosawa [d], Atushi Kawamura [c], Azusa Maruyama [a]

[a] Department of Neurology, Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan
[b] Department of Pediatrics, Kobe University Graduate School of Medicine, Kobe, Japan
[c] Department of Neurosurgery, Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan
[d] Department of Pediatric Critical Care Medicine, Hyogo Prefectural Kobe Children's Hospital, Kobe, Japan

Received 9 March 2018; received in revised form 23 August 2018; accepted 6 November 2018

Abstract

Background: The initial symptoms of Guillain-Barre Syndrome (GBS) can be similar to a case of spontaneous spinal epidural hematoma (SSEH) located at the cervicothoracic junction. Therefore, SSEH may be misdiagnosed as GBS. Case Report: A previously healthy 6-year-old girl presented with a 2-day history of progressive pain in the lower extremities and an inability to walk. On initial evaluation, she was completely paraparetic in the lower extremities. Deep tendon reflexes were absent in the lower extremities, and Babinski reflexes were positive on both sides. She exhibited reduced response to light touch and pinprick with a sensory level below T10, and experienced difficulty during urination. However, the strength, sensation and flexion of upper extremities were normal. Because her presentation and examinations were consistent with GBS, we initiated intravenous immunoglobulin therapy. The next day, she also developed pain and muscle weakness of the right upper extremity. Three days after admission, respiratory depression progressed rapidly. Spinal MRI showed a mass extending from the level of C7-T3, with spinal cord compression. The patient underwent an emergency laminectomy with evacuation of hematoma, and was diagnosed with SSEH. Sixty days after admission, she was transferred to the rehabilitation hospital with severe neurologic sequelae of paralysis in both legs. Conclusion: SSEH might have severe consequences, including neurologic deficits and risk of death. This case report serves to raise the awareness of SSEH that mimics the initial presentation of GBS.

Keywords: Albuminocytologic dissociation; Areflexia; Babinski sign; Cerebrospinal fluid; Guillain-Barre Syndrome; Spinal shock; Spontaneous spinal epidural hematoma

1. Introduction

Spontaneous spinal epidural hematoma (SSEH) is a rare neurologic emergency of the spinal cord, the diagnosis of which is often difficult because of its rarity and atypical symptoms. Initial symptoms typically include neck and back pain, followed by pain and plegia of the extremities, or bladder and rectal disturbances [1–3].

Guillain-Barre Syndrome (GBS) is an acute autoimmune neuropathy. Neurologic deficits of GBS commonly begin as weakness in the lower limbs, migrating upward to the trunk and upper limbs [4,5]. Its initial

* Corresponding author at: Department of Neurology, Hyogo Prefectural Kobe Children's Hospital, 1-6-7 Minatojimaminamimachi, Chuo-Ku, Kobe, Hyogo 650-0047, Japan.
E-mail address: hiyamaguchi_kch@hp.pref.hyogo.jp (H. Yamaguchi).

https://doi.org/10.1016/j.braindev.2018.11.004
0387-7604/

symptoms are very similar to SSEH at the cervicothoracic junction. Therefore, SSEH may be misdiagnosed as GBS.

Here, we present a rare and severe prognostic case of SSEH in a 6-year-old girl, where the initial symptoms and test results mimicked GBS.

2. Case report

A previously healthy 6-year-old girl presented to our emergency department with a 2-day history of progressive pain in the lower extremities and an inability to walk. One month prior to admission, she suffered a neck injury during a sporting event (a friend fell on her neck). However, it did not immediately affect her daily life. The patient exhibited a severe cough and one day of fever, approximately one week prior to admission.

On initial evaluation, she was afebrile, awake, and alert. Neurologic examination revealed significant pain in both lower extremities. The patient was completely paraparetic in the lower extremities. Deep tendon reflexes were absent in the lower extremities, and Babinski reflexes were positive on both sides. She exhibited reduced response to light touch and pinprick with a sensory level below T10, and experienced difficulty during urination. However, strength, sensation and flexion of upper extremities were normal. First, we performed magnetic resonance imaging (MRI) of the lower spine and performed a lumbar puncture. MRI was normal, and cerebrospinal fluid (CSF) examination revealed an increased level of protein (192 mg/dL) without other abnormalities. As her presentation and examinations were consistent with GBS, we initiated intravenous immunoglobulin (IVIG) therapy. However, strength or pain in both of her lower extremities did not improve. The next day, she also developed pain and muscle weakness of the right upper extremity. Three days after admission, respiratory depression progressed rapidly. Emergency spinal MRI showed a mass extending from C7-T3 level with spinal cord compression (Fig. 1). The patient underwent an emergency laminectomy with evacuation of hematoma, and she was diagnosed with SSEH. Operative findings showed a dark brown-colored clot in the liquid state, indicating chronic venous bleeding. There were no apparent vascular malformations.

After the operation, we administered methylprednisolone pulse therapy for 3 days. Although her pain in both lower extremities gradually improved and follow-up spinal MRI revealed no hematoma 12 days after operation, her complete paraplegia had not improved. We performed tracheotomy 15 days after the operation because she experienced tension pneumothorax twice, revealing upper airway obstruction and difficulty expectorating. Fifty-six days after admission, her tracheotomy tube was removed. She could talk and eat, and exhibited no respiratory difficulties. Sixty days after admission, she was transferred to the rehabilitation hospital; she then showed only neurologic sequelae of paralysis in both legs, but could move her toes.

3. Discussion

SSEH is a rare clinical entity, the incidence of which is estimated at 0.1 patients per 100,000; its pathogenesis remains unclear [1,6]. As clinical symptoms depend on the involved vertebral levels, it is often difficult to diagnose. In infants and children, initial symptoms can be non-specific and manifest primarily as irritability or crying [2,3,7]. Thus, there is often a delay in diagnosis. Our patient experienced a neck injury, followed by severe cough, at 6 weeks prior to hospitalization, respectively. However, we did not identify a specific cause.

Physicians should be aware that SSEH may occur in children, mimicking the initial presentation of GBS. Our SSEH case mimicked GBS in terms of progressive weakness of the legs and progressive respiratory disturbances, as well as pain and loss of bilateral leg sensations, early bladder dysfunction, areflexia in legs, and albuminocytologic dissociation on CSF analysis. Most GBS patients show decreased or absent reflexes in arms or legs [8]; SSEH patients may exhibit these altered reflexes if they have "spinal shock." Severe damage of the spinal cord initially results in a profound depression of spinal reflex activity [9]. GBS patients typically show albuminocytologic dissociation on CSF analysis [3]. However, it is possible that SSEH shows this dissociation as a result of blockage of the CSF [10].

There have been two reports of SSEH described as GBS [2,3]. Cakir et al. reported a clinical course of SSEH mimicking GBS in 9-year-old female, with progressive inability to walk and bilateral arm pain [2]. Similar to our case, the clinical course imitated GBS, and IVIG therapy was initiated. An MRI, 3 days after admission, showed thoracic epidural hematoma between T2 and T8. The patent underwent emergency surgery and fully recovered over the following 2 weeks. Although the initial clinical course was quite similar to our case, the outcome of the patient dramatically differed. There are three possible reasons. First, the position of SSEH was higher in our patient (C7-T3) than in the previous case (T2-T8). Second, according to MRI findings, the compression of spinal cord was more severe in our patient. Finally, our patient developed more severe neurologic symptoms, including deterioration of respiratory status, likely related to edema of the upper cervical spine. Importantly, a severe preoperative neurological status has been associated with a worsened prognosis [11]. Lee et al. reported SSEH comorbid with GBS in a 32-month-old with progressive paraparesis [3]. Surprisingly, the patient fully recovered,

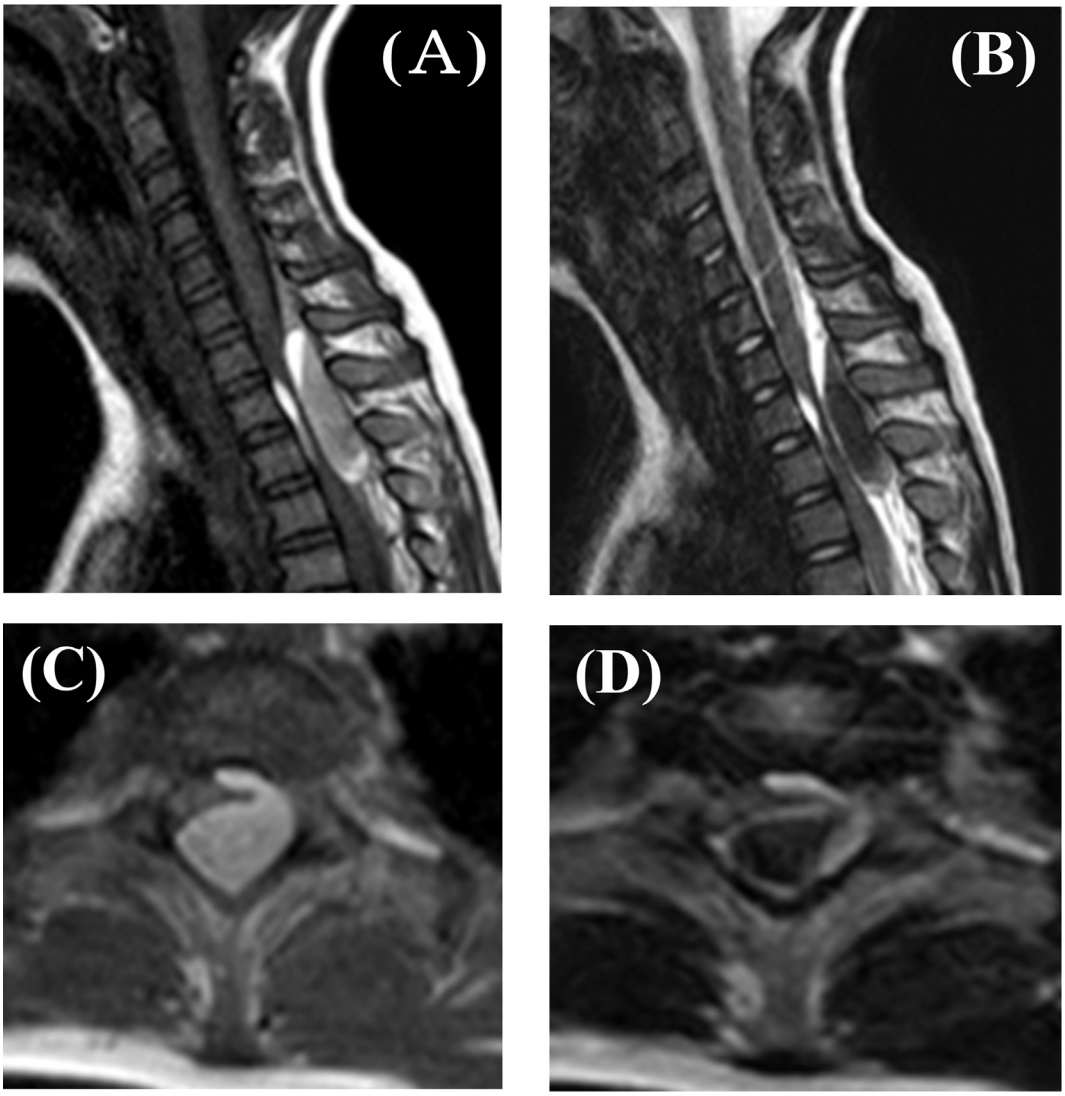

Fig. 1. Preoperative sagittal ((A) (T1-weighted), (B) (T2-weighted)) and axial ((C) (T1-weighted), (D) (T2-weighted)) magnetic resonance images show a longitudinal epidural hematoma, which compresses the spinal cord in the dorsal to the left lateral epidural space at the level of C7-T3. The lesion was isointense to the spinal cord on T1-weighted images, and hyperintense on T2-weighted images. The findings of the hematoma, showing epidural space-occupying lesion, extending from the left side to the opposite site and pushing the dura mater anteriorly, distinguish this as an epidural hematoma, rather than a subdural hematoma.

one week after delayed hemilaminectomy. Preoperative interval for decompressive laminectomy and hematoma evacuation may be one of the key factors in recovery [7]. However, Liu et al. reported that patients with complete neurologic deficits showed disappointing prognosis, even if they underwent operation in <24 h [1]. In summary, because of our patient's degree of spinal compression and level of hematoma, she might show poor prognosis. Unfortunately, prior case reports did not discuss the Babinski sign, which indicates the presence of central or high spinal lesions. Therefore, this sign might facilitate the diagnosis of SSEH.

In conclusion, this case describes SSEH mimicking GBS. In cases of SSEH at the cervicothoracic junction, no symptoms or neurological abnormality of upper extremities may be observed. Cervicothoracic spine MRI is required if the case does not match findings of GBS, such as positive Babinski reflexes.

Funding

The authors received no financial support.

Acknowledgements

We thank Drs. Nobuo Kohara, Keisuke Yoshikawa, and Susumu Kusunoki for their advice regarding the patient. We also thank Dr. Yoshinobu Akasaka, Department of Radiology in Hyogo Prefectural Kobe

Children's Hospital, for his advice regarding the patient's MRI findings.

References

[1] Liu Z, Jiao Q, Xu J, Wang X, Li S, You C. Spontaneous spinal epidural hematoma: analysis of 23 cases. Surg Neurol 2008;69:253–60 [discussion 260].
[2] Cakir E, Karaarslan G, Usul H, Baykal S, Kuzeyli K, Mungan I, et al. Clinical course of spontaneous spinal epidural haematoma mimicking Guillain-Barré syndrome in a child: a case report and literature review. Dev Med Child Neurol 2004;46:838–42.
[3] Lee CH, Song GS, Kim YH, Son DW, Lee SW. spontaneous spinal epidural hematoma coexisting Guillan-Barré Syndrome in a child: a case report. Korean J Spine 2016;13:167–9.
[4] Mitsui Y, Kusunoki S, Arimura K, Kaji R, Kanda T, Kuwabara S, et al. Japanese GBS study group. A multicentre prospective study of Guillain-Barré syndrome in Japan: a focus on the incidence of subtypes. J Neurol Neurosurg Psychiatry 2015;86:110–4.
[5] Sudulagunta SR, Sodalagunta MB, Sepehrar M, Khorram H, Bangalore Raja SK, Kothandapani S, et al. Guillain-Barré syndrome: clinical profile and management. Ger Med Sci 2015;13:Doc16.
[6] Holtås S, Heiling M, Lönntoft M. Spontaneous spinal epidural hematoma: findings at MR imaging and clinical correlation. Radiology 1996;199:409–13.
[7] Schoonjans AS, De Dooy J, Kenis S, Menovsky T, Verhulst S, Hellinckx J, et al. Spontaneous spinal epidural hematoma in infancy: review of the literature and the "seventh" case report. Eur J Paediatr Neurol 2013;17:537–42.
[8] Fokke C, van den Berg B, Drenthen J, Walgaard C, van Doorn PA, Jacobs BC. Diagnosis of Guillain-Barré syndrome and validation of Brighton criteria. Brain 2014;137:33.
[9] Ashby P, Verrier M, Lightfoot E. Segmental reflex pathways in spinal shock and spinal spasticity in man. J Neurol Neurosurg Psychiatry 1974;37:1352–60.
[10] Markham JW, Lynge HN, Stahlman GE. The syndrome of spontaneous spinal epidural hematoma. Report of three cases. J Neurosurg 1967;26:334–42.
[11] Rajz G, Cohen JE, Harnof S, Knoller N, Goren O, Shoshan Y, et al. Spontaneous spinal epidural hematoma: the importance of preoperative neurological status and rapid intervention. J Clin Neurosci 2015;22:123–8.