Spontaneous Spinal Epidural Hematoma in an Infant with Developmental Disabilities

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Abstract:

Introduction: Spontaneous spinal epidural hematomas (SSEHs) are rare in childhood, especially in infants.

Case Report: We present the case of a 17-month-old-boy with trisomy 21 and a large SSEH. He was hospitalized for acute onset paraplegia after 6 days of irritability. Nine days after symptom onset, magnetic resonance imaging (MRI) of the spine revealed an extensive epidural hematoma between C7 and T5 causing severe spinal cord compression. After a coagulation disorder was ruled out (12 days after onset), he underwent emergency hemilaminectomy with evacuation of the hematoma. His neurologic impairment gradually improved, and 4 months after surgery he was back to his neurologic baseline. At 18 months after surgery, he was walking independently, although he had some developmental disabilities due to trisomy 21.

Conclusions: Only 20 cases of SSEH in infancy have been previously reported, and this is the first report of SSEH in an infant with developmental disabilities. Because of the non-specific symptoms and difficulty obtaining MRIs in infants, particularly in those with developmental disabilities, the diagnosis and treatment of SSEH may be delayed. However, early diagnosis with MRI and early evacuation of SSEH in patients with severe neurological impairments is important for good outcomes. Attention must be paid to postoperative spinal deformity in infants.

Keywords:
Spontaneous spinal epidural hematomas, Infant, Developmental disability

Introduction

The incidence of spontaneous spinal epidural hematoma (SSEH) has been reported as 0.1 per 100,000 patients per year\(^1\). Although this condition can occur at any age, it is most frequently seen in the fourth and fifth decades of life\(^2\). SSEH is rare in children, especially in infants. In infants, the diagnosis and treatment of SSEH are often delayed because of the non-specific symptoms and difficulty in obtaining magnetic resonance imaging (MRI). In this study, we reviewed the literature and present the case of a 17-month-old boy with trisomy 21 and a large SSEH.

Case Report

A 17-month-old-boy was admitted to our institution for irritability and abrupt onset paraplegia. He had trisomy 21 and mild developmental disabilities, but he was able to walk with assistance prior to symptom onset. Seven days before admission, he cried after the influenza virus vaccination and became irritable. Six days before admission, he became more irritable and was unable to stand. On initial assessment, his consciousness was clear, but he demonstrated motor weakness in the lower limbs, with movement restricted to his ankles only. The deep tendon reflex was increased in both lower limbs, and both ankles displayed clonus. Cerebrospinal fluid showed high levels of protein and myelin basic protein, indicating demyelination. Ten days after onset, to investigate the cause of his paraplegia, we performed MRI of the head to thoracic region under sedation. As a result, MRI of the cervicothoracic spine revealed a large posterior extradural mass extending from the 7\(^{th}\) cervical vertebra to the 5\(^{th}\) thoracic vertebra, the most severe at the 2\(^{nd}\) thoracic vertebra. The mass showed high and low signal intensity on the T1 and T2 weighted images. The spinal cord was displaced anteriorly and severely compressed by the...
MRI of the spine revealed a large posterior extradural mass extending from the 7th cervical vertebra to the 5th thoracic vertebra, most severe at the 2nd thoracic vertebra. The spinal cord was displaced anteriorly and severely compressed by the large mass.

There was no evidence of fracture, active bleeding, or arteriovenous malformation.

Based on these findings, the patient was diagnosed with severe paraplegia in both lower limbs due to SSEH (C7-T4). Twelve days after onset, emergency surgical evacuation using a posterior approach was performed. A T2 spine hemilaminectomy was performed using a high-speed air drill (Fig. 3a). After the T2 hemilaminectomy, the epidural hematoma was visualized (Fig. 3b). The epidural hematoma was evacu-
Figure 3. Operative findings. a) T2 spine hemilaminectomy was performed using a high-speed air drill. b) After the T2 hemilaminectomy, the epidural hematoma was visualized. c) The epidural hematoma was removed.

Figure 4. X-ray findings 18 months after the surgery. a) AP view. b) Lateral view. There was no evidence of postoperative spinal deformity.

We report a case of SSEH in a 17-month-old male with developmental disabilities. The patient had no evidence of a coagulation disorder or any vascular malformations. Kiehna et al. reported a case of spinal epidural hematoma (SEH) in an infant with hemophilia. Min et al. reported a case of SSEH in an infant with a vascular malformation. The diagnosis of SSEH requires extensive laboratory testing for coagulation disorders, as well as CTA imaging. SSEH is rare in infants; only 20 cases have been reported in the literature (Table 1). To our knowledge, this is the first report of SSEH in an infant with developmental disabilities. Because of the non-specific symptoms and difficulty obtaining MRIs in infants, particularly those with developmental disabilities, the diagnosis and treatment of SSEH may be delayed. SSEH
should be suspected in infants presenting with irritability and paraplegia; early MRI, under sedation if necessary, should be considered. Furthermore, Lim et al. reported that prompt surgical intervention could result in a good outcome even if the diagnosis was delayed\(^{15}\). Based on these reports, patients with severe paralysis should be promptly treated with surgical intervention, even when the diagnosis of SSEH is delayed.

Surgical strategies in SSEH generally include a hemilaminectomy to expose the hematoma. Several authors reported spinal deformity after laminectomy developed in younger patients\(^{22-24}\). Based on these reports, surgical strategies that preserve the posterior ligamentous complex should be employed when possible, including laminoplasty and hemilaminectomy. However, there was no evidence indicating safety in terms of spinal deformity after hemilaminectomy, especially in infants. Therefore, long-term X-ray follow-up is necessary after surgery.

Regarding the pathogenesis of SSEH, most previous reports could not determine its cause. In their review, Schoonjans AS et al. reported that, due to the lack of valves in the epidural venous plexus, sudden increases in intra-abdominal and intrathoracic pressure induced by crying, coughing, straining, and trauma may cause a backflow resulting in a sudden increase in pressure. In this case, he cried after the influenza virus vaccination and then became irritable. One possible mechanism of SSEH is that his crying induced a sudden increase in epidural venous plexus pressure.

In conclusion, we present a case of a 17-month-old-boy with trisomy 21 and a large SSEH. Only 20 cases of SSEH in infancy have been previously reported, and this is the first report of SSEH in an infant with developmental disabilities. Surgical intervention for SSEH in patients with severe neurological impairments appears to result in a good prognosis, even if diagnosis and surgery are delayed. However, attention must be paid to postoperative spinal deformity, particularly in infants.

**Conflicts of Interest:** The authors declare that there are no relevant conflicts of interest.

**Author Contributions:** MuM and MaM conceived this study, performed surgery, and drafted the manuscript. TK, TI, and KN performed the surgery. TN, GI, ES, and ES participated in the design of the study, and KU and MT conceived the study and participated in its design and coordination. All authors have read and approved the final manuscript.

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**Table 1. Literature Review of 21 Infants with Spontaneous Spinal Epidural Hematoma.**

| Author          | Sex/Age (months) | Interval between onset and surgery (days) | Treatment (surgical strategy) | Outcome          |
|-----------------|------------------|------------------------------------------|------------------------------|------------------|
| Shenkin et al.  | F/20             | 14                                       | ope                          | CR               |
| Jackson et al.  | F/14             | NA                                       | ope                          | CR               |
| Helman et al.   | M/21             | NA                                       | conserva                     | Right knee hyperreflexia |
| Vallee et al.   | F/22             | 10                                       | ope                          | CR               |
| Licate et al.   | M/18             | 6                                        | ope                          | CR               |
| Caldarelli et al.| F/24             | 3                                        | ope                          | CR               |
| Caldarelli et al.| F/16             | 9                                        | ope                          | CR               |
| Patel et al.    | M/18             | 7                                        | ope                          | CR               |
| Patel et al.    | M/22             | NA                                       | conserva                     | CR               |
| Pai et al.      | M/15             | 2                                        | ope (laminectomy)            | CR               |
| Liao et al.     | MA/7             | 7                                        | ope (laminectomy)            | paraplegia       |
| Lee et al.      | M/4              | 5                                        | ope (laminectomy)            | CR               |
| Lim et al.      | F/20             | 14                                       | ope                          | NA               |
| Arya et al.     | F/12             | NA                                       | coserva                      | CR               |
| Poonai et al.   | F/11             | 60                                       | ope (laminoplasty)           | PR               |
| Ramelli et al.  | M/7              | 14                                       | ope (laminectomy)            | paraplegia       |
| Hosoki et al.   | M/11             | 3.5                                      | ope (hemilaminectomy)        | PR               |
| Schoonjans et al.| M/8              | 4                                        | ope (laminectomy)            | incomplete       |
| Abbad et al.    | M/5              | NA                                       | ope                          | NA               |
| Postnikoff et al.| F/30             | NA                                       | ope                          | CR               |
| Our case        | M/17             | 12                                       | ope (hemilaminectomy)        | CR               |

NA: non-available, ope: operative treatment, conserva: conservative treatment, CR: complete recovery, PR: partial recovery.
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