

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

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Introduction

The incidence of spontaneous spinal epidural hematoma (SSEH) has been reported as 0.1 per 100,000 patients per year¹⁾. Although this condition can occur at any age, it is most frequently seen in the fourth and fifth decades of life²⁾. 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 7th cervical vertebra to the 5th thoracic vertebra, the most severe at the 2nd 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

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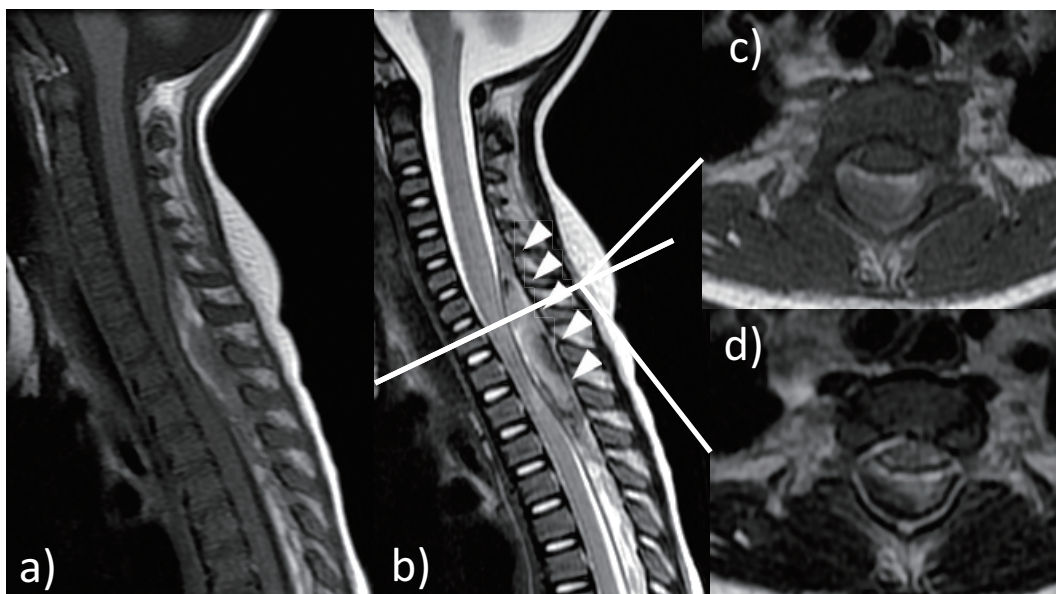


Figure 1. Preoperative magnetic resonance imaging (MRI). a) Sagittal view of T1 weighted image (WI). b) Sagittal view of T2 WI. c) Axial view of T1 WI. d) Axial view of T2 WI.

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.

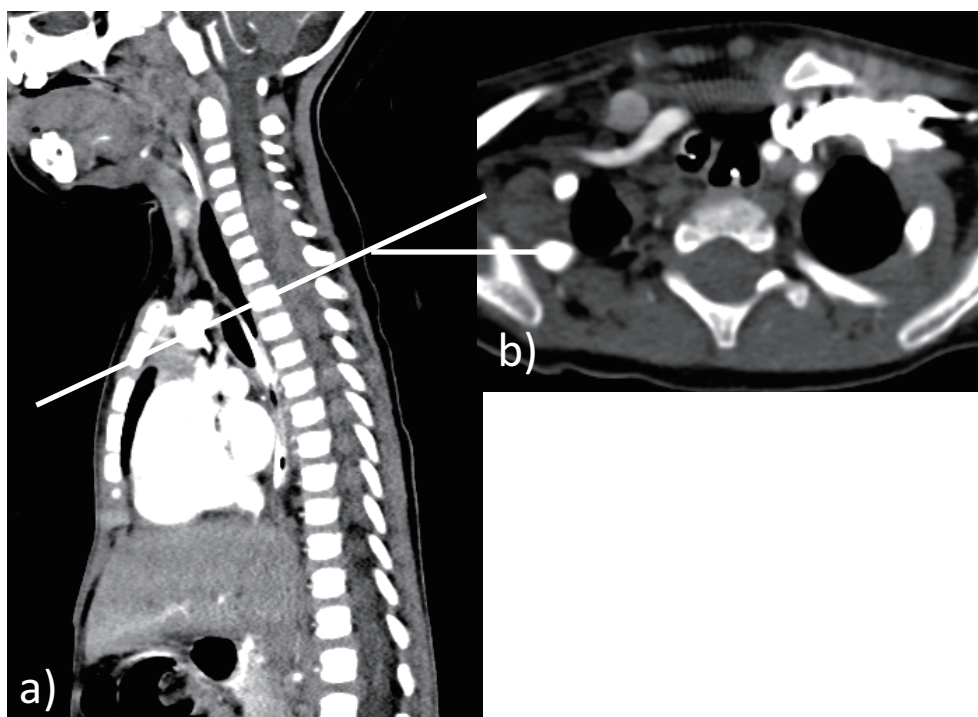


Figure 2. Preoperative computed tomography angiography (CTA). a) Sagittal view of the center of spine, b) Axial view at the T2 level.

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

mass (Fig. 1). Extensive laboratory testing 11 days after onset did not reveal any coagulation defects. Computed tomography angiography (CTA) revealed no evidence of fractures, active bleeding, or an arteriovenous malformation (Fig. 2).

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-

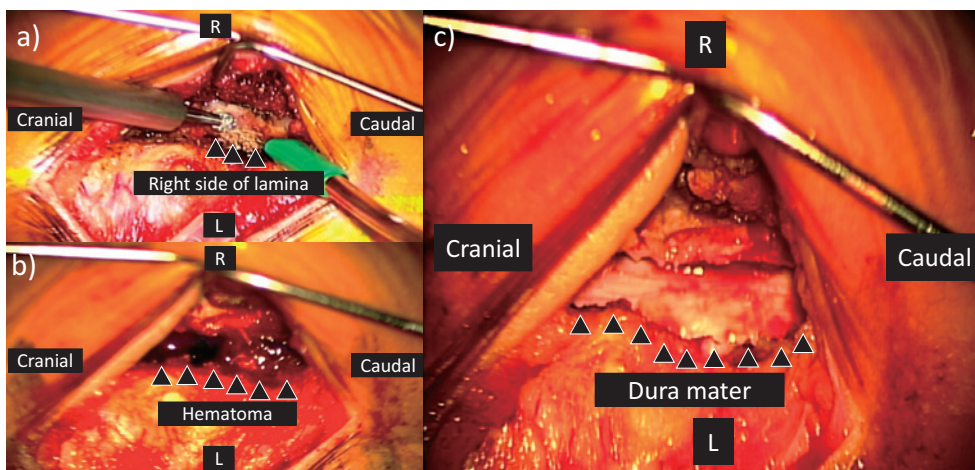


Figure 3. Operative findings. a) T2 spine hemilaminectomy was performed using a high-speed air drill. b) After the T2 hemilaminectomy, the epidural hematoma was visualized. c) The epidural hematoma was removed.

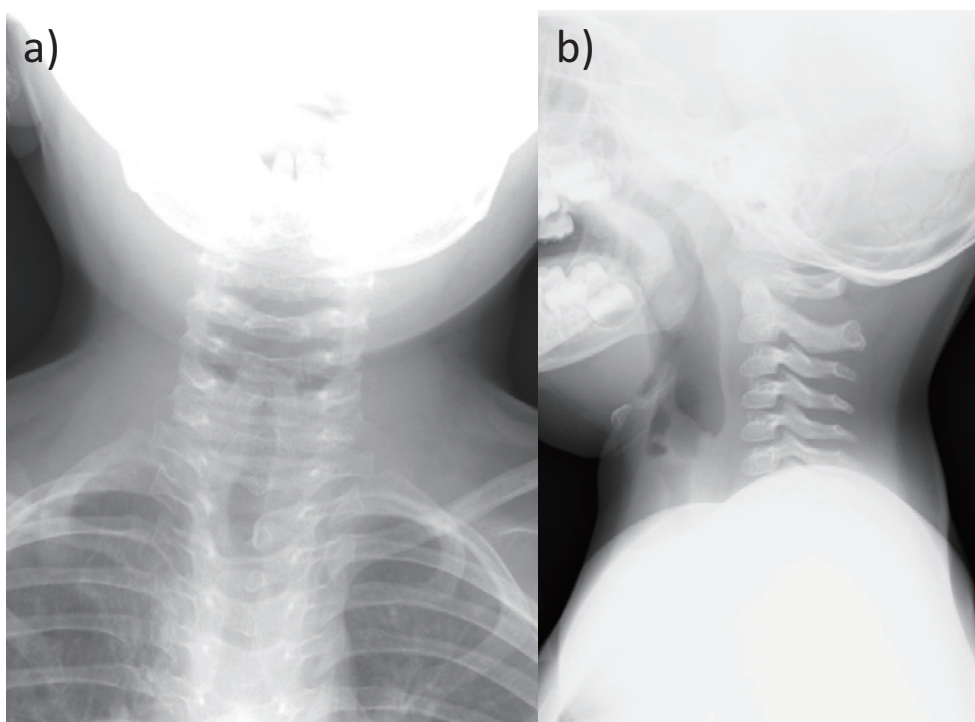


Figure 4. X-ray findings 18 months after the surgery. a) AP view. b) Lateral view. There was no evidence of postoperative spinal deformity.

ated (Fig. 3c). Three days after surgery, he regained motor activity of both lower limbs. Over the next 4 months, his motor skills improved to baseline, and he was able to walk with assistance. By 18 months after surgery, he was walking independently, although he had mild developmental disabilities due to trisomy 21, and he did not display any postoperative spinal deformity (Fig. 4).

Discussion

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 hemophilia³. Min et al. reported a case of SSEH in an infant with a vascular malformation⁴. 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)^{2,5-21}. 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

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
Hehman 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 (hemi-laminectomy)	PR
Schoonjans et al.	M/8	4	ope (laminectomy)	incomplete
Abbad et al.	M/5	NA	ope	NA
Posnikoff et al.	F/30	NA	ope	CR
Our case	M/17	12	ope (hemi-laminectomy)	CR

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

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 delayed¹³. 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 patients²²⁻²⁴. 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, cough, voiding, 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.

References

1. Groen RJ, van Alphen HA. Operative treatment of spontaneous spinal epidural hematomas: a study of the factors determining postoperative outcome. *Neurosurgery*. 1996;39(3):494-508.
2. Schoonjans AS, De DJ, Kenis S, et al. Spontaneous spinal epidural hematoma in infancy: review of the literature and the "seventh" case report. *Eur J Paediatr Neurol*. 2013;17(6):537-42.
3. Kiehna EN, Waldron PE, Jane JA. Conservative management of an acute spontaneous holocord epidural hemorrhage in a hemophilic

- infant. *J Neurosurg Pediatr* 2010;6(1):43-8.
4. Min S, Duan Y, Jin A, et al. Chronic spontaneous cervicothoracic epidural hematoma in an 8-month-old infant. *Ann Saudi Med* 2011;31(3):301-4.
 5. Abbas A, Afzal K, Mujeeb AA, et al. Spontaneous ventral spinal epidural hematoma in an infant: an unusual presentation. *Iran J Child Neurol*. 2013;7(2):47-50.
 6. Arya R, Jain P, Kumar A, et al. Spontaneous spinal epidural hematoma in an infant. *J Child Neurol* 2012;27(12):1577-9.
 7. Caldarelli M, Di RC, La MF. Spontaneous spinal epidural hematoma in toddlers: description of two cases and review of the literature. *Surg Neurol*. 1994;41(4):325-9.
 8. Hehman K, Norrell H. Massive chronic spinal epidural hematoma in a child. *Am J Dis Child*. 1968;116(3):308-10.
 9. Hosoki K, Kumamoto T, Kasai Y, et al. Infantile spontaneous spinal epidural hematoma. *Pediatr Int*. 2010;52(3):507-8.
 10. Jackson FE. Spontaneous spinal epidural hematoma coincident with whooping cough. Case report. *J Neurosurg*. 1963;20(8):715-7.
 11. Lee JS, Yu CY, Huang KC, et al. Spontaneous spinal epidural hematoma in a 4-month-old infant. *Spinal Cord*. 2007;45(8):586-90.
 12. Licata C, Zoppetti MC, Perini SS, et al. Spontaneous spinal haematomas. *Acta Neurochir (Wien)*. 1988;95(3-4):126-30.
 13. Lim JJ, Yoon SH, Cho KH, et al. Spontaneous spinal epidural hematoma in an infant: a case report and review of the literature. *J Korean Neurosurg Soc*. 2008;44(2):84-7.
 14. Pai SB, Maiya PP. Spontaneous spinal epidural hematoma in a toddler--a case report. *Childs Nerv Syst*. 2006;22(5):526-9.
 15. Patel H, Garg BP. Increasing irritability with sudden onset of flaccid weakness. *Semin Pediatr Neurol*. 1996;3(3):192-7.
 16. Patel H, Boaz JC, Phillips JP, et al. Spontaneous spinal epidural hematoma in children. *Pediatr Neurol*. 1998;19(4):302-7.
 17. Poonai N, Rieder MJ, Ranger A. Spontaneous spinal epidural hematoma in an 11-month-old girl. *Pediatr Neurosurg*. 2007;43(2):121-4.
 18. Posnikoff J. Spontaneous spinal epidural hematoma of childhood. *J Pediatr*. 1968;73(2):178-83.
 19. Ramelli GP, Boscherini D, Kehrl P, et al. Spontaneous spinal epidural hematomas in children: can we prevent a negative prognosis?--reflections on 2 cases. *J Child Neurol*. 2008;23(5):564-7.
 20. Shenkin HA, Horn RC Jr., Grant FC. Lesions of the spinal epidural space producing cord compression. *Arch Surg*. 1945;51(3):125-46.
 21. Vallee B, Besson G, Gaudin J, et al. Spontaneous spinal epidural hematoma in a 22-month-old girl. *J Neurosurg*. 1982;56(1):135-8.
 22. Yasuoka S, Peterson HA, MacCarty CS. Incidence of spinal column deformity after multilevel laminectomy in children and adults. *J Neurosurg*. 1982;57(4):441-5.
 23. Papagelopoulos PJ, Peterson HA, Ebersold MJ, et al. Spinal column deformity and instability after lumbar or thoracolumbar laminectomy for intraspinal tumors in children and young adults. *Spine (Phila Pa 1976)*. 1997;22(4):442-51.
 24. Joaquim AF, Cheng I, Patel AA. Postoperative spinal deformity after treatment of intracanal spine lesions. *Spine J*. 2012;12(11):1067-74.

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