

Case Report

Spontaneous spinal epidural hematoma in a 4-month-old infant

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Study design: A case report and a review of literature.

Objectives: To present the first youngest infant of a 4-month-old boy with spontaneous spinal epidural hematoma in cervicothoracic spine.

Setting: National Cheng Kung University Hospital, Tainan, Taiwan.

Methods: A 4-month-old boy who initially presented with irritable crying, neck stiffness, and fever followed by progressive quadriparesis. Magnetic resonance imaging (MRI) of the spine disclosed a space-occupying lesion on the right posterior–lateral aspect of the cervicothoracic spinal canal. Laminectomy with reconstruction *in situ* from C4 to T4 was performed 5 days after the onset of symptoms.

Results: The boy had gradual improvement of his neurological status. Follow-up visit 1 year later, the infant's growth and development was within normal limit without any neurological deficits; his repeat MRI showed complete fusion of each implanted lamina and well expansion of the spinal cord.

Conclusions: Prompt surgical decompression is valuable, irrespective of the time interval between symptom onset and operation in infant.

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Introduction

Spontaneous spinal epidural hematoma (SSEH) is an uncommon neurological emergency. The incidence is estimated to be 0.1 patients per 100 000 patients per year and most patients are beyond the age of 50 years.^{1,2} The rarity occurrence in children is suggested by limited reports in children and most clinical information are extrapolated from the adult literature. However, based on the current data, there are some obvious differences between the adults and the children with SSEH. In adults, there is a male preponderance with a male/female ratio of 1.5, whereas there is no gender difference in children.^{2,3} Moreover, most of the hematomas are located at the thoracolumbar segment in adults, whereas involvement of the cervicothoracic region is more commonly seen in children.^{2,3} In this paper, we describe the first youngest infant of a 4-month-old boy with SSEH in cervicothoracic spine.

Case report

A 4-month-old boy presented with irritability and crying followed by fever 3 days before admission. The caregivers denied any form of trauma, including shaking movements. The family history was negative for a bleeding disorder. His exam was remarkable for neck stiffness and irritability while turning his head. Deep tendon reflexes were increased over his bilateral lower limbs equally. Urinary analysis disclosed pyuria (WBC > 100/HPF). With the presumptive diagnosis of meningitis and urinary tract infection, he was admitted for lumbar puncture and administration of the antibiotics. The cerebrospinal fluid study was within normal limit. On the following day, the infant appeared to develop a decrease in muscle strength and tone as he lacked the ability to roll his body and continued lying in a supine position. Examination revealed decreased deep tendon reflexes in his bilateral upper limbs with increased reflexes in bilateral lower extremities. Muscle power of four limbs was all decreased, especially in the right upper limb. An emergent spinal magnetic resonance image (MRI) showed signs of cervicothoracic spinal cord compression (Figure 1); spinal epidural hematoma was highly suspected. He underwent laminectomy with reconstruction *in situ* from C4 to T4 to

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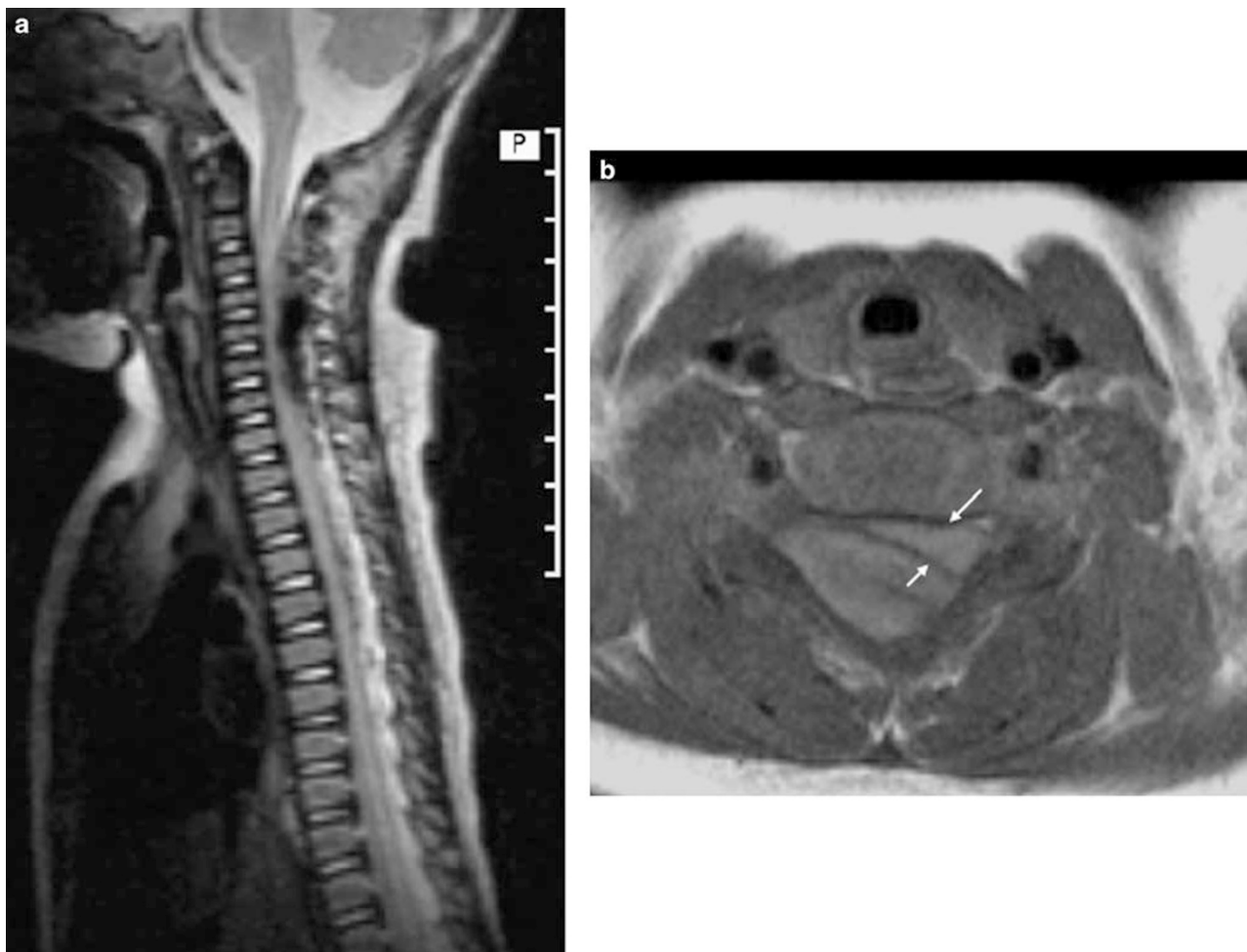


Figure 1 Preoperative MR images. (a) Sagittal T2-weighted image and (b) axial T1-weighted image showing a posterior–lateral lesion with marked spinal cord compression (white arrow) extended from C4–T3

evacuate the epidural hematoma 5 days after the onset of symptoms. Subsequently, the muscle power of his four extremities improved. During his 1-year follow-up period, the infant exhibited normal growth and development without neurological sequelae, while 1-year postoperative MRI showed a well-fused lamina without evidence of cord compression (Figure 2).

Discussion

SSEH is a rare condition in children. In our review of English literature, only eight cases below the age of 2 years had been reported with no age below the first year of life (Table 1).^{4–9} The term ‘crib death’ has been used to describe sudden unexplained death of neonates.¹⁰ In postmortem examination of infants with ‘crib death’, spinal epidural hematomas were the causal factor in 10% of cases.¹¹ Traction and compression force during delivery puts the infant at risk for spinal injury, in which cervical spine is the most site of involvement.

In toddlers, nearly all the reported cases of SSEH involved the cervicothoracic region as in our case.^{4–9} This region is more commonly affected as a result of significant disproportion of the weight ratio of the head to the body in pediatric population, in which the head proportion is larger and heavier than in adults. In addition, gaining the head control is an important neuromuscular development milestone during the first 4–6 months of life. During this period, the control of cervical spine mobility gradually improved. Therefore, the combined effect of the weight of the head and increased cervical mobility in the presence of undeveloped muscular capacity of the neck predispose the cervical spine to injury with sudden flexion and extension movements.

Most authors agree that the rupture of the valveless epidural venous system is the bleeding source of hematoma.^{8–9} The internal vertebral venous plexus is divided into anterior and posterior parts.¹² The anterior part runs fairly constant and is tightly attached to the posterior longitudinal ligament via Hofmann’s liga-

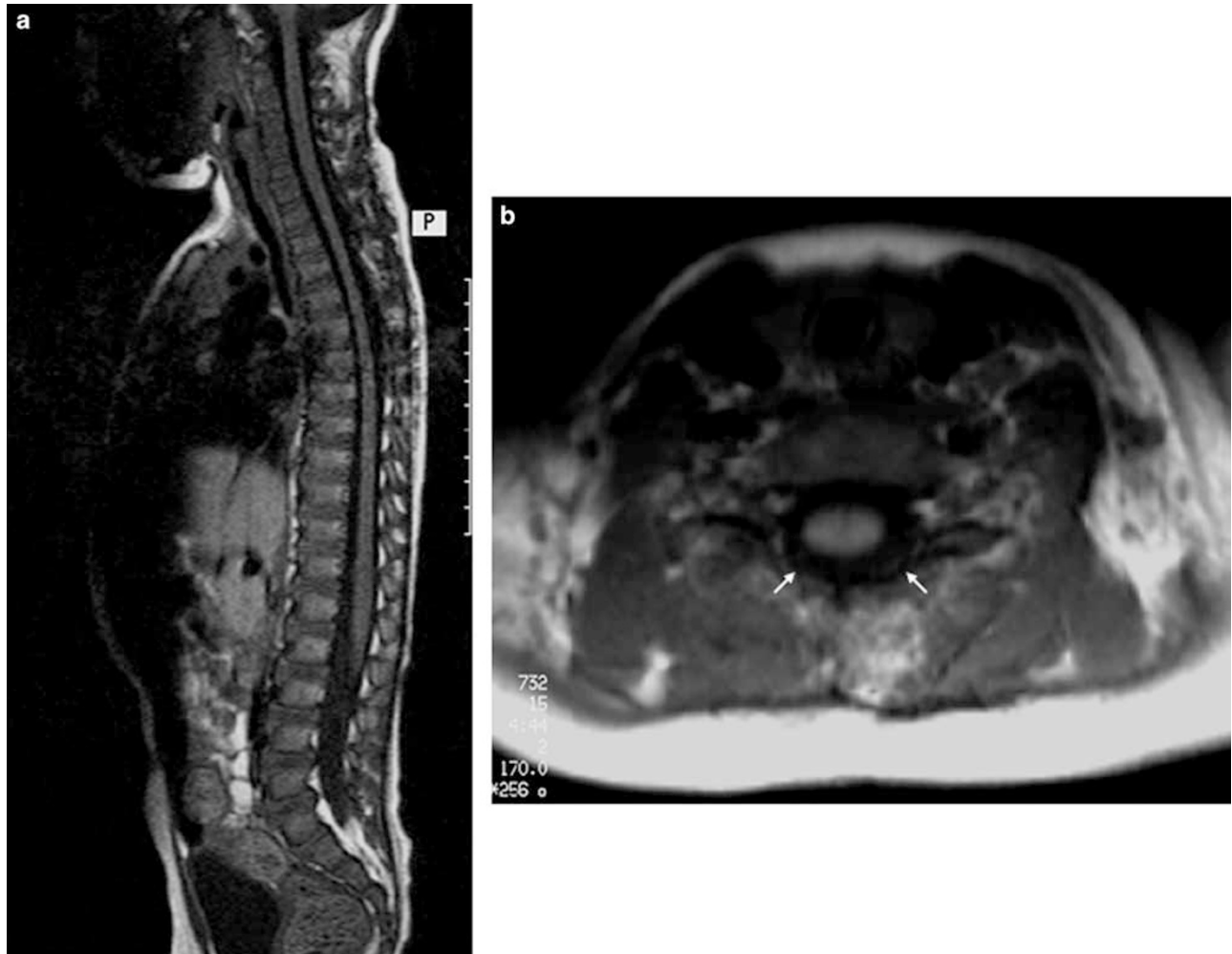


Figure 2 Postoperative MR images, 1 year later. (a) Sagittal T1-weighted image revealing normal curvature of the cervicothoracic segment without spinal cord tethering. (b) Axial sagittal T1-weighted image disclosing rebuilt laminae with fusion to the corresponding vertebra (white arrow) and well expansion of dural sac

ments.^{13,14} On the contrary, the posterior part courses variably and disperses into the loose epidural fat. The sudden elevation of pressure induced by crying, cough, voiding, straining, and trauma can cause a bulk of backflow into the valveless venous system, which makes the loosely supported posterior part of vertebral venous plexus prone to rupture. Thus, the majority of SSEH occurs in the posterior aspect of spinal canal in both adults and children.

The clinical presentations depend on the speed of blood accumulation, location, and length of the hematoma.² Most adult patients complain of a sudden onset of severe pain, sometimes associated with a burning, electric-shock like sensation on the neck or back, followed by ascending numbness and weakness of the legs, which may rapidly develop into paraparesis or paraplegia. The acute neurological deterioration causes them to seek medical consultation immediately. In young children, however, the symptoms and signs highly depend on the observation by their caregivers.

A summary of presenting symptoms and signs from the reported cases includes irritability, abdominal fullness and pain, neck pain, torticollis, rigid neck position, staggering gait, and fever.⁴⁻⁹ The innumerable clinical presentations often result in delaying diagnosis. In our case, the infant initially presented with irritable crying, neck stiffness, and limited range of motion of the neck, followed by neurological deficit as showed by decrease in ability to roll over and decrease limb movements. The occurrence of fever and pyuria can be misleading and point the diagnosis toward an infectious etiology, such as central nervous system infection associated with neurogenic bladder. Lumbar puncture is usually indicated when considering the diagnosis of meningitis. However, placing the patient in a knee-chest position during lumbar puncture may aggravate the spinal cord compression in an undiagnosed spinal epidural hematoma. The important of meticulous clinical observation in infants with SSEH should be kept in mind.

Table 1 Summary of SSEH^a in patients under age of 2 years

Author	Sex/age (months old)	Location	Associated symptoms	Neurological deficits	Interval ^b (days)	Operative method	Outcome
Shenkin <i>et al.</i>	F/20	T1-T7	Fever, irritability	Paraplegia	14	Laminectomy	Good
Jackson FE	F/14	T1-T5	Whooping cough, abdominal distention	Paraparesis	2	Laminectomy	Good
Vallee <i>et al.</i>	F/22	C5-C7	Irritable crying, neck stiffness	Quadripareisis	10	Laminectomy	Good
Licata <i>et al.</i>	M/18	T1-T2	—	Quadripareisis	6	Laminectomy	Good
Caldarelli <i>et al.</i>	F/24	C5-T4	Abdominal pain	Paraplegia	3	Laminotomy	Good
	F/16	C4-C7	Fever, irritability, neck pain	Paraparesis	9	Laminotomy	Good
Patel <i>et al.</i>	M/18	C1-T1	Irritable crying associated with neck movements	Quadripareisis	7	Laminectomy	Good
Pai <i>et al.</i>	M/15	C4-T3	Fever, loss of neck control	Quadripareisis	2	Laminotomy	Good
Present case	M/4	C4-T4	Fever, neck stiffness, pyuria + reconstruction	Quadripareisis Good	5	Laminectomy	Good

^aPatients with obvious history of trauma are excluded

^bTime between the onset of symptoms and operation

MRI is considered to be the diagnostic image modality of choice.^{9,15} The signal intensity on T1-weighted and T2-weighted images varies with the age of hematoma. The former is useful in the follow-up evaluation and the latter is crucial in the diagnosis of SSEH as it provides more information for diagnosis and aides in the differentiation of epidural blood clot from a tumor.¹⁵ In our case, the T1-weighted MRI, displayed isointensity, whereas the T2-weighted MRI disclosed a hypointense foci with iso- to hyperintensity peripherally. This implied that the hematoma contains both deoxy-hemoglobin and methemoglobin, which corresponded to the subacute stage and correlated with the clinical presentation of nearly 5 days.

Without doubt, early surgical decompression is the treatment of choice and most authors agree the time interval between the onset and treatment is the major factor to the functional outcome. The exact time interval had been debatable, with some group proposing 12 h after the onset of symptom, whereas others setting the duration at 48 h.^{2,15,16} In children, however, there is often a delay in diagnosis especially toddlers and infants. The diagnosis of almost all reported case is made after 2 days of presenting symptoms appear. In our review of reported cases in toddlers, most of the cases had complete functional recovery including our patient, suggesting the surgical decompression results in good outcome even with delayed diagnosis.⁴⁻⁹ Laminectomy of the involved segment is the main surgical modality in most cases. However, some authors are concerned that laminectomy will limit normal development in young children, resulting in kyphosis, lordosis, scoliosis or tethered cord. Caldarelli *et al.*⁸ and Pai and Maiya⁹ advocated laminotomy as a good decompression method in children. In our case, we performed the laminectomy with preservation of the lamina in one piece. After removal of the hematoma, the laminae were reconstruction *in situ*. One year after

surgery, follow-up MRI revealed complete fusion of each implanted lamina to the corresponding vertebra (Figure 2b). There is an obvious interface between the dura and paraspinal muscle and no evidence of cord compression, spinal stenosis or defect of laminae suggestive of bone ingrowth on implanted laminae with remodeling.

SSEH is an extremely rare neurological disease in infant. The clinical symptoms and signs are quite diverse which result in difficulty in establishing an early diagnosis. MRI is the imaging modality of choice. Prompt surgical decompression is valuable, irrespective of the time interval between symptom onset and operation, because young children have potentially good recovery.

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