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CASE REPORT

Delayed-onset paralysis induced by spontaneous spinal epidural hematoma communicated with hematoma in the paraspinal muscle in a 6-month-old girl: a case report

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Abstract

Spontaneous spinal epidural hematoma (SSEH) very rarely develops in infants younger than 1 year old. To our knowledge, no previous case of delayed-onset paralysis induced by SSEH communicated with hematoma in the paraspinal muscle has been reported in the literature. The authors present the case of a 6-month-old girl with a tumor mass on her back who developed a paresis of her bilateral lower limbs. On spinal magnetic resonance imaging, the epidural mass appeared to be a dumbbell type and communicated with the mass in the paraspinal muscle through T12/L1 intervertebral foramen at the right side. After excision of the mass in the paraspinal muscle, hemi-laminectomy of T10-L3 was performed. No solid lesion was also present in the spinal canal and it was found to be an epidural hematoma. No malignancy was observed on pathological examination, and vascular and nerve system tumors were negative. When a tumor mass suddenly develops on the back of an infant and motor impairment of the lower limbs develops as the mass gradually enlarges, differential diagnosis should be performed taking SSEH into consideration.

Keywords Infant · Spontaneous spinal epidural hematoma · Spinal cord compression · Dumbbell type

Background

Spontaneous spinal epidural hematoma (SSEH) is a rare disease with an annual incidence of 0.1 in 100,000 people [1, 2]. It may develop at any age, but the peak onset age is 15–20 and 45–75 years old [3]. Its development in infants younger than 1 year old is very rare, and to our knowledge, only nine cases have been reported [4–12]. We report a 6-month-old girl with SSEH and clarify the clinical characteristics of delayed-onset paralysis.

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Case presentation

The patient was a 6-month-old girl whose mother noticed a tumor mass on her back. Six days later, the mother noticed that the child did not move her legs much and brought her to the department of pediatric surgery of our hospital. She demonstrated only weak movement of the lower limbs at hip, knee, and ankle with maximum power of 2/5 in response to sole stimulation, and the deep tendon reflex was normal. On blood testing, no inflammatory reaction was detected, and coagulation factors (PT, APTT, fibrinogen, and platelet count) and tumor markers (AFP, NSE, and sIL-2R) were normal. There was no history of metabolic or hematologic disease, coagulopathies, or coagulation defect tendencies in patient or relatives.

On sagittal spinal magnetic resonance imaging (MRI), a 1.7 × 4.0-cm mass was noted in the paraspinal muscle. Another mass in the spinal canal at the T10-L3 level was also present (Fig. 1A, B). It compressed the spinal cord from the dorsal side. On axial MRI, spinal cord compression was noted in the spinal canal (Fig. 1C, D). It seemed to be a dumbbell type tumor and communicated with the mass in the paraspinal muscle through the T12/L1 intervertebral foramen. Decompression surgery was performed on day 2 after admission. The mass in the paraspinal muscle was a hematoma. After excision of the hematoma, hemi-laminectomy of T10-L3 was performed. The mass in the spinal canal contained no solid lesion and was also a hematoma (Fig. 2). After evacuation of hematoma, it began to bleed from epidural venous plexus. Intraoperative blood loss was 560 ml. The postoperative course was favorable, and the bilateral lower limbs moved in response to sole stimulation on day 2 after surgery. Lower limb muscular strength gradually improved and was completely recovered at 4 weeks after surgery. Histopathological analysis revealed no malignant tumor or vascular malformation. On spinal MRI at 6 weeks after surgery, the spinal cord was sufficiently decompressed, and no recurrence of the mass had occurred (Fig. 3A, B).

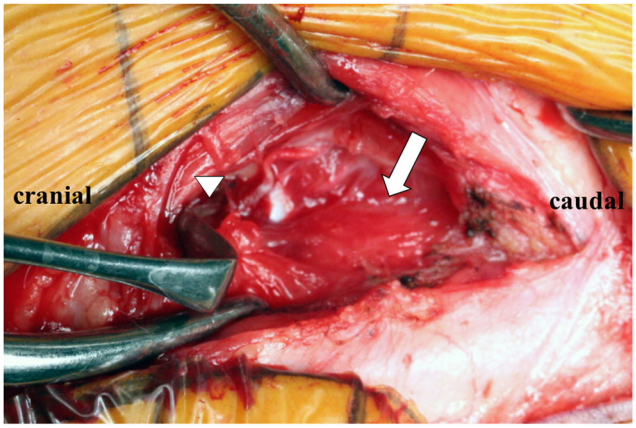

Fig. 2 Intraoperative photograph. The mass in the paraspinal muscle was a hematoma and it was aspirated (arrow). It communicated with the inside of the intervertebral foramen (arrowhead)

Discussion

SSEH is a rare disease with an annual incidence of 0.1 in 100,000 people [1, 2]. The peak onset age is 15–20 and 45–75 years old, and the percentage of male accounted for 64% while female made up 36% [3]. SSEH may develop at any age, but its development in infants younger than 1 year old is very rare and only nine cases have been reported (Table 1) [4–12]. Inducers of SSEH may include blood coagulation abnormality, vascular malformation, pregnancy, tumor hemorrhage, iatrogenic, state of infection, and minor trauma [1, 3, 13]. Groen et al. reported that a certain cause of SSEH was present only in 96 out of 199 cases (48%) [1].

The pathogenesis of SSEH is still unclear. Sudden venous pressure elevation is readily induced by exercise, coughing, and abdominal pressure, and damages blood vessels because of the absence of venous valves [14, 15]. In the current case, no blood coagulation abnormality, vascular malformation, or spinal cord tumor was present, and no cause or inducer of the disease was identified during the surgery or on the pathological examination. However, our patient experienced excessive intraoperative blood loss after evacuation of the hematoma. Possibility of anomaly or vulnerability of spinal vascular cannot be denied.

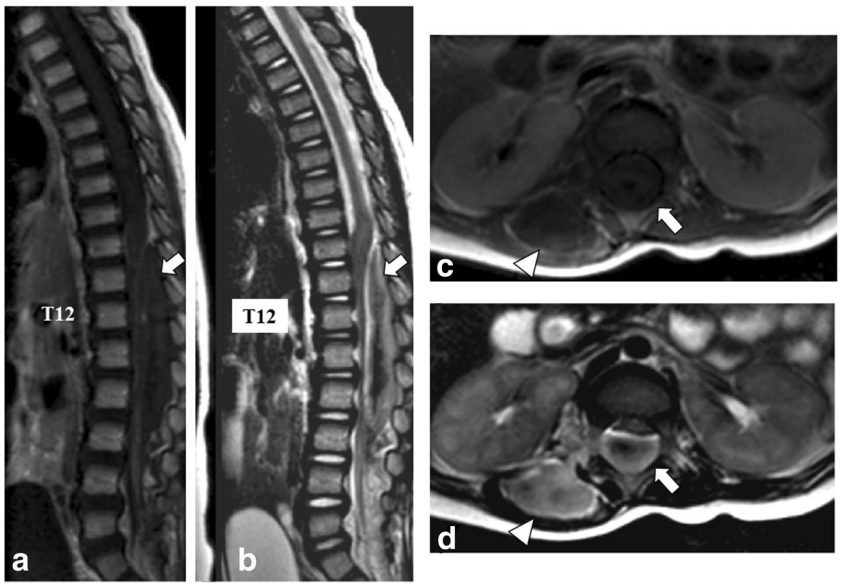

Fig. 1 Preoperative MRI. Sagittal T1-weighted image (**a**), T2-weighted image (**b**), axial T1-weighted image (**c**), and axial T2-weighted image (**d**). The spinal cord was compressed by the hematoma in the spinal canal (arrow). A mass was also present in the paraspinal muscle in the paraspinal muscle (arrowhead)

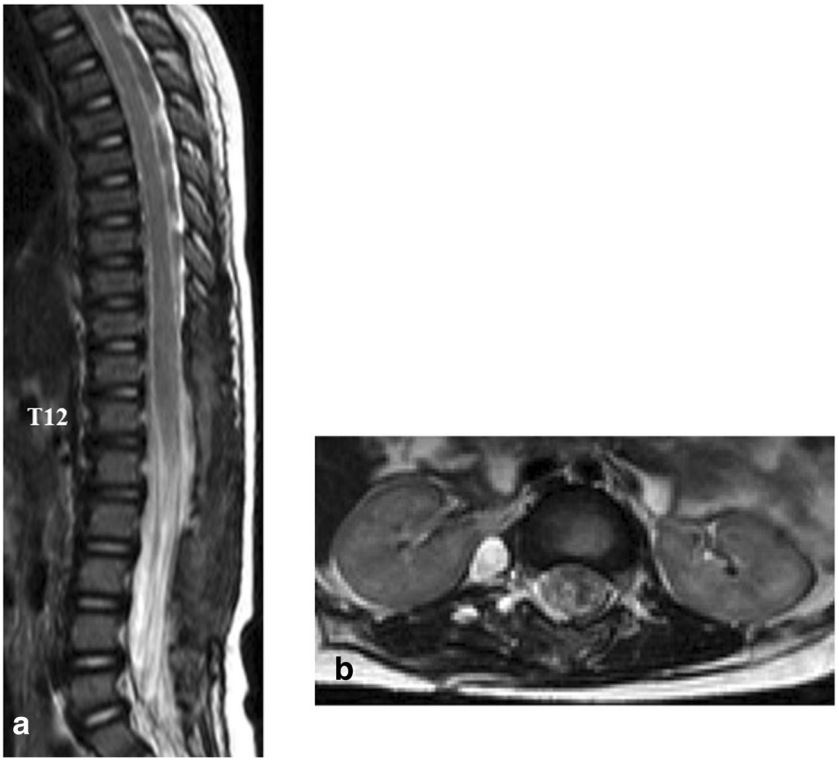

Fig. 3 Postoperative MRI. Sagittal T2-weighted image (**a**) and axial T2-weighted image (**b**). The spinal cord was sufficiently decompressed and no recurrence of the mass was noted

Difficulty in diagnosing SSEH in infants younger than 1 year old is a reason for the small number of reported cases. It has also been reported that the cause of unexplainable sudden death of infants was SSEH in 10% [16, 17]. The most characteristic symptom of SSEH is a sudden sharp pain radiating from the hematoma region to the neck-shoulder and scapula-upper arm when it develops in the cervical lesion, low back pain when it develops in the thoracic-lumbar to lumbar lesion, and motor paralysis and sensory impairment subsequently progress [1, 3]. As it is difficult for infants to tell their symptoms, they may suddenly be in a bad mood or cry [4–12]. Paralysis progresses thereafter, but a definite diagnosis is delayed in many cases. Including current case, 3.5 days to 2 months (mean, 16.2 days) were required to make a definite diagnosis [4–12]. Our case might be acute-on-chronic or chronic spontaneous spinal epidural hematoma because of her normal deep tendon reflex. As for the surgical treatment for SSEH in infants, multilevel decompressive laminectomy should be needed [4–12]. However, postlaminectomy deformity can develop in children without facet injury [18]. We tried not total laminectomy but hemilaminectomy to avoid postlaminectomy deformity, though there was no clear evidence that osteoplastic laminectomy could avoid progressive deformity of the spine [19].

Prognostic factors of SSEH include the severity of neurological manifestation before treatment, presence of spinal edema, and rapid exacerbation of symptoms [20]. As the outcomes of patients with serious neurological manifestation were poor even though they received early surgical evacuation, surgical treatment should be performed before aggravation of neurological manifestations. Groen RJ et al. collected data on 333 cases of SSEH that were reported adequately and confirmed by surgery or autopsy [1]. They concluded that surgical evacuation of SSEH within 36 h in cases involving complete spinal dysfunction and within 48 h in those involving incomplete deficits resulted in favorable outcomes. Paralysis was completely resolved in only five of the ten infants including our case [5, 8, 10, 11] demonstrating that early diagnosis and treatment are key to good neurological recovery.

The initial symptom was the mass on the back and delayed-onset paralysis developed 6 days later. The cause of delayed-onset paralysis may have been the communication between dumbbell type SSEH in the spinal canal and hematoma in the paraspinal muscle through the intervertebral foramen. Inamasu et al. reported that regarding the mechanism of spontaneous absorption of epidural hematoma, the spinal cord can be decompressed by outflow of liquefied hematoma from spinal canal through the intervertebral foramen [21]. The developmental mechanism in the current case was considered to be the following: An epidural hematoma communicated with the paraspinal muscle through the intervertebral foramen and exceeded the limit of expansion in the back. The hematoma no longer had space to escape and expanded in the spinal canal, compressing the spinal cord and causing paralysis. When a tumor mass suddenly develops on the back of an infant and motor impairment of the lower limbs develops as

Table 1 Summary of spontaneous spinal epidural hematoma in infants younger than 1 year old

Author	Sex	Age (month)	Location	Clinical Symptoms	etiology	Neurological deficits	Symptom to operation Interval	Operative method	Outcome
Schoonjans AS et al [4]	male	8	C5-L1	irritability, crying, respiratory ditress	None	paraplesia	4days	laminectomy	incomplete quadriplesia
Min S et al [5]	male	8	C7-T3	irritability, crying	vascular malformation	paraplesia	40days	laminectomy	complete recovery
Hosoki K et al [6]	male	11	T9-L1	irritability	None	paraplesia	3.5 days	hemi-partial laminectomy	partial recovery
Ramelli GP et al [7]	female	7	C6-T7	irritability, crying, respiratory ditress	None	paraplesia	14 days	laminectomy	paraplesia
Lee JS et al [8]	male	4	C4-T4	irritability, crying, fever, neck stiffness	None	quadriparesi s	5 days	laminectomy	complete recovery
Poonai N et al [9]	female	11	C4-T3	irritability, fever	None	paraplesia	2 months	laminoplasty	partial recovery
Liao CC et al [10]	female male	6	C7-T7	irritability, crying, fever, cough	upper respiratory tract infection	complete paraplesia	7 days	laminoplasty	complete recovery
Kalina P et al [11]	male	7	C2-L4	irritability, fussiness, acting uncomfortable	hemophilia	quadriparesi s	N/A	Conservative therapy (Factor VII bolus)	complete recovery
Kim M et al [12]	male	11	C6-T6	irritability	Viral pharyngitis		6 days	Laminectomy	partial recovery
current case	female	6	T10-L3	irritability, back tumor	None		6 days	hemi-partial laminectomy	complete recovery

the mass gradually enlarges, differential diagnosis should be performed taking SSEH into consideration.

Conclusions

We report a 6-month-old girl with SSEH and hematoma in the paraspinal muscle in whom the initial symptom was a tumor mass on the back and paralysis of the bilateral lower limbs developed 6 days later. No case of SSEH communicated with the paraspinal muscle through the intervertebral foramen in an infant has been previously reported. When a tumor mass suddenly develops on the back of an infant and motor impairment of the lower limbs develops as the mass gradually enlarges, differential diagnosis should be performed taking SSEH into consideration.

Authors' contributions

HU performed the study design, analyzed the results, and contributed to the manuscript. YT, MM, MN, HS, and HM participated in the treatment decision. YT made some meaningful suggestions. All authors reviewed and approved the final submitted version.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interests.

Ethics approval and consent to participate This study was approved by the Nihon University Institutional Review Board. Written informed consent was obtained from the patient's parent.

Consent for publication Written informed consent was obtained from the patient's parent for the publication of this manuscript.

Availability of data and materials All data used and analyzed during this study are available from the corresponding author on reasonable request.

Abbreviations *SSEH*, spontaneous spinal epidural hematoma; *MRI*, magnetic resonance imaging

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