J Ped Surg Case Reports 2 (2014) 355–359

Contents lists available at ScienceDirect

Journal of Pediatric Surgery CASE REPORTS

journal homepage: www.jpscasereports.com

Paraplegia induced by mild trauma in a child with thoracic spinal arachnoid cyst☆

Al-Wala Awad [a], Douglas A. Hardesty [b], Krystal Tomei [b], Ratan D. Bhardwaj [b,*]

[a] University of Arizona, College of Medicine-Phoenix, Phoenix, AZ 85004, USA
[b] Division of Neurological Surgery, Barrow Neurological Institute at Phoenix Children's Hospital, Phoenix, AZ, USA

ARTICLE INFO

Article history:
Received 30 June 2014
Received in revised form
11 July 2014
Accepted 11 July 2014

Key words:
Extramedullary intradural arachnoid cysts
Lower extremities paralysis spinal cord compression

ABSTRACT

Spinal arachnoid cysts are rare entities that often present with progressive myelopathy and are treated via surgical excision and fenestration. The acute onset of symptoms from these lesions is not well described in the literature. We report an 18-month-old child with acute onset of paraplegia following a mild trauma, who was found to have a compressive dorsal thoracic intradural spinal arachnoid cyst and emergently treated via surgical decompression and cyst resection. After several months of physical therapy the child achieved meaningful neurologic recovery. Spinal arachnoid cysts can cause acute decompensation in children with serious neurological injury following mild trauma, this risk should be weighed when managing asymptomatic lesions.

Spinal arachnoid cysts (SACs) are rare entities which can arise throughout the spinal column in children or adults. Most commonly these lesions are extra-dural, however intra-dural and even intra-medullary lesions are well described in the pediatric literature which consists mainly of case reports or case series [1–23]. Most often, SACs are found after months or years of symptoms such as progressive lower extremity weakness, back pain, gait spasticity, or other signs of myelopathy, and the management strategy of choice is surgical excision or fenestration of the lesion with restoration of normal cerebrospinal fluid (CSF) flow. Due to the increasing use of routine magnetic resonance imaging (MRI) within the pediatric population, these lesions are at times now found incidentally in an asymptomatic patient; the management strategy and natural history of these lesions are not well-established. The acute onset of symptoms from SACs is exceedingly rare. Here, we report a case of acute paraplegia following mild trauma in a young girl with a dorsal thoracic intradural SAC.

1. Case report

1.1. History and examination

An 18-month-old girl was brought to our hospital's emergency department with acute onset paraplegia. She was otherwise healthy and had developed normally prior to hospitalization. She first walked at approximately 1 year of age and had no gait difficulty prior to presentation. There was no family history of neurological illness or arachnoid cyst.

The evening prior to admission, the child was jumping onto a short plastic children's chair when the seat broke, causing her to fall approximately one foot. She landed first on her feet and immediately fell into a seated position. After the mild trauma, her mother noted some gait clumsiness but the patient was ambulatory. She did not complain of severe back pain. Shortly thereafter she was put to bed.

The next morning, the child awoke unable to move her legs. The family brought her immediately to a local hospital and she was then emergently transported to our facility. We observed flaccid paralysis of the lower extremities and bilateral Babinski signs in our hospital's emergency department. Knee and ankle reflexes were not appreciated. She was otherwise awake, alert, and her upper extremities were full strength. She underwent a spinal MRI that demonstrated a large dorsal heterogenous fluid collection with ventral displacement of the thoracic spinal cord (Fig. 1).

Abbreviations: CSF, cerebrospinal fluid; MRI, magnetic resonance imaging; SAC, spinal arachnoid cyst; SSEP, somatosensory evoked potential.

* Corresponding author. Barrow Neurological Institute at Phoenix Children's Hospital, Ambulatory Building, 4th Floor, 1919 E. Thomas Road, Phoenix, AZ 85016, USA. Tel.: +1 602 933 0196; fax: +1 602 933 0445.
E-mail address: rbhardwaj@phoenixchildrens.com (R.D. Bhardwaj).

2213-5766/$ – see front matter © 2014 The Authors. Published by Elsevier Inc. All rights reserved.
http://dx.doi.org/10.1016/j.epsc.2014.07.006

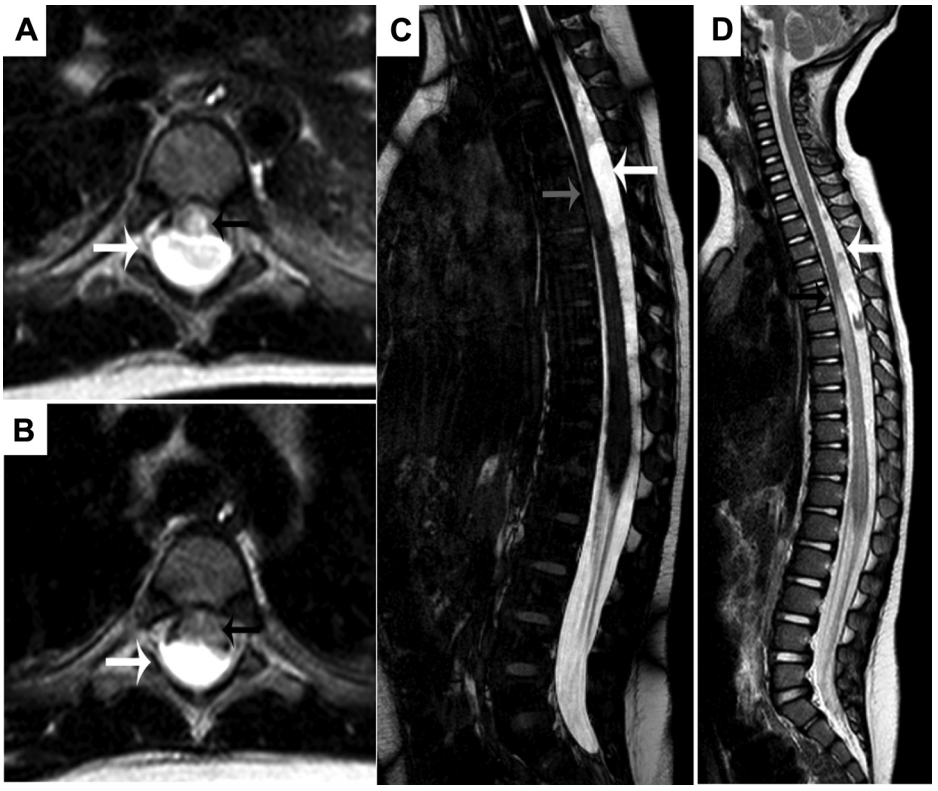

Fig. 1. Axial (A, B) and sagittal (C, D) preoperative T2-weighted MR images demonstrate a dorsal fluid collection consistent with arachnoid cyst (White arrow) in the thoracic spine compressing the spinal cord with early spinal cord (Black/gray arrow) signal change.

Pre-operatively, the cord itself had significant swelling and T2 signal change on MRI. She was taken emergently to the operating room for decompression of the spinal cord in an attempt to maximize functional recovery.

1.2. Operation

Electrophysiological monitoring was established, and initial baseline motor evoked potentials and somatosensory evoked potentials (SSEPs) were absent in the lower extremities prior to skin incision. A standard thoracic laminoplasty was performed, with exposure of T3-T9. The operative microscope was brought into use, and the dura was carefully opened. In the caudal direction of exposure was a thickened arachnoid membrane with clear CSF. There was no evidence of acute or subacute hemorrhage. The venous system of the dorsal spinal cord appeared full and the fluid here was not under significant pressure. However, in the rostral aspect of our exposure, a thinner arachnoid membrane was visualized as well. This was entered sharply, and CSF was expressed under significant pressure. At this rostral site, the dorsal spinal cord vasculature appeared blanched and the cord itself was displaced ventrally (Fig. 2). Upon decompression at this rostral aspect of the lesion, there was a slight improvement in SSEPs, but no return of motor potentials. After resection of the visualized arachnoid membranes and ensuring good CSF flow in all directions, the dura was closed, the lamina were replaced using suture, and the wound was closed in a standard multilayer fashion.

1.3. Postoperative course

Post-operatively, the child returned to the intensive care unit where Mean Arterial Pressure (MAP) was kept elevated (>85 mm Hg) for five days to maximize spinal cord perfusion. An MRI was performed demonstrating excellent decompression (Fig. 3) but with significant progression in spinal cord signal change and edema, consistent with infarct. After one month of intensive

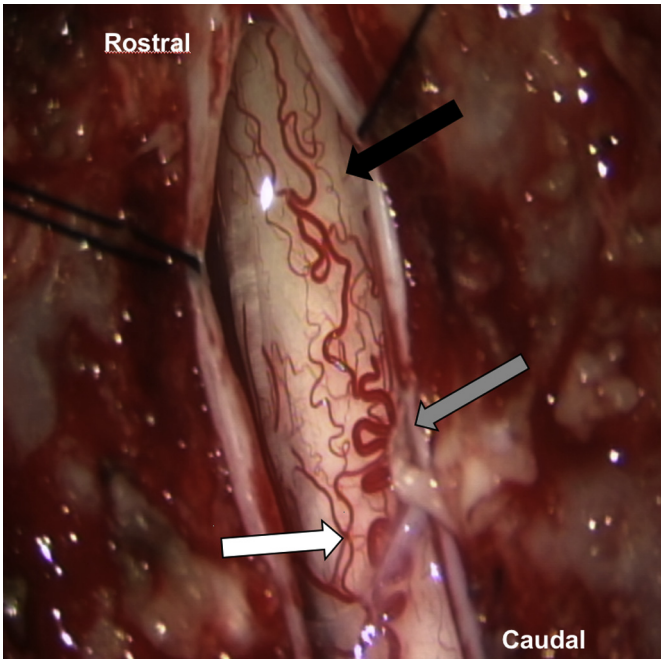

Fig. 2. Intraoperative microscopic view demonstrating operative findings including, paucity of dorsal vessel filling with blanched cord (Black arrow) at site of high-pressure CSF space, remnant of thickened arachnoid membrane caudally (Gray arrow), and caudal cord under the arachnoid cyst with normal blood vessel filling and cord color (White arrow).

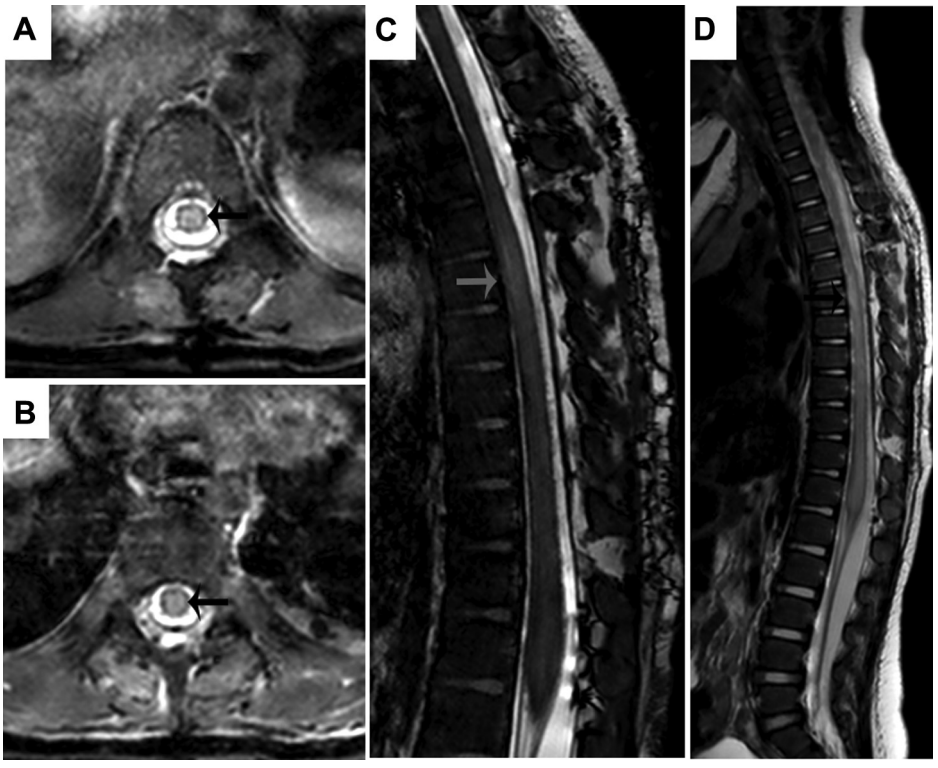

Fig. 3. Axial (A, B) and sagittal (C, D) postoperative T-2 weighted MR images demonstrate excellent decompression of the dorsal fluid collection with restoration of circumferential CSF signal around the spinal cord (Black/gray arrow). Progression of cord edema is noted.

inpatient acute rehabilitation the child was discharged with no return of lower extremity function but with some response to painful stimulus. Subsequently, the patient continued with 2-3x of weekly outpatient physical therapy. After an initial 8-week follow-up, the child regained some spontaneous non-purposeful movement of the lower extremities but no functional strength and remained paraplegic. We continued to monitor the progress of the patient's recovery who has since completed 10 months of 2-3x of weekly outpatient physical therapy. To our surprise, at the most recent follow-up (12 months post injury), the child now has voluntary movements in the bilateral lower extremities including hip flexion. Although the patient continues to be paretic, the patient is also able to stand with assistance, bear weight, and is able to walk short distances with the aid of a walker. Additionally, the child has normal urodynamic testing and exhibits excessive toe biting, suggestive of some sensory function and may be due to dysesthetic pain and paresthesia.

2. Discussion

Spinal arachnoid cysts are uncommon lesions in children that often present with progressive signs and symptoms of myelopathy due to cord compression. Acute symptoms from a previously asymptomatic SAC are exceedingly rare; we found three cases in the literature where a child clearly presented with acute onset (<48 h) of weakness after a mild traumatic injury. One publication reported two cases of patients with ventral cervical SACs presenting with tetraparesis or tetraplegia after mild trauma [17]. Another case series of three patients with SACs described a single pediatric patient with acute onset tetraplegia after mild trauma with a cervicothoracic junction ventral SAC [15]. We found no case reports of dorsal SACs causing acute symptoms after trauma. Overall, the majority of published literature that we reviewed consisted of patients with subacute or chronic symptoms (Table 1).

These lesions are generally considered slow growing, and the etiology of acute onset deficits in children is not obvious. One possibility would be a direct cord contusion from the traumatic insult, in the setting of acquired spinal stenosis from the SAC. This

Table 1
Review of pediatric spinal arachnoid cyst published series.

Author	Year	Total# patients	# of pediatric patients	# patients presenting with weakness	#Pts presenting with acute paraplegia or quadriplegia
Raja	1970	2	0	2	0
Palmer	1974	6	3	5	0
Jensen	1977	1	1	1	0
Herskowitz	1978	1	0	1	0
Fortuna	1983	9	1	8	0
Chan	1985	1	0	1	0
Shih	1990	2	2	2	0
Rabb	1992	11	11	3	0
Chen	1996	1	0	1	0
Kazan	1999	2	1	2	0
Gelabert-González	2001	1	1	1	0
Lee	2001	3	3	3	1
Takahashi	2003	1	1	0	0
Muthukumar	2004	2	2	2	2
Maiuri	2006	1	0	1	0
Gezici	2008	1	1	1	0
Gul	2009	1	1	1	0
Endo	2010	11	1	8	0
Chern	2011	1	1	0	0
Bond	2012	31	31	12	0
Su	2012	1	1	1	0
Evangelou	2013	2	2	2	0

would be akin to a spinal cord injury mechanism similar to that of adult patients with thoracic herniated disc disease who present with acute spinal cord injury after a mild trauma. However, we speculate that a blunt cord injury such as this would be unlikely to occur over multiple levels such as in our patient. Based on the intraoperative findings of a CSF collection under significant pressure, with obvious ongoing cord compression, we suspect a ball-valve mechanism whereby the mild traumatic injury caused a tear within the trabecule of the arachnoid cyst. Unidirectional flow of CSF into such a space then would create an ongoing, expansive mass lesion causing cord compression and ischemia. We believe this theory is more fitting with the intraoperative findings of thickened arachnoid (i.e., the chronic, lower-pressure SAC) below the level of cord compression, and CSF under high pressure within a separate cystic space at the level of cord compression. This theory has also been postulated for the initial formation of SACs, and an exaggerated but similar mechanism via a traumatic tear in the arachnoid could explain acute decompensation [1,3].

As more and more pediatric patients undergo MRI, the incidence of asymptomatic SACs will undoubtedly increase. The optimal management of these asymptomatic lesions is not clear. Some have recommended yearly monitoring with imagining to evaluate for new signal changes and confirm the benign nature of the lesion [24]. However, no consensus exists on the natural history of asymptomatic SACs in the pediatric population, and some may regress or remain stable over years without symptoms.

When SACs present with progressive, chronic symptoms, the choice for surgical decompression to maximize recovery and prevent progression is clear. We found only one case report in which the treating physician opted for conservative management even in the case of mild symptoms [25]. The patient was in his early teens when intermittent symptoms of pain and paresthesiae began, however they were not progressive and underwent no intervention. The patient re-presented as an adult with mild progressive symptoms of pain over the left thigh over the course of one year. An MRI of the patient's spine demonstrated a posterior extradural cysts at the thoracolumbar junction; however due to the mild symptoms the patient and physician did not pursue any surgical intervention and no specific medical treatment was discussed [25]. This was a rare entity and was not associated with trauma; in instances in which there is clear acute progression of neurological injury we believe surgical management remains the responsible choice. In two of the largest series of pediatric patients who were managed with either surgical excision or fenestration, 87–94% had symptomatic improvement or complete remission [1,6]. Open microsurgical fenestration is a more commonly reported procedure over complete excision; however recurrence rates for this procedure ranged from 3 to 12.5% [1,6]. In cases of recurrence, cystoperitoneal shunting has been successfully used as a last report treatment [1,6,13,26]. Recently, minimally invasive endoscopic and MRI guided needle fenestration procedures have been reported [4,5,23]. These procedures have similar outcomes when compared to open surgical intervention, however they have the added benefit of limited blood loss, reduced operative time, and smaller incision and subsequent scar formation [5].

The presented case illustrates the potential for meaningful recovery after acute decompensation and para- or tetraplegia after mild trauma. We were pleasantly surprised by the neurological recovery that was made by our patient after completing several months (1 month inpatient, 7 months outpatient) of aggressive neurorehabilitative therapy. This recovery potential may be unique to the pediatric population. In a large series of severe pediatric spinal cord injuries, functional recovery occurred at higher rates in children (64% vs 31.4%) when compared to adults with similar injury severity at 1 year follow-up [27]. Additionally, functional gains in pediatric patients are dependent on the severity of the initial injury, thus, limiting the acute progression of a neurologic injury is vital for improving outcomes [28]. Following surgical decompression, our patient's MAP was kept elevated (>85 mm Hg) for 5 days to ensure adequate profusion of the spinal cord. Maintaining and elevated MAP after spinal cord injury (SCI) has been shown to improve functional outcomes in patients with SCI, which may be due to limiting the extent of secondary injury caused by ischemia or local profusion changes [29–31].

3. Conclusion

As indicated by our literature review, the onset of acute symptoms following mild spinal trauma in patients with previously asymptomatic spinal arachnoid cyst is exceedingly rare. Although there is clear benefit from surgical intervention in symptomatic patients, the management of asymptomatic patients remains ill-defined. This case however illustrates the potential risk of acute symptomatic progression in this patient population, and makes a compelling case for early and a more aggressive surgical approach to prevent future neurological catastrophe in asymptomatic patients.

Funding and grants

We have no funding or grants to report.

Acknowledgments

We have no acknowledgments to make.

References

[1] Bond AE, Zada G, Bowen I, McComb JG, Krieger MD. Spinal arachnoid cysts in the pediatric population: report of 31 cases and a review of the literature: clinical article. J Neurosurg Pediatr 2012;9:432–41.
[2] Chan RC, Thompson GB, Bratty PJ. Symptomatic anterior spinal arachnoid diverticulum. Neurosurgery 1985;16:663–5.
[3] Chen H-J, Chen L. Traumatic interdural arachnoid cyst in the upper cervical spine: case report. J Neurosurg 1996;85:351–3.
[4] Chern JJ, Gordon AS, Naftel RP, Tubbs RS, Oakes WJ, Wellons JC. Intradural spinal endoscopy in children: report of 4 cases. J Neurosurg Pediatr 2011;8:107–11.
[5] Endo T, Takahashi T, Jokura H, Tominaga T. Surgical treatment of spinal intradural arachnoid cysts using endoscopy: clinical article. J Neurosurg Spine 2010;12:641–6.
[6] Evangelou P, Meixensberger J, Bernhard M, Hirsch W, Kiess W, Merkenschlager A, et al. Operative management of idiopathic spinal intradural arachnoid cysts in children: a systematic review. Childs Nerv Syst 2013;29:657–64.
[7] Fortuna A, Mercuri S. Intradural spinal cysts. Acta Neurochir (Wien) 1983;68:289–314.
[8] Gelabert-González M, Cutrín-Prieto JM, García-Allut A. Spinal arachnoid cyst without neural tube defect. Childs Nerv Syst 2001;17:179–81.
[9] Gezici AR, Ergün R. Cervical anterior intradural arachnoid cyst in a child. Acta Neurochir (Wien) 2008;150:695–8.
[10] Gezici AR, Ergün R. Intramedullary arachnoid cyst: 3-year follow up after bilateral dorsal root entry zone myelotomy. Acta Neurochir (Wien) 2008;150:595–7.
[11] Gul S, Bahadir B, Kalayci M, Acikgoz B. Noncommunicating thoracolumbar intradural extramedullary arachnoid cyst in an 8-year-old boy: Intact removal by spontaneous delivery. Pediatr Neurosurg 2009;45:410–3.
[12] Herskowitz J, Bielawski MA, Venna N, Sabin TD. Anterior cervical arachnoid cyst simulating syringomyelia: a case with preceding posterior arachnoid cysts. Arch Neurol 1978;35:57–8.
[13] Jensen F, Knudsen V, Troelsen S. Recurrent intraspinal arachnoid cyst treated with a shunt procedure. Acta Neurochir (Wien) 1977;39:127–9.
[14] Kazan S, Ozdemir O, Akyüz M, Tuncer R. Spinal intradural arachnoid cysts located anterior to the cervical spinal cord. Report of two cases and review of the literature. J Neurosurg 1999;91:211–5.
[15] Lee HJ, Cho DY. Symptomatic spinal intradural arachnoid cysts in the pediatric age group: description of three new cases and review of the literature. Pediatr Neurosurg 2001;35:181–7.

[16] Maiuri F, Iaconetta G, Esposito M. Neurological picture. Recurrent episodes of sudden tetraplegia caused by an anterior cervical arachnoid cyst. J Neurol Neurosurg Psychiatry 2006;77:1185–6.
[17] Muthukumar N. Anterior cervical arachnoid cyst presenting with traumatic quadriplegia. Childs Nerv Syst 2004;20:757–60.
[18] Palmer JJ. Spinal arachnoid cysts. Report of six cases. J Neurosurg 1974;41:728–35.
[19] Rabb CH, McComb JG, Raffel C, Kennedy JG. Spinal arachnoid cysts in the pediatric age group: an association with neural tube defects. J Neurosurg 1992;77:369–72.
[20] Raja IA, Hankinson J. Congenital spinal arachnoid cysts; report of two cases and review of the literature. J Neurol Neurosurg Psychiatry 1970;33:105–10.
[21] Shih DY, Chen HJ, Lee TC, Chen L. Congenital spinal arachnoid cysts: report of 2 cases with review of the literature. J Formos Med Assoc 1990;89:588–92.
[22] Su DK, Ebenezer S, Avellino AM. Symptomatic spinal cord compression from an intradural arachnoid cyst with associated syrinx in a child: case report. Pediatr Neurosurg 2012;48:236–9.
[23] Takahashi S, Morikawa S, Egawa M, Saruhashi Y, Matsusue Y. Magnetic resonance imaging-guided percutaneous fenestration of a cervical intradural cyst. Case report. J Neurosurg 2003;99:313–5.
[24] Hughes G, Ugokwe K, Benzel EC. A review of spinal arachnoid cysts. Cleve Clin J Med 2008;75:311–5.
[25] Rimmelin A, Clouet PL, Salatino S, Kehrli P, Maitrot D, Stephan M, et al. Imaging of thoracic and lumbar spinal extradural arachnoid cysts: report of two cases. Neuroradiology 1997;39:203–6.
[26] James HE, Postlethwait R. Spinal Peritoneal Shunts for Conditions other than Hydrocephalus and Pseudotumor Cerebri: a Clinical report. Pediatr Neurosurg 2007;43:456–60.
[27] Wang MY, Hoh DJ, Leary SP, Griffith P, McComb JG. High rates of neurological improvement following severe traumatic pediatric spinal cord injury. Spine 2004;29:1493–7.
[28] Garcia RA, Gaebler-Spira D, Sisung C, Heinemann AW. Functional improvement after pediatric spinal cord injury. Am J Phys Med Rehabil 2002;81:458–63.
[29] Vale FL, Burns J, Jackson AB, Hadley MN. Combined medical and surgical treatment after acute spinal cord injury: results of a prospective pilot study to assess the merits of aggressive medical resuscitation and blood pressure management. J Neurosurg 1997;87:239–46.
[30] Blood pressure management after acute spinal cord injury. [Miscellaneous article]. Neurosurg March 2002;2002:50.
[31] Ryken TC, Hurlbert RJ, Hadley MN, Aarabi B, Dhall SS, Gelb DE, et al. The acute cardiopulmonary management of patients with cervical spinal cord injuries. Neurosurgery 2013;72:84–92.