

Compressive Cervicothoracic Adhesive Arachnoiditis following Aneurysmal Subarachnoid Hemorrhage: A Case Report and Literature Review

Gazanfar Rahmathulla^{1,2} Kambiz Kamian²

¹Department of Neurosurgery, Mayo Clinic, Jacksonville, Florida
²Department of Neurosurgery, Cleveland Clinic, Cleveland, Ohio, United States

Address for correspondence Gazanfar Rahmathulla, MD, DNB, Department of Neurosurgery, Cleveland Clinic, 9500 Euclid Avenue S73, Cleveland, OH 44195, United States (e-mail: drgazanfar@gmail.com).

J Neurol Surg Rep 2014;75:e56–e61.

Abstract

We present the case of a 55-year-old woman with diffuse adhesive arachnoiditis in the posterior fossa and cervicothoracic spine following posterior inferior cerebellar artery aneurysmal subarachnoid hemorrhage (SAH). She underwent aneurysm clipping with subsequent gradual neurologic decline associated with sensory disturbances, gait ataxia, and spastic paraparesis. Magnetic resonance imaging revealed diffuse adhesive arachnoiditis in the posterior fossa and cervicothoracic spine, syringobulbia, and multiple arachnoid cysts in the cervicothoracic spine along with syringohydromyelia. Early surgical intervention with microlysis of the adhesions and duraplasty at the clinically relevant levels resulted in clinical improvement. Although adhesive arachnoiditis, secondary arachnoid cysts, and cerebrospinal fluid flow abnormalities resulting in syrinx are rare following aneurysmal SAH, early recognition and appropriate intervention lead to good clinical outcomes.

Keywords

- ▶ aneurysmal subarachnoid hemorrhage
- ▶ adhesive arachnoiditis
- ▶ arachnoid cysts
- ▶ syringohydromyelia
- ▶ syringobulbia

Introduction

Aneurysmal subarachnoid hemorrhage (SAH) is rarely associated with the development of chronic diffuse spinal arachnoiditis.^{1,2} Adhesive arachnoiditis is commonly associated with abnormalities that include the development of a syrinx in the craniospinal axis and arachnoid cysts with mass effect on the cord, possibly at multiple sites giving rise to a variety of gradually progressing neurologic deficits. Although surgery, trauma, and meningitis are the more commonly known causes of adhesive arachnoiditis, hemorrhage appears to cause a severe inflammatory reaction that also results in arachnoiditis at any level of the neuraxis.^{3,4} There have been ~ 15 reported cases of adhesive arachnoiditis following SAH with variable neurologic signs, timing of treatment, type of intervention, and outcomes ranging from poor to exceptionally good neurologic recovery.^{1,5–8}

We present our case of a 55-year old woman operated on for a posterior inferior cerebellar aneurysm (PICA) followed

almost a year later with extensive craniovertebral (CV) junction and cervicothoracic spine involvement with arachnoiditis, arachnoid cysts, and syringomyelia. Surgical intervention at the CV junction, cervical, and thoracic spinal levels with microlysis of adhesions resulted in a regression of symptoms and signs. This is the first case of arachnoid cysts, syrinx, and arachnoiditis to our knowledge where early detection, decompressive surgery, and microlysis at two independent levels led to stabilization and recovery of the syringobulbia, syringohydromyelia with complete neurologic recovery. It is important to recognize this late complication of SAH. Early diagnosis and intervention can lead to stabilization and improvement of this benign pathology.

Case Report

A 54-year-old woman had presented to an outside hospital following rupture of a right PICA with SAH for which she

received
 September 1, 2013
 accepted
 November 3, 2013
 published online
 January 16, 2014

DOI <http://dx.doi.org/10.1055/s-0033-1363506>.
 ISSN 2193-6358.

© 2014 Georg Thieme Verlag KG, Stuttgart · New York

License terms



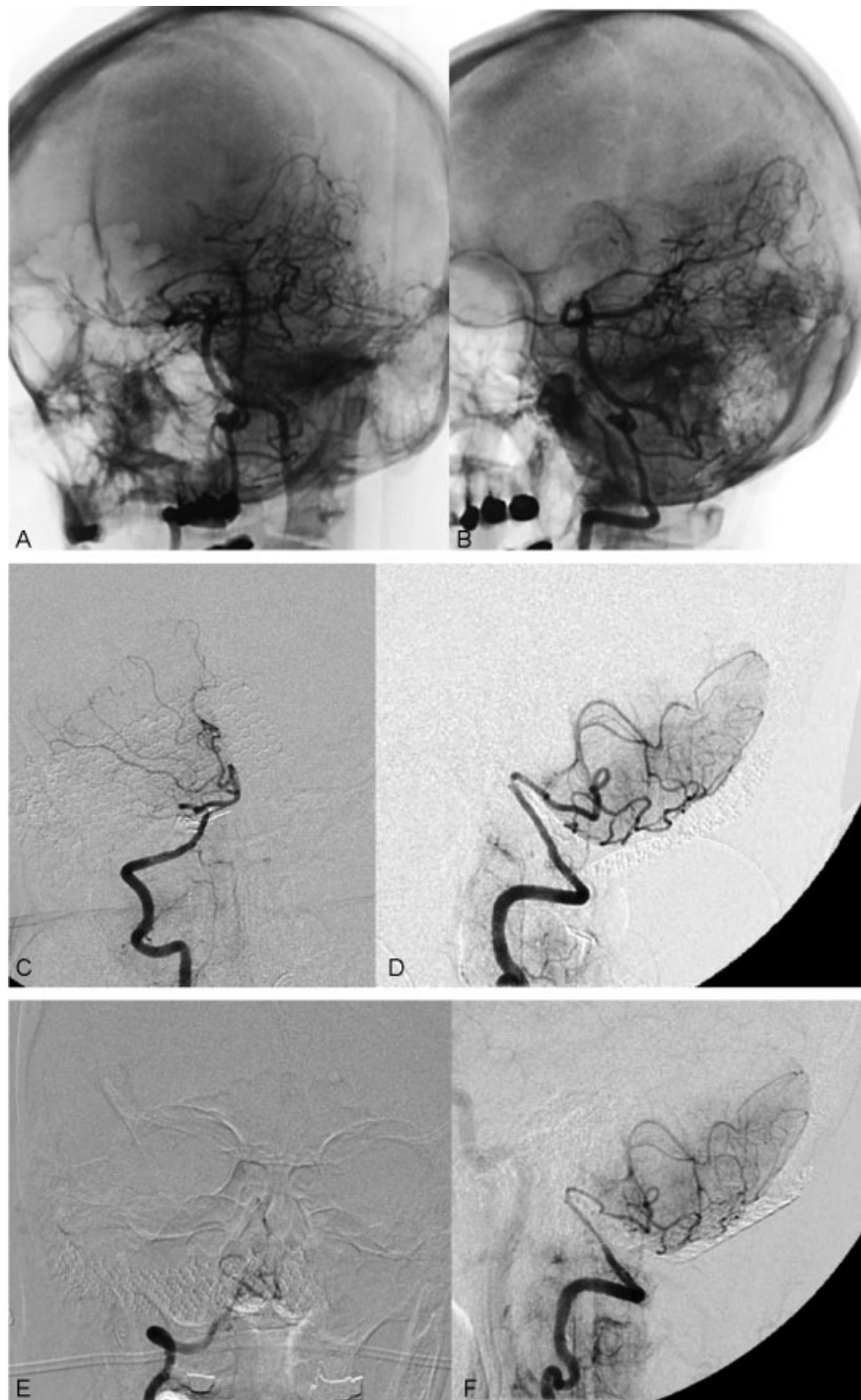


Fig. 1 Preoperative digital subtraction angiogram (DSA) images in the arterial phase. (A) Anteroposterior (AP) and (B) lateral (Lat) views following contrast injection of the right vertebral artery. The DSA reveals the presence of a bilobed aneurysm arising from the right posterior inferior cerebellar artery with no evidence of cerebral vasospasm. (C and D) Postoperative DSA images in (C) AP and (D) Lat views postcontrast show complete clipping of the posterior inferior cerebellar aneurysm with the clip in situ (dotted oval white line outlining the aneurysm clip), no evidence of residual filling, and the absence of vasospasm. (E and F) Postoperative DSA images one week after the aneurysm clipping revealing angiographic evidence of vasospasm in the posterior circulation on the (E) AP angiogram and (F) the lateral view of the posterior circulation on the angiogram.

underwent clipping (►Fig. 1A–D). The SAH was clinically a Hunt and Hess grade 2⁹ with a Fisher grade 3¹⁰ reported on her computed tomography (CT) scan. Her postoperative course was complicated by clinical and angiographic cerebral vasospasm with communicating hydrocephalus a week following surgery (►Fig. 1E, F). She underwent the insertion of a ventriculostomy and following its removal required insertion

of a ventriculoperitoneal (VP) shunt. Shortly after the shunt insertion she developed cerebral venous thrombosis confirmed with a magnetic resonance venogram (MRV) and was started on warfarin. She was discharged from the hospital almost 5 weeks subsequent to her initial surgery with a good recovery. About 4 months later she began to experience a decline in her motor ability, with spasticity, weakness in her

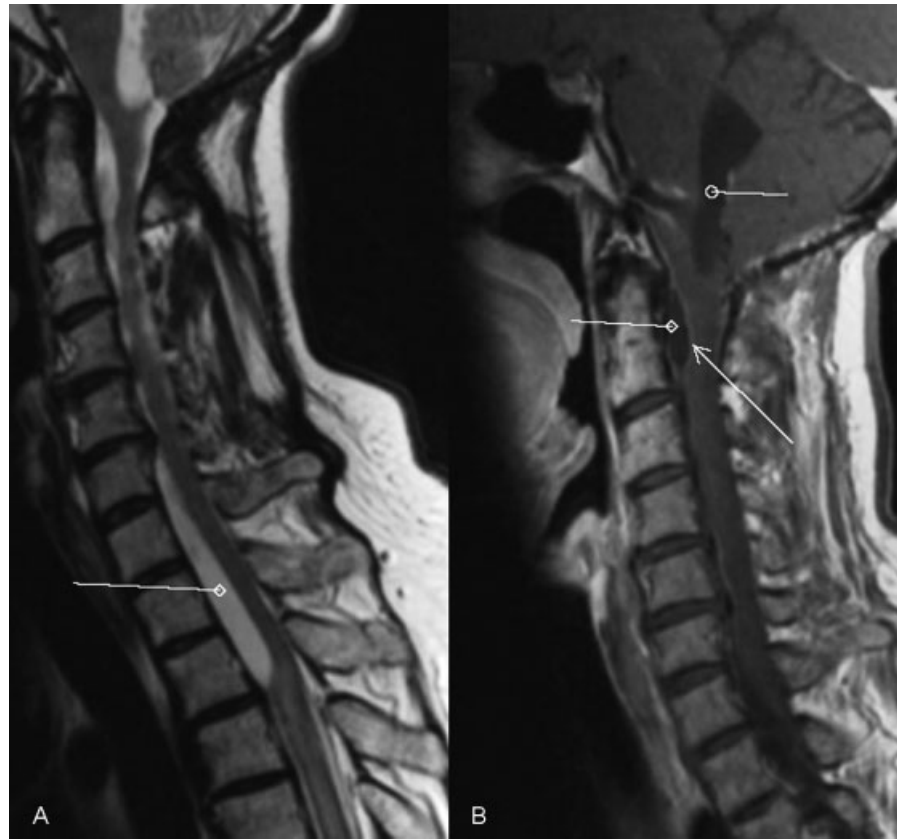


Fig. 2 (A) Preoperative T2-weighted sagittal image of the craniovertebral junction and cervicothoracic spine revealing dilatation of the inferior fourth ventricle with central canal enlargement along with ventral cysts at cervical (C1–2) and cervicothoracic cord (arrow with diamond head). (B) T1-weighted sagittal images postcontrast revealing dilatation of the inferior fourth ventricle (circular arrowhead) with a ventrally placed cyst at C1–2 (diamond head arrow) and enhancement of the adhesions along the ventral aspect of the cervical and upper thoracic cord (open arrowhead).

lower extremities, and requiring assistance to walk. Gradually over the year this progressively worsened, and she required a cane and then a walker to help her ambulate. She also developed progressive unsteadiness along with numbness in her upper and lower extremities. She underwent cranial imaging with a CT scan twice, both times reported as normal.

She presented to us about a year from her initial surgery, and clinical examination revealed hypoesthesia around her lips with spasticity in her lower extremities. She had a motor power of 4+/5 (British Medical Research Council grading)¹¹ in all her extremities. Her upper extremity reflexes were brisk with a positive Hoffman sign. Lower extremity reflexes were exaggerated with a clonus and bilateral upward plantar response. Her CT scan revealed a VP shunt in situ with normal lateral and third ventricles and a dilatation of the lower fourth ventricle along with the central canal of the upper cervical cord. A magnetic resonance imaging (MRI) scan of the craniospinal axis revealed dilatation of the fourth ventricle with compression at the CV junction and the adjacent C1 and C2 levels, and a contrast-enhancing cyst wall at the C1–2 and 5–6 levels with a mass effect on the cervical spinal cord (►Fig. 2A, B). The cervicothoracic scan revealed syringohydromyelia extending upward from the T4–5 level, with ventral cysts and adhesions causing cord compression (►Fig. 3A, B).

The patient underwent a staged decompression at two levels, above at the CV junction and cervical spine (C1–2) and

below at the site of maximal thoracic compression. The posterior fossa craniectomy was reexplored and extended by further resection of the occipital bone across the midline, and the C1 posterior arch was also removed and a C2–C3 laminectomy performed. The dura was opened in a standard Y-shaped manner and densely adherent to the underlying cerebellum and cervical cord. Under the operating microscope, the scarring was grossly visible; evidence of sequelae from arachnoiditis and adhesions was covering the nerve roots, tonsils, and aneurysm clip as well as the PICAs bilaterally. We performed microlysis of the adhesions and intraoperative spinal cord monitoring using somatosensory evoked potentials to identify and avoid any neurologic deficits at the time of adhesiolysis and microdissection. Once the tonsils were released, the fourth ventricle was opened and CSF under high pressure escaped. The large arachnoid cyst at C1–2 compressing the right side of the spinal cord was then dissected and other smaller cysts released as well. Hemostasis was then achieved and a duraplasty was done. Fascia lata was harvested from the right lower extremity and used for the duraplasty that was layered over with DuraGen and Tisseel (fibrin glue) and subsequently layered muscle closure.

About 2 weeks later we performed a standard decompressive laminectomy in the thoracic region extending from T2 to T4. The laminectomy was extended laterally using a modified posterolateral approach to reach the ventral



Fig. 3 (A) Preoperative T2-weighted sagittal image of the thoracic spine revealing dilatation, a cord syrinx with ventral loculated cysts, and adhesions tethering the cord anteriorly and compressing it at the same time (diamond head arrows). (B) T1-weighted sagittal postcontrast images revealing enhancement of the fibrous bands (adhesions) tethering the cord and patchy enhancement of the cyst wall margins (open arrowheads).

arachnoid cyst, one of which had its inferior limit at T2 and another smaller cyst anterior to T3. The cysts were fenestrated and marsupialized. Then intradural microlysis of the adhesions was done using continuous intraoperative neurophysiologic monitoring to avoid injury to the cord while performing the procedure. Watertight closure of the dura was done using 5-0 Prolene and layered with Tisseel, followed by layered closure of the muscle and fascia along with a noninstrumented bony fusion across the operated level.

The patient's recovery following surgery was uneventful with no immediate changes in her neurologic status. She was discharged and her postoperative imaging 6 months following surgery revealed decompression of the posterior fossa and upper cervical and thoracic spine with absence of cystic compression and resolution of the cervicomedullary and thoracic syrinx (→Fig. 4A, B). Her gait, balance, motor, and sensory symptoms had gradually improved at her 18-month follow-up.

Discussion

Numerous reports documenting adhesive arachnoiditis have shown that the risk is significantly higher following rupture of posterior circulation aneurysms when compared with anterior

circulation SAH.^{8,12-14} Although speculation regarding its occurrence has been made in relation to the volume of subarachnoid blood⁶ and the possibility of an associated prolonged chronic inflammatory reaction^{15,16} of the pia-arachnoid, the number of cases remains particularly low in spite of the large aneurysm volumes treated at major tertiary centers. Other mechanisms proposed for the diffuse arachnoiditis seen in this small group of patients have varied from prolonged bed rest, the need for cerebrospinal fluid (CSF) diversion, prolonged intensive care unit stay,¹ the use of fibrin glue,⁸ occult meningitis, and collapse of the subarachnoid space causing adhesions to the pia secondary to multiple lumbar punctures.^{15,16} The arachnoid cysts associated with adhesive arachnoiditis have been seen following both aneurysm clipping as well as endovascular coiling.¹⁷ The sequelae following SAH may lead to a diffuse chronic inflammatory state, fibroproliferation, and possible vasospasm causing adhesion and loculation,^{1,18} with more evidence observed with published data from anesthesiology following epidural patch procedures.^{19,20} Although these cysts are more commonly seen with trauma, infection, or following surgery, the fibrosis and adhesions ensuing SAH tend to loculate CSF into pockets. The continuous production of CSF via the adjacent membranes causes an increase in their size, followed by their mass effect and neurologic symptoms.²¹ An alternative hypothesis put



Fig. 4 Postoperative magnetic resonance images at 6 months following surgery. (A) T1-weighted sagittal image of the craniovertebral junction revealing a wide decompression of the posterior fossa and upper cervical canal with diminished fourth ventricular size, absence of upper cord compression by the cyst, and no visible tethering by adhesions (diamond arrowheads). (B) T2-weighted sagittal image of the cervicothoracic spine showing posterior decompression at the cervical and upper thoracic spine (diamond arrowheads) without cord compression, and resolution of the mass effect of the cyst and syrinx.

forward for the increase in the size of these cysts is the possibility of a unilateral valve mechanism, with gradual inflow and no egress of CSF from these spaces.²² With a disruption in CSF pathways secondary to fibrosis, adhesions, cyst formation, and its mass effect on the cord, an imbalance of CSF circulation effectively results in the formation of a syrinx.^{23–25}

Ventral cord compression is usually associated with motor symptoms, and sensory symptoms such as paresthesias are seen when patients have associated syrinx cavities.^{1,2,8,13} Kok et al¹ identified patients with posterior circulation SAH, those requiring CSF diversion procedures, and those having undergone long-term intensive care as factors resulting in a higher risk of developing diffuse arachnoiditis with or without associated cysts. There have been ~ 15 patients with diffuse arachnoiditis described in the literature; 7 cases were associated with an arachnoid cyst.²

Our patient had many of the factors known to incite the development of diffuse arachnoiditis including prolonged bed rest, a high volume of subarachnoid blood, ventriculostomy insertion, and VP shunting. With clinical and radiologic evidence of obstruction of the subarachnoid CSF pathways at multiple levels (CV junction, upper cervical and thoracic spine), we did not feel the need to investigate with further imaging such as CSF flow studies (Cine-MRI scans) or CT myelogram to identify the level of CSF flow obstruction. The insertion of shunts to divert CSF flow has only shown short-term improvements, and unfavorable outcomes in the long term with syrinx recurrence rates being high.²⁵ Our strategy was to directly tackle the most compressed levels of arachnoiditis at both the CV junction and thoracic spine with

adequate decompression by performing a microlysis of the adhesions, decompression of the cysts, and a duraplasty with an autologous fascia lata graft at the cervical level. We prefer autologous grafts in our practice because in our practice we have seen the use of other materials appears to incite an intense inflammatory reaction that we wanted to avoid. Prior cases have been treated with microlysis, expansive laminoplasty and duraplasty^{6,26} with good outcomes. Because of the rare nature of this pathology, there is no uniform consensus on the best treatment strategy, and the surgical approach has to be individualized, taking into consideration the clinical presentation and neuroimaging findings.

Conclusion

Spinal arachnoiditis, arachnoid cysts, and syringomyelia following aneurysmal SAH is a rare entity. Clinicians treating these patients should have a high degree of suspicion, especially when they present with an undiagnosed progressive neurologic decline. Early diagnosis and targeted intervention may be the key strategy to prevent late complications of SAH by directly relieving compression. Attempting to normalize CSF flow dynamics may obviate the neurologic sequelae in these difficult cases.

References

- 1 Kok AJ, Verhagen WI, Bartels RH, van Dijk R, Prick MJ. Spinal arachnoiditis following subarachnoid haemorrhage: report of two cases and review of the literature. *Acta Neurochir (Wien)* 2000; 142(7):795–798; discussion 798–799

- 2 Tumialán LM, Cawley CM, Barrow DL. Arachnoid cyst with associated arachnoiditis developing after subarachnoid hemorrhage. Case report. *J Neurosurg* 2005;103(6):1088–1091
- 3 Klimo P Jr, Kestle JR, MacDonald JD, Schmidt RH. Marked reduction of cerebral vasospasm with lumbar drainage of cerebrospinal fluid after subarachnoid hemorrhage. *J Neurosurg* 2004;100(2):215–224
- 4 Sajanti J, Majamaa K. Detection of meningeal fibrosis after subarachnoid haemorrhage by assaying procollagen propeptides in cerebrospinal fluid. *J Neurol Neurosurg Psychiatry* 1999;67(2):185–188
- 5 Augustijn P, Vanneste J, Davies G. Chronic spinal arachnoiditis following intracranial subarachnoid haemorrhage. *Clin Neurol Neurosurg* 1989;91(4):347–350
- 6 Ishizaka S, Hayashi K, Otsuka M, et al. Syringomyelia and arachnoid cysts associated with spinal arachnoiditis following subarachnoid hemorrhage. *Neurol Med Chir (Tokyo)* 2012;52(9):686–690
- 7 Klekamp J, Iaconetta G, Batzdorf U, Samii M. Syringomyelia associated with foramen magnum arachnoiditis. *J Neurosurg* 2002;97(3, Suppl):317–322
- 8 Taguchi Y, Suzuki R, Okada M, Sekino H. Spinal arachnoid cyst developing after surgical treatment of a ruptured vertebral artery aneurysm: a possible complication of topical use of fibrin glue. Case report. *J Neurosurg* 1996;84(3):526–529
- 9 Hunt WE, Hess RM. Surgical risk as related to time of intervention in the repair of intracranial aneurysms. *J Neurosurg* 1968;28(1):14–20
- 10 Fisher CM, Kistler JP, Davis JM. Relation of cerebral vasospasm to subarachnoid hemorrhage visualized by computerized tomographic scanning. *Neurosurgery* 1980;6(1):1–9
- 11 Medical Research Council. Aids to the Investigation of Peripheral Nerve Injuries (War Memorandum No. 7). 2nd ed. London, UK: His Majesty's Stationery Office; 1943
- 12 Brodbelt AR, Stoodley MA. Syringomyelia and the arachnoid web. *Acta Neurochir (Wien)* 2003;145(8):707–711; discussion 711
- 13 Eneling J, Boström S, Rossitti S. Subarachnoid hemorrhage-associated arachnoiditis and syringomyelia. *Clin Neuroradiol* 2012;22(2):169–173
- 14 Tjandra JJ, Varma TR, Weeks RD. Spinal arachnoiditis following subarachnoid haemorrhage. *Aust N Z J Surg* 1989;59(1):84–87
- 15 Koyanagi I, Iwasaki Y, Hida K, Houkin K. Clinical features and pathomechanisms of syringomyelia associated with spinal arachnoiditis. *Surg Neurol* 2005;63(4):350–355; discussion 355–356
- 16 Sercombe R, Dinh YR, Gomis P. Cerebrovascular inflammation following subarachnoid hemorrhage. *Jpn J Pharmacol* 2002;88(3):227–249
- 17 Marcoux J, Roy D, Bojanowski MW. Acquired arachnoid cyst after a coil-ruptured aneurysm. Case illustration. *J Neurosurg* 2002;97(3):722
- 18 Choi JU, Kim DS. Pathogenesis of arachnoid cyst: congenital or traumatic? *Pediatr Neurosurg* 1998;29(5):260–266
- 19 Aldrete JA. Neurologic deficits and arachnoiditis following neuroaxial anesthesia. *Acta Anaesthesiol Scand* 2003;47(1):3–12
- 20 Aldrete JA, Brown TL. Intrathecal hematoma and arachnoiditis after prophylactic blood patch through a catheter. *Anesth Analg* 1997;84(1):233–234
- 21 Basaldella L, Orvieto E, Dei Tos AP, Della Barbera M, Valente M, Longatti P. Causes of arachnoid cyst development and expansion. *Neurosurg Focus* 2007;22(2):E4
- 22 Rohrer DC, Burchiel KJ, Gruber DP. Intraspinous extradural meningeal cyst demonstrating ball-valve mechanism of formation. Case report. *J Neurosurg* 1993;78(1):122–125
- 23 Caplan LR, Norohna AB, Amico LL. Syringomyelia and arachnoiditis. *J Neurol Neurosurg Psychiatry* 1990;53(2):106–113
- 24 Chang HS, Joko M, Matsuo N, Kim SD, Nakagawa H. Subarachnoid pressure-dependent change in syrinx size in a patient with syringomyelia associated with adhesive arachnoiditis. Case report. *J Neurosurg Spine* 2005;2(2):209–214
- 25 Chang HS, Nakagawa H. Theoretical analysis of the pathophysiology of syringomyelia associated with adhesive arachnoiditis. *J Neurol Neurosurg Psychiatry* 2004;75(5):754–757
- 26 Ohata K, Gotoh T, Matsusaka Y, et al. Surgical management of syringomyelia associated with spinal adhesive arachnoiditis. *J Clin Neurosci* 2001;8(1):40–42