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Journal of Craniovertebral Junction and Spine

Case Report

Syringomyelia and spinal arachnoiditis resulting from aneurysmal subarachnoid hemorrhage: Report of two cases and review of the literature

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Journal of Craniovertebral Junction and Spine 2014, 5:10

Abstract

Syringomyelia resulting from arachnoiditis secondary to aneurysmal subarachnoid hemorrhage (SAH) is an extremely rare clinical entity with few cases reported in the literature. The presentation, management, and pathogenesis of syringomyelia in this setting is poorly understood. We describe the presentation, radiology, management, and outcomes in two patients with syringomyelia resulting from arachnoiditis secondary to aneurysmal SAH and review the literature on this rare condition. Case number 1 was treated successfully with syrinx-subarachnoid shunt after extensive lysis of adhesions. Case number 2 was treated with syringoperitoneal shunt. Both patients had radiographic decreased syrinx size postoperatively. These patients add to the small literature on syringomyelia occurring secondary to SAH-associated arachnoiditis. The radiographic outcomes demonstrate that in the appropriately selected patient, syrinx-subarachnoid or syringoperitoneal shunting are viable options.

Key words: Paraplegia, spinal arachnoiditis, subarachnoid hemorrhage, syringomyelia

INTRODUCTION

Aneurysmal subarachnoid hemorrhage (SAH) is a devastating neurosurgical condition with significant morbidity for up to 50% of survivors.[1] An extremely rare complication of aneurysmal SAH is delayed spinal arachnoiditis that can occur months to years after the causative SAH.[2,3] Post-SAH spinal arachnoiditis was first described in 1943 by Nelson,[4] but several more cases have been reported since that time.[2,3,5-7] Even more rare than SAH-associated arachnoiditis is the development of syringomyelia secondary to arachnoiditis after aneurysmal SAH.[8] Due to the rarity of post-SAH syringomyelia, little is known about its presentation, pathogenesis, and management. In this report, we describe two cases of post-SAH spinal arachnoiditis with holochord syringomyelia and review the literature on this rare condition.

Access this article online	
Quick Response Code:	Website: www.jcvjs.com
	DOI: 10.4103/0974-8237.135227

CASE REPORTS

Case 1

History and physical

A 66-year-old retried lawyer presented for evaluation of known cervicothoracic syringohydromyelia. Eleven years prior to his presentation he suffered a SAH from a right fusiform vertebral artery aneurysm, which was clipped through a right suboccipital approach. After his SAH, he was hospitalized for 2 months and was in a coma for more than a month. Eight years after his SAH, he developed neck pain that lead him to obtain magnetic resonance imaging (MRI) demonstrating a cervicothoracic syrinx. He was seen by several neurosurgeons and eventually referred to the senior author (A.H.M.) University of Iowa Hospitals and Clinics.

On examination, he kept his neck flexed. Extraocular movements were full and his gag response was absent bilaterally. He had normal strength in his bilateral upper and lower extremities. Sensation was diminished to pain and light touch in the right neck, right trunk, and right upper extremity, but otherwise sensation was intact throughout. There were no pathologic reflexes. Imaging demonstrated a syringohydromyelia extending from the C1 level into the thoracic spine [Figure 1a]. Given stable imaging over several years and stable symptoms of neck pain and right upper extremity sensory change, a decision was made to follow him closely and monitor the syrinx.

One year later, the patient returned with difficulty ambulating, requiring the assistance of a walker over the previous 2 weeks. He had new bowel incontinence. Sensation was diminished in both lower extremities and right upper extremity. Strength was 4/5 in the right upper extremity and 3/5 in the right lower extremity. He also had 4/5 left hamstring strength, but otherwise all muscle groups on the left side were intact. Computerized tomographic myelogram via lumbar puncture was obtained and demonstrated cessation of contrast flow at the T9 level. Follow-up MRI demonstrated an increase in the syrinx from the levels of C1-C7 [Figure 1b].

Operation

Given progression of symptoms, the patient underwent C5-C6 osteoplastic laminotomies. The dura was opened with care to preserve the underlying arachnoid membrane. The arachnoid was grossly abnormal and appeared white, opaque, and unusually vascular [Figure 2a]. The dense "arachnoid" was opened and lysis of adhesions was performed [Figure 2b]. The subarachnoid space was opened, allowing placement of a syrinx to subarachnoid shunt [Figure 2c]. A midline myelotomy was performed to expose the syrinx cavity and the syrinx-subarachnoid shunt was then placed and secured [Figure 2d]. The arachnoid membrane was then closed over the shunt tubing [Figure 2e]. The patient tolerated the surgery well without complication.

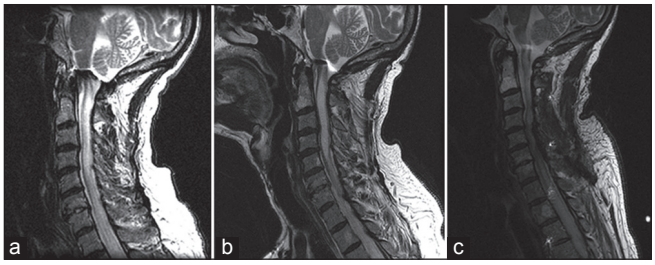

Figure 1: Sagittal T2-magnetic resonance imaging showing syringomyelia at presentation to UI neurosurgery. (a) One year after presentation. (b) Five months postoperatively. (c) The syrinx is decreased in size after syrinx-subarachnoid shunt

Postoperative course

By postoperative day 4, the patient had improved strength (grade 4+/5) in his right upper and lower extremities. Immediately after surgery, he had worsened proprioception in his right lower extremity, however this improved by postoperative day 6. Subjectively, the patient's sensation improved on the right side. He was able to ambulate with minimal assistance prior to discharge.

Five months following his operation, the patient returned to the hospital with new symptoms of right hemiparesis and hoarseness. On exam, he had decreased right facial sensation and decreased gag along with his right side hemiparesis. Brain and spinal MR were obtained and demonstrated an infarct in the right dorsolateral medulla. He was started on aspirin and plavix.

Magnetic resonance imaging of the neuraxis 2 years later demonstrated deflation of syringomyelia from 11 mm width to 3 mm [Figure 1c]. The patient has not followed-up since.

Case 2

History and physical

A 48-year-old woman with a 5-month history of progressive gait disturbance to the point of requiring a walker and eventually being wheelchair bound. Her past-medical history was significant for grade IV SAH from a ruptured right superior cerebellar artery aneurysm 8 years prior [Figure 3a and b] treated at our institution. She had been treated via ventriculostomy and coil embolization of her aneurysm and did well. On examination, she had 5/5 strength in the upper extremities and 4/5 strength in her lower extremities. Sensation

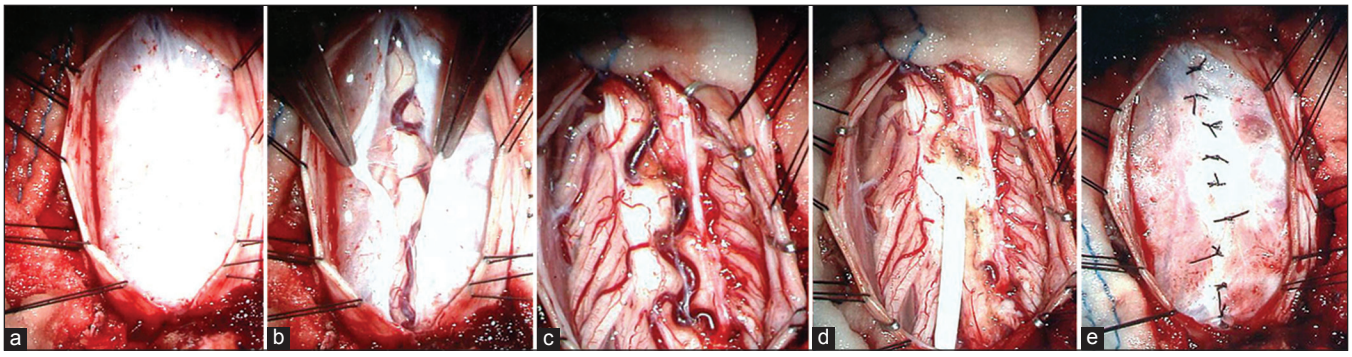

Figure 2: (a) Intraoperative microscope photographs of Case number 1 syrinx-subarachnoid shunt placement. The arachnoid was densely opaque. (b) Opened to initiate lysis of adhesions. (c) The subarachnoid space was satisfactorily opened. (d) To allow for placement of syrinx-subarachnoid shunt. (e) Subsequent arachnoid closure

was decreased in the bilateral lower extremities. There was no change in bowel or bladder function. Neuraxis MR imaging was obtained and demonstrated holocord syringohydromyelia extending from C4 to the conus with arachnoid enhancement on contrast administration [Figure 4].

Operation

Given progressive symptoms of weakness and sensory changes along with imaging findings of large syrinx, the patient underwent syrinx-peritoneal shunt placement. Osteoplastic laminectomies were performed at T7 and T8 to expose the dura. The arachnoid was thickened, white, and opaque with adhesions to the spinal cord and dorsal nerve roots [Figure 5a]. Given the degree of arachnoiditis, the decision was made to perform a syrinx-peritoneal shunt rather than a syrinx-subarachnoid shunt. A midline myelotomy was performed at T7 to expose the syrinx and a catheter was set in place and secured to the pia-arachnoid [Figure 5b]. The dura was closed around the proximal catheter in a standard fashion [Figure 5c]. This was then tunneled to a right flank incision for insertion into the peritoneal cavity. The patient tolerated the surgery well without complications.

Postoperative course

By postoperative day 7, the patient had improved lower extremity strength to a grade of 4+/5. Six weeks after surgery, her symptoms remained stable. She continued to require a wheelchair for ambulation; however her weakness did not worsen further. MR imaging obtained 2 years after surgery demonstrated significant decreased in distension caused by syringohydromyelia.

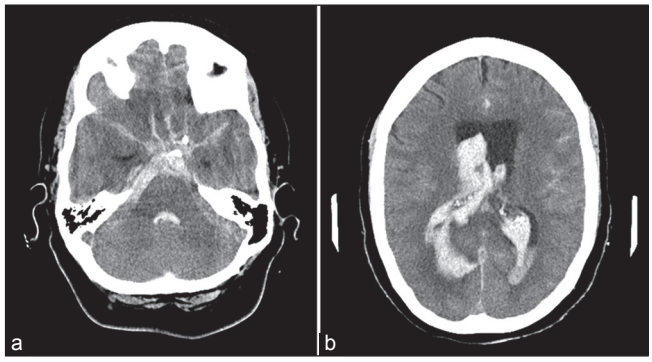

Figure 3: Axial noncontrast computerized tomographic images demonstrating aneurysmal subarachnoid hemorrhage in Case number 2. There is massive hemorrhage in the basal cisterns (a), hemorrhage occluding the IVth ventricle (a), and substantial intraventricular hemorrhage (b)

DISCUSSION

In this report, we document the presentation and management of two patients with delayed development of syringomyelia after aneurysmal SAH. This is an extremely rare complication of SAH with few cases reported in the literature.[8] The pathophysiology of syrinx formation after SAH is poorly understood, but is likely to result from arachnoiditis causing impaired cerebrospinal fluid dynamics (CSF) dynamics around the spinal cord.[9]

Spinal arachnoiditis is a chronic inflammation of the pia-arachnoid that occurs after meningitis, spinal trauma, SAH, and intrathecal injection.[10,11] The range of severity is broad for spinal arachnoiditis with mild cases characterized by faint arachnoid thickening and severe cases noted to have effacement of the subarachnoid space with the formation of arachnoid cysts.[3] Spinal arachnoiditis is associated with meningeal thickening, adhesions with cord deformity, meningeal contrast-enhancement, arachnoid cyst formation, and syrinx.[12] The presentation of spinal arachnoiditis is varied with many patients being completely asymptomatic, but patients with more severe pathology developing severe myelopathy or radiculopathy.[10] The mechanisms of syringomyelia in patients with spinal arachnoiditis are incompletely understood and recently reviewed by Klekamp.[13] Spinal arachnoiditis is thought to cause syringomyelia by two mechanisms: Obstruction of normal CSF flow and spinal cord tethering.[13] Arachnoiditis adhesions slow flow though the subarachnoid space, which increases subarachnoid pressure rostrally and intramedullary pressure caudally.[9] This pressure gradient leads to intramedullary edema and ultimately syringomyelia.[13]

The pathophysiologic mechanisms that mediate spinal arachnoiditis after SAH are incompletely understood. Many of the patients reported to develop spinal arachnoiditis after SAH had posterior fossa aneurysms (including the cases presented here), which may be due to blockage of the basal cisterns by blood.[13] This supports the idea of hematomyelia being the instigating stimulus for arachnoidopathy after SAH. After rupture of a cerebral aneurysm, a high concentration of blood

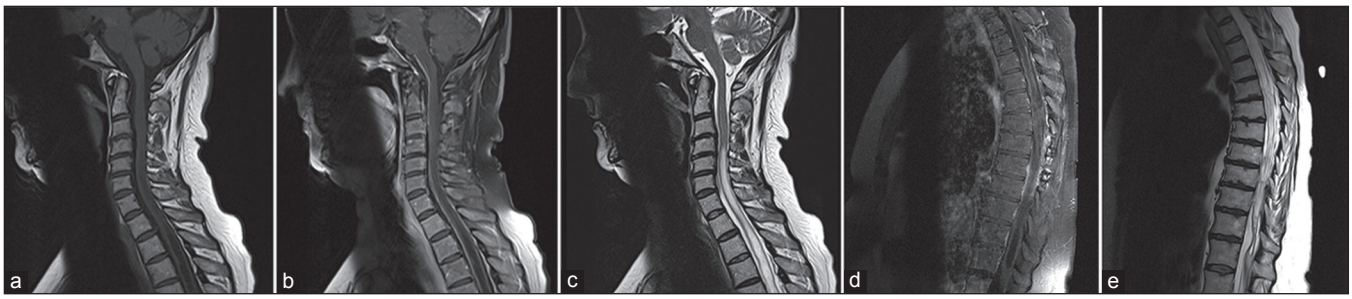

Figure 4: Cervicothoracic syrinx demonstrated by sagittal T1 precontrast magnetic resonance (MR) (a) sagittal T1 postcontrast MR (b and d) Sagittal T2 MR. (e) Postcontrast images demonstrate significant arachnoid enhancement secondary to arachnoiditis (b and d)

products accumulates in the subarachnoid space leading to meningeal irritation and ultimately arachnoiditis. The presence of these blood products in the posthemorrhagic period initiates an inflammatory response and in turn induces a fibroproliferative state and this has potential to cause arachnoiditis anywhere in the neuraxis.[14,15] The initial phase of this response involves a rapid influx of inflammatory cells leading to local release cytokines.[16] The released cytokines signal a fibroproliferative response by attracting fibroblasts which ultimately leads to increased collagen synthesis.[16] This pathway may ultimately lead to obliteration of the subarachnoid space (with or without arachnoid cysts), adhesions, and spinal cord tethering that are characteristic of spinal arachnoiditis.

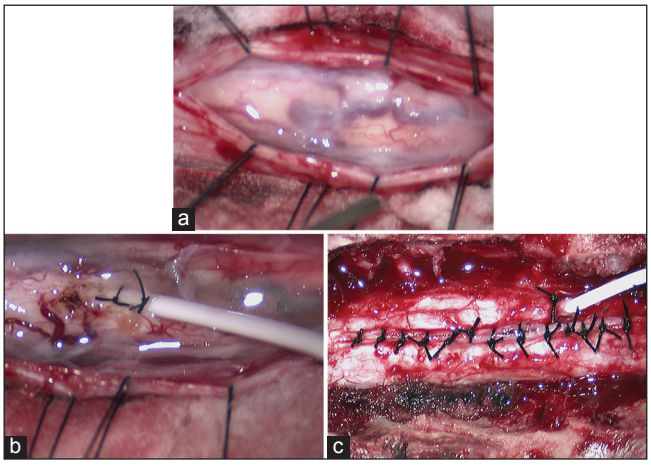

Figure 5: (a) Intraoperative microscope photographs from Case number 2.The arachnoid was opaque. (b) A syringoperitoneal shunt was placed. (c) The dural closed around the proximal catheter

Spinal arachnoiditis resulting from aneurysmal SAH is rare with few cases reported in the literature.[2,3,5-8,13,17,18] In 1943, Nelson was the first to describe spinal arachnoiditis after SAH.[4] His patient was a 50-year-old man with thrombocytopenic purpora who became myelopathic years after suffering an intracranial hemorrhage. On autopsy, the patient was found to have diffused intracranial and spinal arachnoiditis with syringomyelia. In 1961, Lombardi *et al.* mentioned an additional three patients who developed spinal arachnoiditis after SAH, however there is no comment on the etiology of SAH in his patients.[19] In 2000, Kok *et al.* reviewed the literature on spinal arachnoiditis after SAH and found only 14 cases.[3] Of the cases reviewed, nearly all of them presented with symptoms of myelopathy and were found to have findings of arachnoiditis on neuroimaging when it was available. When a cerebral aneurysm rupture is the etiology of SAH precipitating spinal arachnoiditis, it is more commonly the result of aneurysm in the posterior fossa. In the review by Kok *et al.*, in five of six cases in which the aneurysm location was known, the aneurysm originated from an artery in the posterior fossa.[3] More recently, Klekamp documented three of four patients with syringomyelia after SAH with posterior fossa aneurysms.[13]

Syringomyelia occurs in a small subset of patients who develop spinal arachnoiditis after aneurysmal SAH and therefore it is very rare.[8,9,13,18] The characteristics of patients described in detail in the literature are summarized in Table 1. In 2012, Nakanishi *et al.*

Table 1: Summary of syringomyelia cases caused by arachnoiditis associated with aneurysmal SAH

Case	Age/sex	Aneurysm location	Presentation	Latency	Radiographic findings	Treatment	Outcome
Eneling *et al.*, 2012: Case number 1	57/female	Acomm	Gait disturbance, urinary retention, LE weakness	18 months	Arachnoiditis with thoracic syrinx	Laminectomy and SP shunt	Improvement in symptoms
Eneling *et al.*, 2012: Case number 2	65/female	L PICA	LE weakness, urinary retention	8 years	Arachnoiditis with cervicothoracic syrinx	Laminectomy and SP shunt	Radiographic improvement with no improvement in symptoms
Nakanishi *et al.*, 2012: Case number 1	54/female	L MCA	LE weakness, hyper-reflexia	20 months	Arachnoiditis with C7 – T10 syrinx	Laminectomy and syrinx-subarachnoid shunt	Radiographic improvement with incomplete improvement in symptoms
Nakanishi *et al.*, 2012: Case number 2	49/male	L VA	Diplopia, hoarseness, dysphagia, ataxia	5 months	Arachnoiditis with medulla — C6 syrinx	FM decompression with IVth ventricle — subarachnoid shunt	Radiographic improvement. Dysphagia and hoarseness improvement. Incomplete recovery of diplopia and ataxia
Abel *et al.*, 2013: Case number 1	66/male	R VA	Neck pain	11 years	Arachnoiditis with C1 — mid thoracic syrinx	Laminectomy, lysis of adhesions, syrinx-subarachnoid shunt	Symptomatic improvement and radiographic decrease in syrinx
Abel *et al.* 2013: Case number 2	48/female	R SCA	LE weakness progressing to wheelchair dependence	8 years	Arachnoiditis with C4 — conus syrinx	Laminectomy with syrinx-peritoneal shunt	Symptomatic improvement and radiographic decrease in syrinx

Acomm: Anterior communicating artery; PICA: Posterior inferior cerebellar artery; MCA: Middle cerebral artery; VA: Vertebral artery; SCA: Superior cerebellar artery; LE: Lower extremity; SP: Syringoperiotoneal; SAH: Subarachnoid hemorrhage

described two patients who presented with myelopathy and were found to have syringomyelia on spinal MR imaging.[18] The first patient developed myelopathy 20 months after rupture of a left MCA aneurysm and was found to have a C2-T10 syrinx, which was subsequently treated with a syrinx– subarachnoid shunt. The second patient presented 5 months after SAH and underwent IVth ventriculo-subarachnoid shunt for a medulla — C6 syrinx. Both patients demonstrated radiographic reduction in syrinx, but incomplete symptom relief. Also in 2012, Eneling *et al.* described two patients who presented with myelopathy after aneurysmal SAH who were found to have syringomyelia.[8] Both were treated with syrinx-periotoneal shunts and improved radiographically and symptomatically.

Arachnoiditis-associated syringomyelia may be treated with laminectomy with lysis of adhesions and duraplasty, syringosubarachnoid shunt, or syringoperitoneal shunt.[20] The most appropriate management is situation dependent and heavily debated. In Case number 1, lysis of adhesions was satisfactorily opened into the subarachnoid space to enable placement of syrinx-subarachnoid shunt. When distant adhesions are evident on preoperating MR imaging, local lysis of adhesions is unlikely to open the subarachnoid space well enough for effective syrinx to subarachnoid drainage. In these cases, syringoperitoneal shunt are necessary and can have satisfactory results as we have demonstrated in Case number 2.

CONCLUSIONS

Subarachnoid hemorrhage-associated arachnoiditis is an extremely rare cause of syringomyelia that can produce symptoms from months to years after aneurysmal rupture. The pathogenesis of arachnoiditis in this setting is not fully understood, but is associated with posterior fossa aneurysms suggesting that blood product hemolysis in the spinal subarachnoid space is an important factor. Successful treatment depends on restoration of CSF flow via lysis of adhesions with or without syringosubarachnoid shunt or syringoperitoneal shunting.

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How to cite this article: Abel TJ, Howard MA, Menezes A. Syringomyelia and spinal arachnoiditis resulting from aneurysmal subarachnoid hemorrhage: Report of two cases and review of the literature. J Craniovert Jun Spine 2014;5:47-51.
Source of Support: Nil, **Conflict of Interest:** None declared.