ISSN 1220-8841 (Print)
ISSN 2344-4959 (Online)

ROMANIAN
NEUROSURGERY

Vol. XXXIX | No. 1 March 2025

Syringomyelia resulting from aneurysmal subarachnoid haemorrhage. One case report and literature review

Alexandru Ioan Bira,
Horia Ples

DOI: 10.33962/roneuro-2025-001

Romanian Neurosurgery (2025) XXXIX (1): pp. 9-13
DOI: 10.33962/roneuro-2025-001
www.journals.lapub.co.uk/index.php/roneurosurgery

Syringomyelia resulting from aneurysmal subarachnoid haemorrhage. One case report and literature review

Alexandru Ioan Bira[1], Horia Ples[1,2]

[1] Emergency County Hospital, Timisoara, ROMANIA
[2] "Victor Babes" University of Medicine and Pharmacy, Timisoara, ROMANIA

ABSTRACT

Syringomyelia, characterized by cystic cavitation in the spinal cord, can be primary or secondary to various conditions.[1] This article focuses on syringomyelia resulting from subarachnoid haemorrhage (SAH). A case study of a 45-year-old female with SAH is presented. Despite successful aneurysm clipping, the patient developed post-SAH syringomyelia, a rare complication occurring in less than 1% of SAH cases. [2] The pathophysiology involves arachnoiditis disrupting cerebrospinal fluid dynamics, leading to spinal cord tethering. Multiple hypotheses, including inflammatory responses and disruptions in glymphatic flow, contribute to syrinx formation.[4] Surgical options, from arachnoid lysis to various shunting procedures, aim to address progressive symptoms. The choice remains case-specific, with debates on long-term shunt efficacy. Overall, syringomyelia post-SAH poses diagnostic and therapeutic challenges, emphasizing the need for further research in understanding and managing this rare complication.

Keywords
syringomyelia,
cerebral aneurysm,
subarachnoid haemorrhage

Corresponding author:
Horia Ples

"Victor Babes" University of Medicine and Pharmacy, Timisoara, Romania

horia.ples@gmail.com

INTRODUCTION

Syringomyelia or syrinx represent the cystic cavitation of the spinal cord.

Syrinx can be either primary, idiopathic, without an identifiable cause, or secondary to a myriad of conditions, including Chiari type I malformation, spinal cord trauma, inflammatory/ postinfectious conditions such as arachnoiditis, and both spinal and posterior fossa neoplasms, among others. [1]

Syringomyelia largely affects children and young adults. The prevalence has been estimated at 9 per 100,000 people, and an incidence of 0.44 cases per year has been cited in the literature, but epidemiological data on syringomyelia is limited. Chiari malformations are responsible for nearly half of the affected population, while spinal cord trauma and arachnoiditis account for another quarter of adult patients with syringes. [1]

Milhorat developed a classification of syringomyelia based on pathophysiology by comparing information obtained from 175

ISSN online 2344-4959
© Romanian Society of Neurosurgery

First published
March 2025 by
London Academic Publishing
www.lapub.co.uk

autopsies with clinical data on 927 patients with syringomyelia. Syringes are divided into four categories on the basis of pathologic findings: (1) dilations of the central canal that are anatomically continuous with the fourth ventricle (communicating syringomyelia); (2) noncommunicating syringomyelia, including dilations of the central canal that do not communicate with the fourth ventricle and extracanalicular syringes that originate in the spinal cord parenchyma and do not communicate with the central canal or fourth ventricle (primary parenchymal cavitations); (3) atrophic syringes occurring with myelomalacia (syringomyelia ex vacuo); and (4) neoplastic cyst (Table 1). [6]

In this article, we will focus our attention on syringomyelia resulting from subarachnoid hemorrhage.

Table 1.

Class Type of Syringomyelia
I. Communicating syringomyelia • Central canal dilations 1. Communicating hydrocephalus (posthemorrhagic, postmeningitic) 2. Complex hindbrain malformations (Chiari type II, encephalocele) 3. Dandy-Walker malformation
II. Noncommunicating syringomyelia • Central canal/paracentral syringes 1. Chiari malformation 2. Basilar invagination 3. Spinal arachnoiditis (posttraumatic, postmeningitic) 4. Extramedullary compression (spondylosis, tumors, cysts) 5. Tethered cord 6. Acquired tonsillar herniation (hydrocephalus, intracranial mass lesions) • Primary parenchymal cavitations 1. Spinal cord trauma 2. Ischemia/infarction 3. Intramedullary hemorrhage
III. Atrophic cavitations (syringomyelia ex vacuo)
IV. Neoplastic cavitations
Modified from Milhorat TH, Johnson RW, Milhorat RH, et al. Clinicopathological correlations in syringomyelia using axial magnetic resonance imaging. *Neurosurgery.* 1995;37:206-213.

Case presentation

We present the case of a 45-year-old female patient, first admitted to our clinic in 2018 with subarachnoid hemorrhage Fischer Grade II, Hunt and Hess Grade II, and WFNS Grade I. Cerebral CT angiography revealed cerebral aneurysms of the anterior communicating artery and the left posterior inferior cerebellar artery.

Analyzing the subarachnoid hemorrhage pattern, the rupture of the anterior communicating artery aneurysm was identified, leading to surgical intervention for aneurysm clipping. The neurological outcome was favorable.

However, 7 days after the surgical procedure, the patient's neurological status deteriorated abruptly. A cerebral CT scan revealed infratentorial subarachnoid hemorrhage with intraventricular extension (Fischer Grade IV) and secondary hydrocephalus. Emergency neurosurgical intervention included the placement of an external ventricular drain and clipping of the left posterior inferior cerebellar artery aneurysm. Subsequent progress was slow, but favorable.

In the course of recovery, the patient required the placement of a permanent ventriculo-peritoneal shunt. She was discharged conscious, Glasgow Coma Scale (GCS): 15p, ambulating with assistance, and a modified Rankin Scale (mRS) score of 2, one month after admission.

In 2022, the patient presented to our clinic complaining of neck pain, paresthesia in the upper and lower limbs, progressive spastic tetraparesis and loss of thermal and pain sensitivity especially in upper limbs. The symptoms had started six months prior to the presentation. The MRI revealed obstruction of the outflow pathways of the fourth ventricle and holocord syringomyelia. (Fig. 1)

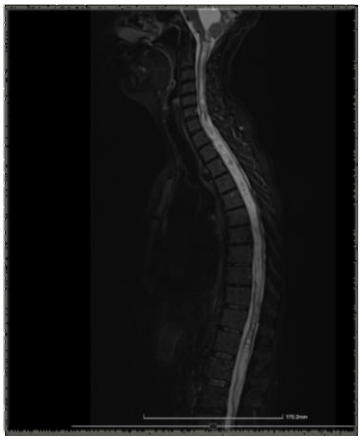

Figure 1.

Considering the imaging findings and the symptoms, a decision was made to perform posterior fossa decompression through adhesiolysis and

the placement of a cervical syringo-subarachnoid shunt via midline myelotomy and duraplasty.

Immediately postoperatively, there was an worsening of motor deficits (from muscular force 4/5 to 3/5), and she develop neurogenic bladder. After a few days, the symptoms subsided. The patient is discharged 10 days postoperative. At discharge, she is conscious, Glasgow Coma Scale (GCS) score of 15, mobilizing with assistance, with pre-operative spastic tetraparesis with overall muscle strength of 4/5, preservation of sensory disturbances, and improvement in pain symptoms.

At the 6-month follow-up, a reduction in the size of the syringomyelic cavity at the cervical level was observed, but the syringomyelia in the thoracic-lumbar region persisted. Currently, the patient is not inclined towards undergoing another intervention.

Discussion

Post-subarachnoid hemorrhage syringomyelia occurs in less than 1% of subarachnoid hemorrhage cases.[2] Given the rarity of this complication and the absence of specific guidelines and recommendations, we sought information in the literature. Searching on PubMed with the keywords "syringomyelia" and "subarachnoid hemorrhage," we identified 10 articles addressing this subject. Searching in the literature, we identified several risk factors in the development of this late complication, including ruptured aneurysms in the posterior circulation, intraventricular extension of hemorrhage, and the need for cerebrospinal fluid drainage [2]. These risk factors align with the case of our patient.

Given the physiopathology of this entity, it can be stated that the involved mechanism is not fully understood. Several hypotheses have been proposed.

It is considered that the presence of blood in the subarachnoid space acts as a trigger. Hemoglobin degradation products generate activation and recruitment of inflammatory cells. Ultimately, this inflammatory mechanism leads to fibroblast activation with collagen synthesis and the development of chronic arachnoiditis. The formation of adhesions in the subarachnoid space leads to tethering of the spinal cord and disturbances in cerebrospinal fluid dynamics. Adhesions in the cerebellomedullary cistern obstruct the outlet of the fourth ventricle thereby inhibiting the cerebrospinal fluid flowing out of the fourth ventricle and leading to hydrocephalus, which directly impacts and dilates the spinal cord central canal, thus ultimately leading to syringomyelia. In the case of our patient, this mechanism seems to be involved, considering the obstruction of the obex by adhesions.[2,6] (Fig. 2)

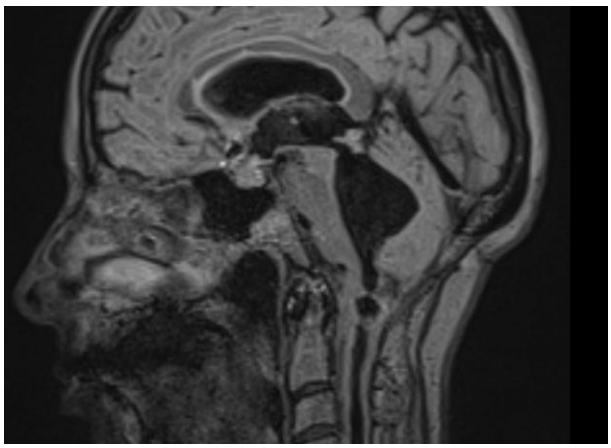

Figure 2.

This mechanism bears resemblance to the hydrodynamic theory proposed in the 1960s by Gardner. The theory suggested that cerebrospinal fluid (CSF) pulsations originating from the choroid plexus typically contribute to the expansion of the neural tube during development. According to the theory, imbalanced CSF pulsations between the supratentorial and infratentorial spaces during development could lead to the formation of a small posterior fossa, tonsillar ectopia, and the redirection of CSF from the fourth ventricle into the central canal due to the obstruction of fourth ventricular outflow tracts at the foramen magnum.[1]

Another mechanism that could be involved is represented by disruptions in the glymphatic flow. The glymphatic system is the internal mechanism through which the central nervous system accomplishes metabolic clearance. It has been demonstrated that within the spinal cord, there exists the perivascular network necessary for the functioning of this system. [7,8]

We know from studies conducted on the brain that the presence of blood in the subarachnoid space leads to a blockage of the glymphatic system pathways [8]. We have no reason to believe that, in the context of presence of subarachnoid hemorhage in spinal subarachnoid space, blood cannot disrupt the functioning of the spinal glymphatic flow.

In a 2017 study examining glymphatic system in normal pressure hydrocephalus, the intrathecal administration of an MRI contrast agent (gadobutrol), serving as a cerebrospinal fluid tracer, revealed indications of delayed glymphatic clearance in iNPH patients compared to a reference group[9]. Consequently, it is possible that the glymphatic flow may contribute to hydrocephalus in patients with subarachnoid hemorrhage, both in the acute phase and later through its permanent impairment. Although we are aware of the speculation we are raising, we consider that the disruption of glymphatic flow is involved in the development of syringomyelia following subarachnoid hemorrhage. Of course, the mechanism is not well-defined, and further studies are absolutely necessary.

Regarding the surgical treatment, the recomandation it is to treat the patient with progressive symptoms.

Surgical options for treatment of symptomatic syringomyelia include direct lysis of arachnoid adhesions with or without duraplasty and a variety of shunting procedures, including syringo-subarachnoid shunt, syringo-pleural, and syringo-peritoneal. [1,2,3]

Certain authors argue that the primary treatment objective should be the release of adhesions through duraplasty. Advocates of this approach assert that arachnoid lysis addresses the fundamental disruption in cerebrospinal fluid (CSF) flow and enlarges the subarachnoid space, as opposed to merely redirecting CSF. Klekamp and colleagues observed that among 67 patients treated for progressive symptoms, 97% of those who underwent syrinx shunting experienced recurrence during follow-up, whereas 78% of those who underwent arachnoid release and duraplasty remained stable.[10]

Shunting of the syrinx to the subarachnoid, pleural, or peritoneal space is still widely accepted as the treatment of choice for patients with syringomyelia caused by arachnoiditis. Shunting of the syrinx to the subarachnoid or peritoneal cavity was associated with a recurrence rate of 60%, whereas microsurgical dissection of the arachnoid scar and decompression of the subarachnoid space had a recurrence rate of 33%, with a mean follow-up period of 28 months. Successful long-term management of the syrinx was associated with microsurgical dissection of the arachnoid scar and decompression of the subarachnoid space. [2,3]

These data appear to be supported by the conclusions of other articles. In a study published in 2011 the conclusion is that shunting proves effective in alleviating the pressure of the syrinx on the spinal cord tissue, but its long-term impact is limited, with recurrence rates varying from virtually all patients experiencing a recurrence to 60% . This outcome is not unexpected, considering that shunting does not address the root cause of syrinx formation; rather, it merely alleviates the symptoms.[4]

Regarding the effectiveness of shunts no statistically significant difference in efficacy between these methods was highlighted.

In a meta-analysis published in 2021, assessing the complication rates of syringo-subarachnoid, syringo-peritoneal, and syringo-pleural shunts, regardless of the cause that led to the development of syringomyelia, syringopleural shunts may offer the lowest rate of reoperation.[6]

In conclusion, our opinion is that in the case of syringomyelia resulting from post-subarachnoid hemorrhage arachnoiditis, the primary goal of the intervention is the lysis of adhesions and the opening of the subarachnoid space. If distal to the site of adhesiolysis there are no other areas of chronic arachnoiditis, we recommend the placement of a syringo-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, so we recommand syringo-pleural or syringo-peritoneal shunt. [2]

Conclusions

Syringomyelia as a complication following subarachnoid hemorrhage is a rare occurrence, with limited reported cases in the literature, accounting for less than 1% of SAH patients. The exact pathophysiology of syringomyelia post-SAH is not fully understood but may involve arachnoiditis, which disrupts cerebrospinal fluid dynamics and leads to spinal cord tethering. Surgical management is a viable option for patients with progressive symptoms, and until this moment we have more questions than answers about this entity.

References

1. Youmans and Winn Neurological Surgery, 4-Volume Set, 7th Edition.
2. Syringomyelia and spinal arachnoiditis resulting from

aneurysmal subarachnoid hemorrhage: Report of two cases and review of the literature Taylor J. Abel, Matthew A. Howard, III, Arnold Menezes.

3. Spinal syringomyelia following subarachnoid hemorrhage Kinya Nakanishi ⇑ , Takuya Uchiyama, Naoki Nakano, Norihito Fukawa, Kimito Yamada, Tomonari Yabuuchi, Amami Kato.
4. Subarachnoid Hemorrhage-associated Arachnoiditis and Syringomyelia J. Eneling · S. Boström · S. Rossitti.
5. Syrinx shunts for syringomyelia: a systematic review and meta-analysis of syrin-gosubarachnoid, syringoperi-toneal, and syringopleural shunting. Robert J Rothrock, Victor M Lu, Allan D Levi.
6. Classification of syringomyelia , T H Milhorat.
7. The ultrastructure of spinal cord perivascular spaces: Implications for the circula-tion of cerebrospinal fluid Magdalena A Lam 1, Sarah J Hemley 2, Elmira Najafi 2, Nicole G F Vella 3, Lynne E Bilston 4, Marcus A Stoodley.
8. The Glymphatic System: A Beginner's Guide. Nadia Aalling Jessen 1, Anne Sofie Finmann Munk 2, Iben Lundgaard 2, Maiken Nedergaard.
9. Glymphatic MRI in idiopathic normal pressure hydrocephalus. Geir Ringstad,1,2 Svein Are Sirirud Vatnehol,3 and Per Kristian Eide.
10. Treatment of syringomyelia associated with arachnoid scarring caused by arach-noiditis or trauma, J Klekamp, U Batzdorf, M Samii, H W Bothe.