

Case Report

Gall stone pancreatitis in a syringomyelia patient: case report and historical review

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ABSTRACT

Syringomyelia is a rare neurological disorder generally regarded as chronic, characterized by slowly progressing fluid-filled cyst (syrinx) forms within the spinal cord. Here we describe the historical review of a patient suffering from syringomyelia for the past 16 years and present gastric condition- gall stone induced pancreatitis. A quadriparetic female patient who had previously undergone surgical treatment for syringomyelia with the early signs including muscle weakness and wasting (atrophy), loss of reflexes, loss of sensitivity to pain and temperature and also stiffness in back, shoulders, spinal curvature (scoliosis) presenting complains of multiple episodes of hematemesis and severe constipation since 5 days. Ultrasonography imaging and laboratory investigations revealed gallbladder stone induced acute pancreatitis also associated. The patient underwent multiple surgical interventions for syrinx removal and associated surgical complications were managed adequately. Currently, the pancreatitis was managed by laparoscopic corrections. Postoperatively, there was a prompt resolution of pancreatitis and associated gastric symptoms. In conclusion, early diagnosis of syringomyelia with magnetic resonance imaging is critical in the prophylaxis of spinal cord compression and potential neurologic injury. Surgical interventions are recommended in symptomatic cases. This case report describes clinical manifestations and management of acute pancreatitis in a chronic sufferer of syringomyelia.

Keywords: Syringomyelia, Syringo-subarachnoid shunting, Gall stone pancreatitis

INTRODUCTION

Syringomyelia is a rare disorder in which a cyst forms within the spinal cord. As this fluid-filled cyst, expands and lengthens over time, it compresses and damages part of the spinal cord from its centre outward and it causes pain, weakness, and stiffness, among other symptoms. The symptoms of syringomyelia usually develop slowly over time; it mainly occurs with a condition which brain tissues protrude into the spinal canal (Chiari malformation). It is caused by the protrusion of brain tissues into the spinal cord; symptoms may generally

begin between ages 25 and 40. Early signs of syringomyelia include muscle weakness and wasting (atrophy), loss of reflexes, loss of sensitivity to pain and temperature and also stiffness in back, shoulders, arms and legs, pain on neck, arms and back, bowel and bladder function problems, muscle weakness and spasms in legs, spinal curvature (scoliosis). The complications include nerve damage within the spinal cord, scoliosis (an abnormal curve of the spine), chronic pain, and motor difficulties.¹ If a physician suspects syringomyelia, patients may be recommended to do a magnetic resonance imaging (MRI) or computed tomography (CT)

scan. Syringomyelia is treated by surgery; the surgery can reduce the pressure on the brain and spinal cord, restore the normal flow of cerebrospinal fluid, and may improve or resolve syringomyelia. However there is no permanent cure, but continued research is carried out in this area.

Gall stone pancreatitis is an acute condition which develops from gallstone blockage in the pancreatic duct. The round muscle called sphincter of Oddi at the opening of the bile duct into the small intestine is the affected part. The signs and symptoms of acute pancreatitis include upper abdominal pain, fever, rapid pulse, tenderness when touching the abdomen it can cause serious complications including pseudocyst. Acute pancreatitis can cause fluid and debris to collect in cyst-like pockets in the pancreas and large pseudocyst that ruptures can cause complications such as internal bleeding and infection. In most countries, the presence of gallbladder stones represents the most frequent and significant risk factors for developing acute pancreatitis and underlying gallstone disease accounts for between 30% and 50% of cases with pancreatitis.² Other causes include heavy alcohol use, cystic fibrosis, and other inherited disorders, a high level of calcium or fat in the blood, some medicines and autoimmune conditions. Laparoscopic Cholecystectomy is considered as the first-line treatment option in this acute condition.

CASE REPORT

A 62 year-old Indian lady who has undergone surgical intervention for syringomyelia to the emergency department with complaints of epigastric pain, multiple episodes of haematemesis, constipation for 5 days. The patient had no history of loose stools or malena.

Disease background - syringomyelia

The patient first presented with complaints of the weakness of the hand and lower limbs (LL) from 2003, also had difficulty in sitting from supine posture, complaints stiffness and pain in both LL. By 2009 she was walking with support. She had decreased sensation for both pain and touch in hands, especially on medical aspects. Sustaining painless burns in both hands and clawing of both hands was present. She couldn't feel slippers in both feet for 5-8 years from the onset of symptoms. Pain and touch sensation was reduced below the ankle in both LL and she experienced pain in LL knee joints.

The magnetic resonance image morphology is suggestive of the expanded spinal cord with associated syringohydromelia from the fifth cervical vertebra (C5) to sixth dorsal vertebrae (D6) level with intervening septations. The proximal segment of the lesion [from fifth cervical vertebrae to seventh cervical vertebra (C5- C7)] showed irregularly thickened and ill-defined walls with different signal intensity (hyperintense) of the cyst fluid in proton density-weighted imaging. On postcontrast images, mild

peripheral enhancement of this lesion is seen suggestive of the tumoral component. Compared to the previous magnetic resonance image the tumour components didn't show significant changes. The inferior extent of the syrinx has extended to sixth dorsal vertebra (D6) level.

Table 1: Canal measurements.

C2	14 mm
C3	12 mm
C4	12 mm
C5	10 mm
C6	9 mm
C7	10 mm

She underwent third dorsal vertebra (D3) laminectomy and syringo-subarachnoid shunt procedure in January 2011. Postoperative imaging showed the reduced size of the syrinx in lower cervical upper dorsal with some contrast enhancement possibly postoperative inflammatory changes at the surgical site. Shunt to tip was well inside. Postoperatively she was very well for 45 days, following which she started appreciating tract flexor spasms. She was restarted on pregabalin, baclofen. As the anti-spasticity drugs stepped up to relieve spasticity she experienced worsening of weakness which forced lowering of anti-spasticity drugs. With less adjustment of drugs, her symptomatology slightly improved and was able to attend daily activities with support. At the stage, patient needed a lot of emotional, psychological support and explanation of realistic chances of improvement. Since counselling alone did not improve her mood satisfactorily and she continued to have wasting limbs, antidepressants were added to her treatment regimen. She was advised to undergo physiotherapy, gait training, regular exercise at home.



Figure 1: MRI after subarachnoid shunt procedure in 2011.

Since June 2012 she again developed progressive LL weakness. She also developed pain in both LL for the same duration. Recurrence of symptoms was probably due to a malfunction of the syringo-subarachnoid shunt in view of high morbidity of re-do surgery and arachnoidolysis. The patient was planned for conservative management. However, theco-peritoneal (TP) shunt was done in January 2013 to improve cerebrospinal fluid (CSF) drainage which might improve syringo-subarachnoid shut function. The patient developed an acute subdural hematoma (SDH) following TP shunt and TP shunt was blocked and later on removed. Further management planned was to repeat plain CT brain after 3 months to look for resolution of SDH and to repeat spine MRI after 6 months to look for an increase in the size of the syrinx. Re-do surgery would be planned in case of symptomatic deterioration or an increase in the size of the syrinx, otherwise, she will be continued on conservative management.

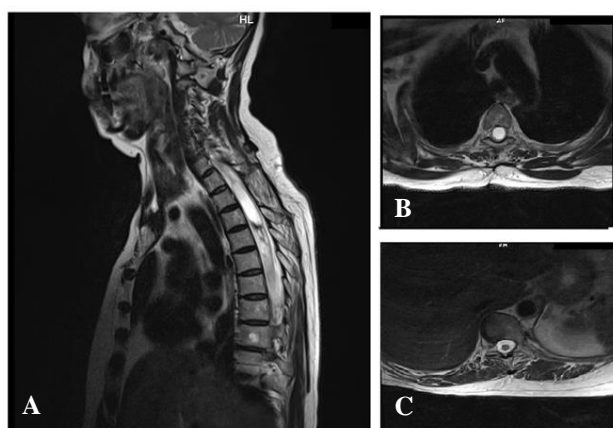


Figure 2: (A) Sagittal MRI showing multiple pockets of syrinx; (B and C) axial image depicting syrinx.

By late 2014 she again developed shunt infection and shunt blockage. Magnetic resonance imaging (MRI) findings showed that lower cervical and upper dorsal spinal cord appears bulky and it is seen replaced by multiple pockets of fluid signal intensity abnormality from the fifth cervical vertebra (C5) to fifth dorsal vertebrae (D5) level. Dorsal scoliosis to right was also identified. Then she underwent second thoracic vertebra (T2) laminectomy, syringo-subarachnoid shunt and lumbo-peritoneal (LP) shunt in January 2015.

Currently, she is on alprazolam 0.5 mg at night, baclofen 10 mg two times daily, pregabalin 75 mg at night. She is having severe constipation for which she is taking combination group of sodium picosulfate (3.33 mg) + liquid paraffin (1.25 ml) + milk of magnesia (3.75 ml) syrup 10-25 ml regularly.

The course of the disease: acute pancreatitis

The patient presented to the emergency department with complaints of multiple episodes of hematemesis,

constipation for 5 days. The patient had no history of loose stools or malena. Her physical examination revealed pallor, epigastric tenderness. No palpable splenomegaly, lymphadenopathy or jaundice was noted.

Laboratory investigations showed a low haemoglobin level of 9.1 g/dl (12-16 g/dl). Her renal, liver function tests, serum electrolytes were normal. Her serum amylase levels were much elevated 753.7 U/I (10-100 U/I).

She was managed with metoclopramide 10 mg intravenous one time daily, rabeprazole 20 mg intravenous two times daily, lorazepam 1 mg intramuscular at night, cinitapride 1 mg orally three times daily, sodium picosulfate 20 mg orally at night and cefoperazone+sulbactam 2 gm injection intravenous two times daily initially.

Her ultrasonography results of the abdomen revealed bulky pancreas, cholelithiasis, traces of pleural effusion and ascites and acute pancreatitis. The patient was suggested for laparoscopic cholecystectomy and the procedure was done on the seventh day of her admission.

Patient was better and symptomatically improved and was discharged after 10 days of hospital stay with discharge medications cefuroxime 500 mg two times daily, diclofenac 25 mg two times daily for 5 days, domperidone (30 mg) + rabeprazole (20 mg) at night for 10 days, ursodeoxycholic acid 150 mg two times daily for 15 days along with old medicines.

DISCUSSION

The diagnosis of acute pancreatitis (AP) requires the presence of 2 of the following 3 criteria: (a) characteristic abdominal pain, (b) serum amylase and/or lipase ≥ 3 times the upper limit of normal, and (c) CT scan findings compatible with AP.³⁻⁵ In AP, usually, serum lipase increases within 4–8 hours after onset of symptoms, peaks at 24 hours, and returns to normal after 8–14 days.^{4,5} The specificity and sensitivity of amylase and lipase are reported to be substantially dependent on the detection method used, ranging from 74 to 100% and from 34 to 100% for serum lipase, and from 70 to 100% and from 33 to 89% for serum amylase, respectively.⁶

It is becoming increasingly clear that syringomyelia cannot be regarded as a condition with a uniform pathology. Syringomyelia is a condition in which fluid-filled cavities develop in the spinal cord. Syringomyelia can be broadly classified into four groups: communicating, non-communicating, atrophic cavitation and neoplastic cavitation. Obstruction of CSF pathways, located distal to the fourth ventricular outlets, creating a syrinx that communicates with the fourth ventricle causes communicating syringomyelia. In non-communicating syringomyelia, the obstruction is usually located at or below the level of the foramen magnum, syrinx and dilated central canal do not communicate with the fourth

ventricle.⁷ The following is the most common type of syringomyelia, found in approximately 50% of all cases, where CSF flow from the posterior fossa to the caudal space is blocked, and the most common underlying cause is an Arnold-Chiari type I malformation.^{7,8} Other common causes of syringomyelia include neoplastic disease, hindbrain anomalies, inflammatory conditions and trauma.^{5,6,9,10}

Myelography was the method for radiographic diagnosis of syringomyelia until the widespread availability of magnetic resonance imaging rendered the radiographic diagnosis of syringomyelia completely non-invasive. Magnetic resonance imaging also provided a convenient tool to monitor changes in diameter and length of the syrinx after surgery, which is one method of evaluating the effectiveness of surgery.¹¹⁻¹⁸

Treatment depends on clinical features and associated findings. Symptomatic treatment commonly suffices in stable courses of disease, while patients with progressive neurological deterioration require surgical decompression.¹⁹ The main cause of the poor outcome and high recurrence after shunting procedure is shunt obstruction and dysfunction.^{20,21} It is seemingly most often the result of the collapse of the syrinx cavity around a shunt tip. Batzdorf et al identified proximal obstruction as the most common source of shunt malfunction and demonstrated in the growth of glial tissue into the perforations of the tube as the cause of shunt failure.²¹ One of the most significant contributors to shunt malfunction is occlusion from arachnoiditis and adhesions around the catheter tip. This is influenced by the individual characteristics of the arachnoid space and the integrity of the arachnoid membrane, ultimately determined by the underlying pathology.

Various literature demonstrates the use of a myringotomy tube for syringe-subarachnoid shunting for the surgical management of a syrinx with good results. This tube is preferred over other shunts due to three reasons. Firstly fluid can communicate according to the pressure gradient voluntarily between the syrinx, the subarachnoid space, and the peritoneal cavity. Secondly, the shunt catheter migration is preventable due to the T- tube dural anchoring is easy and postoperative. Finally, shunt revision with a T tube is easy because only one arm of the T tube is inserted into the intraspinal syrinx cavity.²²

CONCLUSION

Among patient with syringomyelia, periodic application of MRI plays a decisive role in syrinx progression tracking. It is also of great value in identifying the most convenient site for myelotomy and shunt placement if desired. Assessment of surgical outcome can be performed with MRI postoperatively. However, there is no direct correlation between the intensity of the neurological deficits and the size of the syrinx.

As we have described, various techniques are being employed for the surgical management of syringomyelia with varying success. Shunt dislocation is another major cause which leads to shunt dysfunction, frequently occurring in syringo-subarachnoid shunt using the linear catheter. No technique has produced superior results when compared to the others; therefore, various shunting procedures such as syringo-subarachnoid, syringo-peritoneal and syringo-pleural are practised and can be used in different settings. The treatment of syringomyelia remains a difficult endeavour due to a multitude of factors..

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REFERENCES

1. Mayo Clinic. Syringomyelia. 2017. Available at: <https://www.mayoclinic.org/diseases-conditions/syringomyelia/symptoms-causes/syc-20354771>. Accessed on 06 August 2019
2. Pancreapedia. Gallstone-related pathogenesis of acute pancreatitis. 2016. Available at: <https://www.pancreapedia.org/reviews/gallstone-related-pathogenesis-of-acute-pancreatitis>. Accessed on 06 August 2019.
3. Banks PA, Freeman ML. Practice guidelines in acute pancreatitis. *Am J Gastroenterol.* 2006;101:2379–400.
4. Cartier T, Songi P, Perruche F, Meyniard O, Claessens YE, Dhainaut JF, et al. Normal lipase serum level in acute pancreatitis: a case report. *Emerg Med J.* 2006;23:701–2.
5. Shah AM, Eddi R, Kothari ST, Maksoud C, DiGiacomo WS, Baddour W. Acute pancreatitis with normal serum lipase: a case series. *JOP.* 2010;11:369–72.
6. Khan FY, Matar I. Chylous ascites secondary to hyperlipidemic pancreatitis with normal serum amylase and lipase. *World J Gastroenterol.* 2007;13:840–42.
7. Milhorat TH. Classification of syringomyelia, *Neurosurg Focus.* 2000;8(3):1–6.
8. Roy AK, Slimack NP, Ganju A. Idiopathic syringomyelia: retrospective case series,

- comprehensive review, and update on management. *Neurosurg Focus*. 2011;31(6):15.
9. Larner AJ, Muqit MMK, Glickman S. Concurrent syrinx and inflammatory central nervous system disease detected by magnetic resonance imaging: an illustrative case and review of the literature. *Medicine*. 2002;81(1):41-50.
10. Samanci Y, Celik SE. Syringomyelia associated with Cervical Spondylosis: a case report. *Romanian Neurosurg*. 2014;21(2):220-3.
11. Attal N, Parker F, Tadié M, Aghakani N, Bouhassira D. Effects of surgery on the sensory deficits of syringomyelia and predictors of outcome: a long term prospective study. *J Neurol Neurosurg Psychiatr*. 2004;75:1025-30.
12. Barbaro NM, Wilson CB, Gutin PH, Edwards MS. Surgical treatment of syringomyelia. Favorable results with syringoperitoneal shunting. *J Neurosurg*. 1984;61:531-8.
13. Heiss JD, Patronas N, DeVroom HL, Shawker T, Ennis R, Kammerer W, et al. Elucidating the pathophysiology of syringomyelia. *J Neurosurg*. 1999;91:553-62.
14. Isu T, Iwasaki Y, Akino M, Abe H. Hydrosyringomyelia associated with a Chiari I malformation in children and adolescents. *Neurosurg*. 1990;26:591-6.
15. Sansur CA, Heiss JD, DeVroom HL, Eskioglu E, Ennis R, Oldfield EH. Pathophysiology of headache associated with cough in patients with Chiari I. *J Neurosurg*. 2003;98:453-8.
16. Shannon N, Symon L, Logue V, Cull D, Kang J, Kendall B. Clinical features, investigation and treatment of post-traumatic syringomyelia. *J Neurol Neurosurg Psychiatry*. 1981;44:35-42.
17. Steinbok P. Ethical considerations relating to writing a medical scientific paper for publication. *Childs Nerv Syst*. 1995;11:323-8.
18. Ventureyra EC, Tekkok IH. Syringostomy using myringostomy tube: technical note. *Neurosurg*. 1997;41:495-7.
19. Amboss. Syringomyelia. 2019. Available at: <https://www.amboss.com/us/knowledge/Syringomyelia>. Accessed on 07 August 2019.
20. Aghakhani N, Baussart B, David P, Lacroix C, Benoudiba F, Tadie M, et al. Surgical treatment of posttraumatic syringomyelia. *Neurosurg*. 2010;66(6):1120-7.
21. Batzdorf U, Klekamp J, Johnson JP. A critical appraisal of syrinx cavity shunting procedures. *J Neurosurg*. 1998;89:382-8.
22. Kim S, Choi S, Youm J, Kwon H. Syringosubarachnoid-Peritoneal Shunt Using T-Tube for treatment of post-Traumatic Syringomyelia. *J Korean Neurosurg Soc*. 2012;52(1):58-61.

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