Case Report

Post-Traumatic Syringomyelia: A Case Series

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ABSTRACT

It is relatively common to occur in a Tertiary Care Neurosurgical setup to get a case of Syringomyelia proximal or distal to a space-occupying lesion (SOL) or site of spinal cord compression. In this case series, we are presenting two cases in which syringomyelia developed after traumatic spinal cord injury. On initial radiological investigations, the first case presented as an old D12 fracture with Post-traumatic syrinx formation but on complete workup for the extent of the syrinx, another lesion was found incidentally in the form of an intradural extramedullary SOL at the level of cervicomедullary junction. The SOL turned out histologically as WHO Grade I Meningioma. The second case presented as syrinx formation after gunshot (fire-arm) penetrating spinal cord injury to the D11-12 vertebrae. Treatment plans of both these patients are presented here in detail along with the literature review.

Keywords: Syringomyelia, meningioma, cervicomедullary junction, post-traumatic, penetrating spinal cord injury, SOL.

INTRODUCTION

Syringomyelia is defined as the formation of abnormal fluid-filled cavity or syrinx within spinal cord parenchyma containing cerebrospinal fluid (CSF), secondary to spinal cord compression.\textsuperscript{1} The etiology of syringomyelia is multifactorial.\textsuperscript{2,3,4} Although road traffic accidents and fall from height cause most Spinal cord injuries (SCI); 13-17\% of injuries are caused by penetrating spine injury (PSI) (primarily gunshot wounds [GSWs]) which makes them 3\textsuperscript{rd} leading cause of SCI.\textsuperscript{20} Traumatic spinal cord injury(SCI) canal cause a late, relatively uncommon, but potentially harmful complication known as Post-traumatic syringomyelia (PTS). PTS is characterized clinically by the often subtle progression of pain and loss of sensorimotor function that may present after many years of a traumatic spinal cord injury. Loss
of function, chronic pain, respiratory failure, or even death may occur if PTS is left untreated.\textsuperscript{5} Syrinx may extend cranially or caudally to the lesion or site of spinal cord compression. It may extend up to the foramen magnum, involving the brainstem, causing syringobulbia.\textsuperscript{6} Magnetic Resonance Imaging (MRI) is preferably the imaging of choice to see the spinal cord compression and syrinx formation with its extent and probable causation.\textsuperscript{7} The total extent of syrinx should be evaluated by MRI. We report two cases of Post-traumatic syringomyelia: one caused by D12 vertebral fracture with an incidental finding of meningioma at cervicomedullary junction while evaluating for the upper extent of the syrinx and the second caused by gunshot injury to D11-12 vertebrae causing complete spinal cord injury and syrinx formation from D12 up to C2 vertebral level. The history, examination, investigations, and treatment of both cases are presented in detail.

\begin{case-report}
\textbf{Case 1}

A 42 years old Male patient presented in the Outpatient Department of Neurosurgery at Sharif Medical City Hospital on 16-01-2021, with complaints of progressive weakness and numbness of bilateral upper and lower limbs, breathing difficulty, and urinary and fecal incontinence for 3 months. The patient had a preceding history of falling on his back from a trolley, 3 years back, developed severe low back pain (8 on the numeric rating scale) and sudden numbness and weakness of both legs with no urinary or fecal incontinence but was recovered in few days and began walking normally. The patient then remained asymptomatic for 2 years and 9 months. On Neurological Examination, the patient had weakness of all four limbs with Power 1/5 in all limbs involving all key myotomes C4, C5, C6, C7, C8, T1, L2, L3, L4, L5, and S1. Showing upper motor neuron signs with probable phrenic nerve involvement causing respiratory difficulty. Decreased pain and touch sensations at C3 and below dermatomes. Decrease anal tone and perianal sensations. Plantar reflex bilateral up going. The weakness in the upper limb was significantly progressed in the last 3 months along with the involvement of respiration.

MRI Dorso-lumbar spine was initially done which showed D12 collapse vertebral fracture with kyphosis causing significant spinal cord compression with syrinx formation extending upwards up to upper dorsal spinal cord (Figure 1).

To determine the upper extent of the syrinx, MRI cervical spine with contrast was done which

\begin{figure}[h]
\centering
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\caption{Preoperative MRI Dorso-lumbar spine T2WI image sagittal view showing D12 collapse vertebral fracture with syrinx formation extending upwards up to dorsal spinal cord.}
\end{figure}
showed heterogeneously enhancing probably intradural and extramedullary soft tissue mass measuring 16 × 19 × 37mm (AP, Transverse and CC dimensions) located posteriorly in upper cervical canal extending from craniocervical junction till upper-end plate of C3 vertebrae, being iso-hypo intense on T1WI and intermediate intensity on T2WI, likely displacing spinal cord anteriorly, suggesting meningioma, significantly compressing spinal cord at this level. Heterogeneous contrast enhancement of lesion was also seen. Syrinx formation was appreciable distal to this lesion upto the upper dorsal spinal cord (Figure 2).

CT scan Brain plain was also done showing unremarkable brain parenchyma.

Based on MRI cervical spine, differential diagnosis was made of Spinal tumors e.g., meningioma, neurofibroma, lymphomas, metastatic lesion, and spinal infective lesions. As the MRI Cervical spine with contrast showed an incidental finding of a lesion at the craniocervical junction so the decision was made to treat the proximal lesion first surgically. The surgery was performed for excision of cervical SOL using the posterior approach on 01-02-2021, after taking high-risk informed consent from the patient and relatives.

Under Aseptic measures and proper positioning of the patient, exploration of occipitocervical junction from C1 to C3 level. As shown in Figure 3.

Operative findings included firm to hard, greyish white, mildly vascular, calcified intradural and extramedullary lesion in cervical spine extending from C1 to C3 vertebral level, pushing spinal cord anteriorly, attached to dura mater (Figure-4).

Figure 2: Preoperative MRI Cervical Spine with IV Contrast showing heterogeneously contrast enhancing intradural and extramedullary soft tissue mass extending from craniocervical junction to C3 vertebrae compressing spinal cord, with Syrinx formation.

Figure-3: Perioperative post-exploration view showing occipital bone, C1 vertebral posterior arches and C2 vertebral laminae with spinous process.

Figure-4: Perioperative view after opening of dura showing gross appearance of tumor.
Complete Surgical excision with securing of hemostasis was done, SIMPSON Grade 2 excision was carried out (Figures-5, 6).

Figure-5: Perioperative view showing dissection of tumor and preservation of brain stem and spinal cord.

Figure-6: Perioperative view showing complete excision of tumor from C1 to C3 vertebral level.

Occipitocervical fixation was done to minimize the cervical spine instability (Figure 7).

Figure-7: Perioperative view showing Occipito-cervical Fixation after tumor excision.

Histopathology confirmed that the cervical tumor was Psammomatous Meningioma (WHO Grade I).

The postoperative course was uneventful without complications.

The patient has advised chest and body physiotherapy with breathing exercises.

The patient was followed up after 1 week of discharge in OPD, Stitches were removed and examination was done that revealed improvement in breathing and neurological symptoms: improved power in all myotomes (from 1/5 to 3/5). The patient has advised a follow-up MRI Cervical spine with contrast. But patient developed a COVID-19 infection and was treated at home by a local G.P and got expired in 3rd week.

Case 2
A 50 years old Male patient presented initially in the Emergency Department of Farooq Hospital on 25-06-2019 after suffering a gunshot wound to the back on the lower Thoracic region with no exit wound, with complaints of sudden weakness of bilateral lower limbs and loss of all kinds of
sensations on the lower part of the abdomen and both legs with urinary and fecal incontinence. Neurological examination revealed the weakness of bilateral lower limbs with Power (Grade:) 0/5 in both lower limbs involving all key myotomes L2, L3, L4, L5, and S1, having upper motor neuron (UMN) type paraplegia. Complete loss of all types of sensations at T12 and below dermatomes on both sides. Increased anal tone and absent perianal sensations. Plantar reflex bilateral up going.

CT scan Dorso–lumbar spine was done which showed Fire-arm bullet injury to D11 and D12 vertebrae which caused complete spinal cord injury at D12 level (Figure-8). MRI Dorso-lumbar spine was not done due to the presence of a bullet.

![Figure-8: Preoperative CT scan Dorso-Lumbar Spine plain](image)

After initial management and wound debridement, got discharged with no improvement in signs and symptoms, with subsequent weekly follow-up in OPD. The patient remained in this condition for almost 2 months. After that, he started developing gradual weakness and numbness of bilateral upper limbs with respiratory difficulty. On Neurological Examination, the patient had weakness of all four limbs with Power (Grade:) 3/5 in upper limbs involving all key myotomes C4, C5, C6, C7, C8, and T1, showing upper motor neuron signs with probable phrenic nerve involvement causing respiratory difficulty and Power (Grade:) 0/5 in both lower limbs involving all key myotomes L2, L3, L4, L5, and S1, having UMN type paraplegia with decrease muscle bulk in both lower limbs. Decreased pain and touch sensations at C3 and below dermatomes and complete loss of all types of sensations below T12 dermatomes on both sides. Increased anal tone and absent perianal sensations. Plantar reflex bilateral up going. To evaluate for the cause of quadriparesis, MRI cervical spine was required, which was not possible in the presence of a bullet. So open surgery was performed with the informed consent of the patient and the bullet was removed from D11-12 level after laminectomy under fluoroscopic guidance on 10-08-2019. MRI whole spine plain was done at 3rd Post-operative day following bullet removal, which showed syrinx formation in spinal cord parenchyma extending from C3 to spinal cord injury at D12 level (Figure 9, 10).

![Figure-9: MRI Cervical Spine plain](image)

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The patient was managed conservatively following bullet removal for 1 month with regular chest and body physiotherapy, but the condition remained static and could not improve. After 1 month, the condition worsened and Syringo-subarachnoid shunting was done for syringomyelia at C5-6 vertebral level using the posterior approach on 17-09-2019, after taking high-risk informed consent from the patient and relatives.

The postoperative course was uneventful without further complications.

The patient was advised chest and body physiotherapy with breathing exercises.

The patient was followed up after 1 week of discharge in OPD, Stitches removed and examination was done that revealed improvement in breathing and neurological symptoms: improved power in all myotomes of both upper limbs (Power Grade from 3/5 to 5/5) with normal sensations in dermatomes upto D12 bilaterally. However, the neurological status of the lower limbs did not improve. The patient was been advised follow-up MRI whole spine with contrast which showed resolution of the syrinx in the cervical and dorsal spine (Figure-11). Patient is stable with intact neurology in the upper limp since last more than 2 years.

**DISCUSSION**

Syringomyelia is a progressive neurological disorder, defined as the formation of the syrinx within spinal cord parenchyma, containing cerebrospinal fluid (CSF), secondary to spinal cord compression. The different types and causes of Syringomyelia are shown inFigure-12. The exact pathophysiology of syringomyelia is unknown according to the literature. However, different theories regarding pathophysiology have been proposed in the literature. According to the proposed theories in the literature, 3 distinctive types of spinal cord cavities may form depending upon the causes. First is hydromyelic/communicating form which is located centrally and there is communication with the fourth ventricle cranially/rostrally, more commonly associated with congenital defects of the fourth ventricle. Second is the syringomyelic/non-communicating form in which the cranial end of the cavity is closed before it conjoins the fourth ventricle. Third is extracanalicular form which is secondary to traumatic spinal cord injury. However, all theories relate to CSF flow dynamics disorders and abnormalities.

Post-traumatic syringomyelia (PTS) is one of the acquired causes of (secondary) syringomyelia,
first narrated by Bastian in 1867.\textsuperscript{5} Approx. 3 – 4% of patients with traumatic spinal cord injury (SCI) develop clinically symptomatic PTS.\textsuperscript{5,13} Mortality from PTS can result from brainstem respiratory center involvement and surgical complications. As males have a higher incidence of SCI, they have a higher incidence of PTS as compared to females. PTS is unrelated to the age of the patient and time after traumatic SCI.\textsuperscript{5} Magnetic resonance imaging (MRI) plain with IV contrast is the modality of choice in diagnosing syrinx formation with its extent and its probable cause.\textsuperscript{7}

In the first case, the patient presented with 3 months history of progressive quadripareis after 3 years of spinal injury, differential diagnosis was made of post-traumatic syringomyelia. In this case, an MRI of the dorsal spine revealed an old healed wedge fracture of D12 resulting in Kyphotic deformity and PTS. Since in MRI the upper-end Syrinx was not visible and symptoms were progressive involving cervical spine and brain stem. It was therefore decided to do an MRI of the cervical spine to localize the upper extent of Syrinx. CT scan was done to see the possible
Hydrocephalus. To our surprise an intradural extramedullary mass was detected incidentally which was enhancing with contrast, extending from Craniocervical junction to C3 vertebra. Syrinx was extending from the lower pole of this mass to the upper border of the fractured vertebra. It was suggesting meningioma of the craniocervical junction at the upper extent of the syrinx with normal brain study on CT scan Brain plain.

Most of craniocervical meningioma are benign. As meningiomas of the craniocervical junction are in close relationship to the brainstem, cranial nerves, and critical vascular structures, complete surgical excision (Simpson Grade I) of the tumor, which should be the primary goal of surgery, is often difficult to achieve, which is why a higher risk of recurrence might be expected.

So, as proposed by Levine et al., Fox et al. and Oldfield et al., the formation of the syrinx in this patient was attributed to spinal trauma at the dorsal spine as well CSF obstruction at the craniocervical junction by the spinal tumor. And the resection of the tumor will cause the elimination of the mass effect of the tumor on the spinal cord and the obstruction to the flow of CSF in its physiological pathway. The surgical excision of the tumor with occipito-cervical fixation was done with planned spontaneous resolution of syrinx which would be followed up in subsequent follow-up visits in planned intervals. The dorsal spine fracture was to be dealt with in second stage surgery after the proposed 3 months interval if syringomyelia had not resolved. However, the patient showed improvement in neurological symptoms after 2 weeks and all of his stitches were removed. Follow-up MRI cervical spine with contrast was advised and 3 months follow up to check the resolution of Syrinx but unfortunately, the patient got expired due to COVID-19 infection.

In the second case, the patient developed syringomyelia as a consequence of a gunshot wound leading to penetrating spine injury which caused the vertebral fracture, complete spinal cord injury, and CSF obstruction. Although the patient had paraplegia, with complete sensory loss in both lower limbs along with urinary and fecal incontinence, he developed bilateral upper limbs weakness and numbness with respiratory difficulty after 2 months, suggesting cervical spine involvement. To evaluate for the cause, the first bullet was removed using laminectomy of D11 and D12 vertebras under fluoroscopy guidance and MRI whole spine plain was done which showed syrinx formation in cervical spine parenchyma extending from C2-3 vertebral level upto the level of dorsal spinal cord injury. As the patient was having a progressive neurological deficit in upper limbs as well as respiratory difficulty in brief 1-month history following bullet removal without any improvement, to resolve the syrinx in the cervical spine and decrease its pressure on the spinal cord, Syringo-subarachnoid shunt placement was planned. After successful surgery, the patient was followed up in the OPD and showed improvement in the neurology of both upper limbs and regained normal Power (Grade 5) 5/5 in all muscle groups of both upper limbs and improvement in respiration as well regained normal sensations upto D12 level. Follow-up MRI cervical with dorsal spine plain with IV contrast revealed resolution of the syrinx. In this case, the spinal cord injury was attributed to direct impact by the bullet together with its fragments as well as indirect injury by its velocity, rotational force, and kinetic energy. The syrinx formation was probably due to CSF obstruction by spinal cord injury and also due to subclinical infection or chemical changes in cord and CSF caused by a bullet and/or its fragments. No lesion was found at the upper end of the syrinx. So syringo-subarachnoid shunting was done to divert the path of CSF above the level of its obstruction, thereby normalizing the flow of CSF from the spine to the brain and reducing the pressure effect of the syrinx in the spinal cord especially in the cervical region.
CONCLUSION

It is mandatory to define the extent of Syrinx. If it is extending up to the cervicomедullary junction, one must rule out the presence of Hydrocephalus in the Brain. Any negligence may lead to wrong planning and inappropriate treatment. In case of lesions at both ends, one must treat the proximal lesion first followed by the serial MRI cervical spine to see whether the Syrinx is resolving or not. In case of resolution and neurological improvement, the patient is followed carefully both clinically and radiologically. Second stage surgery may be planned to resolve the neurological symptoms related to the second lesion. In case Syrinx is not resolved even after the treatment of both ends which is quite a rare possibility then direct treatment of Syrinx itself with Syringo-subarachnoid shunt must be considered.

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Additional Information
Disclosures: Authors report no conflict of interest.
Ethical Review Board Approval: The study was conformed to the ethical review board requirements.
Human Subjects: Consent was obtained by all patients/participants in this study.
Conflicts of Interest:
In compliance with the ICMJE uniform disclosure form, all authors declare the following:
Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.
Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.
Source of Funding: None.

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| Sr.# | Author’s Full Name       | Intellectual Contribution to Paper in Terms of:                                      |
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| 1.   | Babar Butt               | 1. Operating Surgeon of Case 1 and Primary Author.                                    |
| 2.   | Syed Ali Zunair          | 2. Assistant Surgeon, Literature review, Data interpretation and manuscript writing. |
| 3.   | Aman-ur-Rehman           | 3. Assistant Surgeon, Data collection and analysis, referencing.                      |
| 4.   | Alia Latif               | 4. Pre-operative and Post-operative management of patients.                           |
| 5.   | Nasir Raza Awan          | 5. Review of Manuscript                                                              |
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