Early Development of Syringomyelia after Spinal Cord Injury: Case Report and Review of the Literature

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We report a case of post-traumatic syringomyelia (PTS), which developed 2 months after spinal cord injury (SCI). A 20-year-old man who was involved in a motorcycle accident sustained a complete SCI resulting from a burst fracture of the T5 vertebral body. He underwent posterior fixation with decompression at another hospital 2 days after the injury. Postoperative imaging suggested that spinal stenosis endured at the T4 level and swelling of the spinal cord above that level. Two months later, he felt dysesthetic pain in his forearms and hands, but the cause of the pain was not examined in detail. Four months after the injury, he presented with motor weakness in the upper extremities. Magnetic resonance imaging (MRI) showed syringomyelia ascending from the T3 level to the C1 level, and he was referred to our hospital immediately. The imaging studies suggested that PTS was caused by congestion of the cerebrospinal fluid (CSF) at the T3 level. The patient was treated with syringosubarachnoid (SS) shunt at the T1-T2 level, whereby neurological symptoms of the upper extremities were immediately relieved. Postoperative MRI showed shrinkage of the syrinx. At the latest follow-up 2 years postoperatively, there was no sign of recurrence. It is noteworthy that PTS potentially occurs in the early phase after severe SCI. We discuss relevant pathology and surgical treatment through a review of previous literature.

Keywords: early-developing post-traumatic syringomyelia, spinal cord injury, syringosubarachnoid shunt

Introduction

Post-traumatic syringomyelia (PTS) is recognized as a cause of delayed neurological deficits in patients with spinal cord injury (SCI). The interval between SCI and the onset of clinical neurological deterioration has been reported to be more than 10 years.1,5 Although PTS usually develops years after SCI, it potentially occurs within 6 months.3–5 Without knowledge of this epidemiology, early incidence of PTS may be overlooked. This article describes a case of PTS that developed only 2 months after SCI as a result of T5 burst fracture. Although the patient had undergone posterior decompression and fusion surgery at another hospital immediately after SCI, the PTS was extending from the injured spinal levels to the craniovertebral junction. He was successfully treated with syringosubarachnoid (SS) shunt at the T1-T2 level, with no recurrence up to the latest follow-up at 2 years after the operation. In addition to the present case, we comment on early of PTS and discuss its pathology while reviewing the previous literature.

Case Report

A 20-year-old man was involved in a motorcycle accident and sustained a complete SCI (American spinal cord injury association impairment scale A [ASIA A]) caused by a burst fracture of the T5 vertebral body (Figs. 1A and 1B). He underwent posterior decompression at the T5 level and posterior fusion at the T3-T8 levels at another hospital on the second day after SCI. A postoperative computed tomography scan showed that bony decompression of the spinal canal was achieved at the T5 level, but stenosis remained between the T4 lamina and the retropulsed posterior wall of the T5 vertebral body (Fig. 2A). Two weeks later, T2-weighted magnetic resonance imaging (MRI) showed swelling of the spinal cord above that level and diffuse intramedullary hyperintensity from the T4 to the T2 level (Fig. 2B). On retrospective review, T1-weighted images showed a small syrinx at the T2 level that was not noticed at the time (Fig. 2C). The patient’s symptoms were unchanged and stable. He was transferred to the other hospital for rehabilitation 1 month after surgery. At 2 months, he began to feel dysesthetic pain in his forearms and hands. The doctors in charge considered that the pain was caused by overuse during rehabilitation and did not examine the symptoms in detail. At 4 months, the patient presented with bilateral weakness of the arms and hands. MRI showed syringomyelia extending from the T3 to the C1 level (Fig. 2D). The syrinx had a conical-shaped base at the T3 level, suggesting that it originated from that spinal level. He was referred to our hospital and immediately underwent SS shunt at the T1-T2 level. After general anesthesia was introduced, the patient was positioned prone on a four-point frame for spinal surgery. After partial laminectomy was performed on the left laminae at T1-T2 level, durotomy was performed with attention paid to preserving the arachnoid membrane. Arachnoid scarring or spinal tethering was not observed at that spinal level (Fig. 3A). The arachnoid membrane was incised along the durotomy, and a small myelotomy was made below the T2 dorsal root exit zone. A shunt tube with a diameter of 1.2 mm and a length of 50 mm (Create Medic, Yokohama, Kanagawa, Japan) was inserted from the myelotomy into the rostral side of the syrinx (Fig. 3B). The tube was fixed with a suture to pia...
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mater, and the caudal side of the tube was positioned at the anterior side of the spinal cord. The patient was relieved of the dysesthetic pain immediately after the operation, and motor weakness of the upper extremities also improved. Postoperative MRI revealed shrinkage of the syrinx (Figs. 4A and 4B). At the latest follow-up 2 years postoperatively, no recurrence was observed (Fig. 4C).

Discussion

PTS usually develops over 10 years after SCI. The reason for a long latent period of PTS after the injury might be derived from its mechanism. There are distinct mechanisms for initial traumatic cyst formation and subsequent syrinx development. Initial traumatic cyst formation is likely to result from direct injury including hemorrhage, excitotoxic injury, and ischemia. After initial cyst formation, hydrodynamic factors are likely to play an important role in PTS development. Subsequent respiratory pressure differentials on the external cord, including fluid movement within the cavity, dissect the cord and propagate the syrinx. In addition to these mechanisms, subarachnoid adhesions and subsequent alteration of cerebrospinal fluid (CSF) flow increases its flow into the syrinx following which neurological deterioration occurs in some patients with enlarged syringomyelia. Although a previous study reported that PTS potentially occurs in the earlier period, only five patients with PTS developing within 3 months of injury have been specifically reported (summarized in Table 1). Several authors described early syrinx formation found on autopsy, but presented no evidence confirming that clinical neurological deterioration had been caused by the syrinx. These results suggest that the pathology of early-developing PTS is probably different from that of the more common PTS.

Other than PTS, subacute post-traumatic ascending myelopathy (SPAM) is known as a cause of neurological deterioration after SCI. SPAM occurs within 3 weeks after injury, but is distinct from secondary damage to the spinal cord caused by the inflammatory cascade occurring within 48 hours of SCI. SPAM is defined as neurological deterioration ascending more than four vertebral levels above the initial injured site, and is diagnosed by MRI findings revealing

Fig. 1 Computed tomography (A) and MRI (B) on the day of the traffic accident showing the burst fracture of the T5 vertebral body (arrow in A) and compression of the SCI with intramedullary signal change (arrowhead in B). MRI: magnetic resonance imaging, SCI: spinal cord injury.

Fig. 2 Imaging studies after posterior decompression and fusion at another hospital. Computed tomography scan shows that spinal canal decompression was obtained at the T5 level but with residual stenosis between the T4 lamina and the retropulsed posterior wall of the T5 vertebral body (A). T2-weighted MRI 2 weeks after the injury shows swelling of the spinal cord above the T4 level with intramedullary hyperintensity at the T2-T4 level (B) while T1-weighted image shows a small syrinx at the T2 level (arrow, C). MRI at 4 months shows syrinx extending from the T3 to the C1 level, with a conical-shaped base at the T3 level (D). MRI: magnetic resonance imaging.
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The present case did not meet the criteria of SPAM. In the present case, spinal stenosis was still apparent at the T4 level after posterior decompression and fusion surgery. We consider that the stenosis caused alteration of the CSF flow, which generated the small syrinx with swelling of the spinal cord in the early period. Postoperative MRI at 4 months showed that the syrinx had a conical-shaped base at the T3 level. This finding appears to support our hypothesis that early PTS was caused by congestion of the CSF flow at the spinal stenosis.

Although surgical treatment is recommended for progressive and symptomatic PTS, the approach is still controversial. In the 1970s and 1980s, shunting of the syrinx to the subarachnoid, pleural, or peritoneal space was widely performed to treat patients with syringomyelia caused by trauma or arachnoiditis. However, the effectiveness of the shunt was poor in patients with arachnoid scarring or tethering of the spinal cord. For such cases, some reports recommended decompression surgery such as untethering, expansive duraplasty, or arachnolysis, resulting in better outcomes. Another reported that cordectomy was applicable for patients with PTS who suffered from complete paraplegia or tetraplegia and had undergone several ineffective procedures including decompression and shunt. In previous reports of early-developing PTS, the patients were treated by shunt operations (Table 1). Although good surgical

intramedullary hyperintensity above the original level of injury without generation of a syrinx. The present case did not meet the criteria of SPAM.

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Fig. 3  Intraoperative view. Right side of photographs is rostral and the top is mid. Microscopic view after right hemilaminectomy at the T1-T2 level. There is no arachnoid adhesion or spinal tethering. An arrowhead shows the right T2 root (A). After a small myelotomy below the dorsal root entry zone at the T2 level, one side of the SS shunt tube was implanted into the rostral side of the syrinx by suturing to the pia mater (B), and the other side of the tube was inserted into the caudal side of the cord. SS: syringosubarachnoid.

Fig. 4  MRI at 1 week (A), 6 months (B), and 2 years (C) after surgical treatment with SS shunt, showing that shrinkage of the syrinx was achieved and preserved. MRI: magnetic resonance imaging, SS: syringosubarachnoid.
results were obtained directly after the operation, the long-term postoperative course was not declared in these reports. Therefore, long-term effectiveness of the shunt operation for early-developing PTS is yet to be established. In the present case, SS shunt at the T1-T2 level was chosen because we considered that the syrinx was caused not by arachnoid adhesion but by congestion of CSF flow at the T4 level. Arachnoid adhesion was not observed at the T1-T2 level in the surgical field (Fig. 3A). As we expected, the SS shunt had worked well and led to a good surgical outcome both neuropsychologically and radiographically until the latest follow-up 2 years after surgery.

Furthermore, previous literature has reported several factors that may affect the prediction of earlier PTS; specifically, PTS occurs earlier (1) in aged patients, (2) in SCI at cervical and thoracic spinal level compared with lumbar spine, (3) in complete SCI, (4) in dislocation fractures, and (5) in patients treated with initial surgery without decompression. Therefore, careful attention should be paid to neurological deterioration in such patients.

We assessed the pathology of the present case according to the serial imaging studies and consequently selected the appropriate surgical approach. This pathological profile may not be common in other patients with PTS developing in the early period after SCI. We believe, however, that this report provides useful information for correct interpretation of the pathology and selection of the surgical method.

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### Conflicts of Interest Disclosure

None of the authors have any conflicts of interest to disclose.

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### Table 1  Summary of patients with early-developing PTS (within 3 months)

| Author   | Age (y)/sex | Mode of injury | Level | ASIA | Skeletal injury | Deterioration | Time since accident | Diagnosis           | Tx                  | Result               |
|----------|-------------|----------------|-------|------|-----------------|---------------|---------------------|---------------------|----------------------|----------------------|
| Lyons²¹  | nd          | Fall           | T6    | A    | T5/6 F          | C8-T5 temperature loss | 3 m            | CCT                 | Syrinx (T5-C3 and C4-medulla) | SS shunt             | Gradually improved |
|          | nd          | CA             | T2    | A    | T2/3/4 F-D      | T2 paresthesia     | 3 m            | CCT                 | Syrinx (C4-7)     | SC shunt             | Complete recovery   |
| Yarkony²² | 44/M        | Fall           | C4-6  | B    | C4/5/6 F        | C5/6 weakness      | 75 d           | MRI                 | Complete block (C5-C7) syrinx (C5/6) | SS shunt             | Partially improved  |
|          | 19/M        | Fall           | C7    | C    | C6/7 F-D and locked facet | Paresthesia       | 65 d           | MRI                 | Syrinx (medulla to C7) | SD shunt             | Partially improved  |
|          | 23/M        | Fall           | C5    | A    | C5 burst F      | Weakness           | 57 d           | MRI                 | Syrinx, edema (C5-T1)  | Conservative         | Shunt operation 2 y later |

ASIA: American Spinal Injury Association Impairment Scale, CA: car accident, CCT: contrast computed tomography, d: days, D: dislocation, F: fracture, M: male, m: months, MRI: magnetic resonance imaging, myelo/CT: myelography with computed tomography, nd: not described, SC: syringocisternal, SD: syringosubdural, SP: syringoperitoneal, SS: syringosubarachnoid, Tx: therapy, y: years.
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