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CASE REPORT

Post-traumatic syringomyelia refractory to surgical intervention: a series of cases on recurrent syringomyelia

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OBJECTIVE: The objective of this study was to analyze a series of traumatic spinal cord injury (SCI) patients with a diagnosis of syrinx who had recurrence of symptomatic syrinx following surgical intervention.
DESIGN: This is a patient series.
SETTING: The study was conducted in an acute inpatient rehabilitation facility.
PARTICIPANTS: Participants included patients ($N = 6$) with post-traumatic syringomyelia (PTS) who had recurrent syrinx despite surgical repair.
INTERVENTIONS: Not applicable.
MAIN OUTCOME MEASURES: The main outcome measures were time period between injury and clinical manifestations of syringomyelia, time to recurrence and presenting symptoms.
RESULTS: Among the six patients, there is great variability between time of the initial SCI and the development of syrinx. In terms of time periods between diagnosis of syrinx and recurrence of syrinx, there is also some variability (ranging from 6 to 936 weeks). The median length of time to recurrence was 104 weeks. In all cases, the presenting symptom was ultimately weakness, and in most cases it was associated with rising sensory deficits. Notably, all patients were male, aged 31–55 years, had suffered diffuse traumatic SCI and four of six patients lost the ability to ambulate because of syrinx formation.
CONCLUSIONS: This series suggests that there may be risk factors for developing post-traumatic syringomyelia. We question whether there is a relationship between American Spinal Injury Association Impairment Scale grade and recurrence of PTS. We need to look closely at these patients to see whether there are modifiable risk factors that may minimize their chance of developing PTS. Once these are identified, there may be a role in routine screening of all patients and particularly those who may be at an increased risk for PTS to avoid loss of ambulatory function.

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INTRODUCTION

Syringomyelia is the formation of a cavity within the spinal cord that can increase in size and result in neurologic deterioration. The condition is most often associated with congenital Chiari malformation, or it could arise from a spinal cord insult, such as tumor or traumatic injury. The etiology of posttraumatic syringomyelia (PTS) is not well understood. Although many theories exist, it may be that PTS develops in areas of myelomalacia where there is potential for cyst formation. Scar tissue either within the cord or along the meninges may cause tethering that may interfere with cerebrospinal fluid (CSF) flow. This blockage of the CSF within the subarachnoid space leads to accumulation of fluid and expansion of the cavity.[1,2]

The purpose of this study is to analyze a series of traumatic spinal cord injury (SCI) patients with diagnosis of PTS, who had recurrence of symptomatic syrinx following surgical intervention. We review the risk factors, American Spinal Injury Association (ASIA) level, timing of development to recurrence, type of operation and neurologic outcomes.

CASES

Patient 1

Patient 1 is a 39-year-old man with T1 ASIA B SCI. Twenty-one years after SCI, the patient presented with worsening weakness, spasticity and loss of ambulation. Imaging revealed syringomyelia. He underwent T2–T11 detethering and fusion. He did not regain the ability to walk after this intervention. For the worsening spasticity, he had a baclofen pump placed. Two years after the fusion, he had progressive lower-extremity weakness. A year later, MRI revealed a recurrent syrinx. He underwent reopening of the thoracic laminectomy, lysis of adhesions, with shunt placement, and release of the tethered cord. The baclofen pump was reprogrammed with normal saline. The patient's neurologic level progressed from a T1 ASIA B to a T2 ASIA B.

Patient 2

Patient 2 is a 31-year-old man with T5 ASIA D SCI. After initial rehabilitation stay, he was walking independently using braces and a walker. After 5 years, he developed new numbness and weakness that extended through his left arm and face. Imaging revealed a syrinx, and he underwent untethering and shunt placement. There was a brief improvement initially, but then he had progressive loss of strength in his left upper extremity. One year after initial surgical intervention, he had a syringoperitoneal shunt placed and laminectomy for decompression and detethering. Approximately 4 months later, he was found to have decreased muscle mass and decreased sensation in his left upper extremity. MRI revealed shunt obstruction. Two months later, he

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underwent posterior fixation of T4–T11 and laminectomies T4–T8. Despite these interventions, his neurologic level had progressed from a T5 ASIA D to a C1 ASIA A.

Patient 3

Patient 3 is a 51-year-old man with T12 ASIA A SCI. Ten years after the injury, he had worsening upper-extremity weakness, and imaging revealed the development of a syrinx, and thus he underwent syringopleural shunt placement. Eighteen years later, he began to notice that his arm, neck and chest pain worsened with coughing and sneezing, as well as progressively worsening bilateral upper-extremity weakness. Approximately 3 months later, he had acute onset of severe neck, arm and chest pain. MRI revealed lower cervical and upper thoracic syringomyelia with cystic dilatation of the central spinal canal that extended superiorly to C4–C5. He underwent thoracic laminectomy and placement of a syringopleural shunt. His neurologic level had progressed from a T12 ASIA A to T4 ASIA A.

Patient 4

Patient 4 is a 29-year-old man with C3 ASIA D SCI with C2 type 1-odontoid fracture. After rehabilitation, he had regained ambulatory function. Four years post injury, he had a decline in strength and was diagnosed with cervical syrinx. Imaging revealed a syrinx from the obex of the medulla, extending inferiorly to T5. He underwent placement of a subarachnoid shunt and T1 hemilaminectomy. Two years later, he had neurologic deterioration to the point where he became ventilator-dependent. Several months later, he was found to have recurrent syrinx in the inferior aspect of the medulla through T2. Repeat imaging 1 year later showed further extension of the syrinx to the body of T5. His neurologic level had progressed from a C3 ASIA D to a C2 ASIA A. There was no further surgical intervention.

Patient 5

Patient 5 is a 37-year-old man with T7 ASIA D SCI. He had completed rehabilitation and had regained the ability to ambulate with braces. Two years after injury, the patient presented with changes in sensation and muscle strength, and he had lost the ability to ambulate. This prompted reimaging and diagnosis of syringomyelia. After laminectomy with detethering, followed by inpatient rehabilitation, the patient regained ambulatory function. Six weeks after initial surgery, the patient had acute onset of bilateral lower-extremity weakness. He underwent additional detethering and syringosubarachnoid shunt placement and again participated in acute inpatient rehabilitation. He had symptomatic relief and regained function to the point where he was ambulating with braces and a rolling walker. Twenty-two weeks after the initial procedure, the patient had rapidly worsening lower-extremity weakness and was found to have ascending syringomyelia warranting repeat decompression with detethering and revision of the shunt. His neurologic level had progressed from a T7 ASIA D to a T6 ASIA C. He completed a month of inpatient rehabilitation, but a month later he presented to his neurosurgeon with expanding fullness in his back that was diagnosed as a pseudomeningocele. Initially, he remained at his neurologic baseline but then began to report worsening lower-extremity weakness. He underwent posterior wound irrigation and debridement with the removal of thoracic posterior segmental instrumentation, repair of the CSF leak and placement of a lumbar drain and VP shunt placement. After the procedure and acute inpatient rehabilitation, the patient's neurological classification was T6 ASIA B.

Patient 6

Patient 6 is a 54-year-old man with C7 ASIA A SCI. He was diagnosed with a syrinx 39 years after his initial injury and underwent cervical detethering and fusion. Two years later, he had ascending paralysis with intrinsic hand weakness and was found to have an increasing syrinx on MRI. He underwent posterior cervical fusion laminectomy with detethering of the cord. Postoperative course was complicated by a CSF leak, which prompted washout and placement of a lumbar drain. His neurologic level progressed from a C7 ASIA A to a C6 ASIA A.

RESULTS

There were a total of six patients with a confirmed diagnosis of recurrent PTS. They were all male, and were aged 29–54 years at the time of syrinx diagnosis. At the time of the initial spinal cord injury, four of the six patients had an ASIA Impairment Scale (AIS) grade D and were ambulatory with assistive devices. Two of the patients were tetraplegic. The presenting symptom of recurrent PTS varied; however, all patients presented with weakness as an initial symptom (Table 1).

Development of the initial syrinx ranged from 104 to 2028 weeks. (Table 2) These patients were all referred for neurosurgical evaluation and underwent surgical intervention. Three patients had both shunt placement and detethering, whereas two patients underwent detethering alone and one patient underwent only shunt placement.

Among the six patients, there is great variability between the time of the initial SCI and the development of syrinx. In all cases,

Table 1. Demographics

Case no.	*Gender*	*Age*	*AIS Grade*	*C/I*[a]	*Presenting symptom*
1	M	39	T2 B	I	Weakness/spasticity
2	M	31	T5 D	I	Weakness/sensory loss
3	M	29	C3 D	I	Weakness
4	M	37	T7 D	I	Weakness/sensory loss
5	M	51	T12 A	C	Weakness/pain
6	M	54	C7 A	C	Weakness

Abbreviation: AIS, American Spinal Injury Association (ASIA) Impairment Scale. [a]C, complete injury; I, incomplete injury.

Table 2. Time course for syrinx presentation (weeks)

Case no.	*Initial*	*Recurrent*	*Re-recur*
1	1092	156	–
2	260	52	16
3	208	104	–
4	104	6	30
5	520	936	–
6	2028	104	–
Median	390	104	23

Table 3. Neurologic outcome post surgical intervention

Case no.	*Pre-syrinx*	*Post syrinx*	↑*/↓**
1	T2 B	T1 B	↓
2	T5 D	C1 A	↓
3	C3 D	C2 A	↓
4	T7 D	T6 B	↓
5	T12 A	T4 A	↓
6	C7 A	C6 A	↓

*↑ = Improvement/**↓ = Decline.

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the presenting symptom was weakness. After surgery, time to recurrence ranged from 6 to 936 weeks with a median of 104 weeks. (Table 2) The patients all had a decline in neurologic status. Two of three patients with AIS grade D worsened to AIS A, and the third patient with AIS grade D progressed to AIS B. Four of six patients were AIS incomplete. (Table 3) All patients underwent detethering and four also had shunt placement. The type of operation did not influence neurologic outcome. None of the four patients who had previously been ambulatory were able to walk afterward.

DISCUSSION

This series suggests that the development of PTS occurs as frequently in incomplete as compared with complete SCI. This is in contrast to a recent retrospective cohort study that saw that the majority of cases occurred in AIS grade A injuries.[3] Those with incomplete injuries tended to have a greater degree of functional deterioration. This is evident in the fact that none of the patients who were ambulatory before development of syrinx were able to walk after recurrence, despite surgical intervention. Time to development of PTS in relation to initial injury demonstrates a great degree of variability. Half of the cases occurred within the first 5 years, which is consistent with findings in the literature.[4]

Cord tethering results in obstruction of normal CSF flow through the spinal canal, which may result in the formation of syringomyelia. Detethering with duraplasty is preferable for initial treatment, as it addresses the mechanical cause for fluid collection.[3–5] Shunt procedures should be reserved for recurrent PTS, cases in which complete detethering cannot be achieved or those in which tethering is not the primary mechanism of syrinx formation.[5]

When syringopleural shunts are performed alone, there is an increased risk of complications and recurrence. There was no correlation between the type of surgical intervention and outcome in our recurrent syrinx population, as has been proposed in the first-time occurrence of syringomyelia. One study saw that there was a greater rate of revision for syringomyelia treated with shunting rather than duraplasty.[3] This was not observed in our patient population.

The majority of patients in this series did not recover and lost neurologic function following surgical intervention. This finding is consistent with Klekamp *et al*., who showed that pain relief but not neurologic recovery was the primary benefit of surgery.[6] In addition, it showed that after 5 years 39% had neurologic deterioration after surgical intervention in AIS C and D injuries versus 14% in AIS A and B injuries. Half of our cases of recurrent syrinxes were AIS D, and two out of three of these progressed to complete injury despite the surgical intervention. In our population, incomplete injuries with recurrent syrinxes were also more likely to progress. In contrast to prior studies, none of the patients in our population of recurrent PTS noted any improvement in neurologic function or pain following surgical intervention.[6]

CONCLUSION

We question whether there is a relationship between various factors, including AIS and/or activity level and recurrence of PTS. This series examined the characteristics of patients who developed recurrent PTS but is limited by the relatively small number of cases. The presentation of weakness in all of our patients as opposed to pain or sensory abnormalities is unusual. The incidence of PTS is known to be as high as 29%, although most are asymptomatic. If strength is not monitored closely within the first 5 years, which is when it is most likely to develop, the diagnosis of PTS may be missed. Furthermore, AIS D had greater functional decline than those with AIS A or B, which could indicate that AIS incomplete injuries may be a factor in outcome prediction. We question whether there is a relationship between AIS grade and recurrence of PTS. We need to look closely at these patients to see whether there are modifiable risk factors that may minimize their chance of developing PTS. Once these are identified, there may be a role in routine screening of all patients and particularly those who may be at an increased risk for PTS to avoid loss of ambulatory function.

COMPETING INTERESTS

The authors declare no conflict of interest.

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