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CASE REPORT

Post-traumatic syringomyelia with holocord involvement: a case report

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INTRODUCTION: Syringomyelia is a disorder in which a cyst, or syrinx, develops within the spinal cord. Historically, syringomyelia in post-traumatic spinal cord injury has been uncommon; however, its diagnosis has been increasing due to the advances in medical technology. Syringomyelia that involves the entire spinal cord, or holocord, is rare after traumatic spinal cord injury, with only a few cases reported in the literature.

CASE PRESENTATION: We present a case of a 57-year-old male who had a motorcycle accident 30 years ago resulting in a spinal cord injury, who presented a rapid decline in the function of his left upper extremity. Imaging studies were reviewed to reveal an expansive T2 hyperintense intramedullary spinal cord lesion from C1 inferiorly to the conus medullaris. The patient underwent a T6–T7 laminectomy for the placement of a syringosubarachnoid shunt. The patient was then transferred to acute inpatient rehabilitation where he underwent an intense course of therapy for 3 weeks while being monitored closely by physiatrists.

DISCUSSION: The patient was able to make significant recovery and was successfully discharged home. There are a limited number of reports published about post-traumatic holocord syringomyelia. It is important to recognize this diagnosis during follow-up visits with spinal cord injury patients.

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INTRODUCTION

Syringomyelia is a disorder in which a cyst, or syrinx, forms within the spinal cord. This syrinx can expand over time, leading to the destruction of the existing spinal cord. Although syringomyelia is most commonly a result of an Arnold–Chiari malformation, it is increasingly seen in post-traumatic spinal cord injury (SCI) patients. Depending on the author, the rates were thought to be 1–8%. However, with the help of magnetic resonance imaging (MRI), the rates are thought to be as high as 22%.[1,2]

The time of the initial presentation of syringomyelia varies as well, from several months after SCI to many years later. The syrinx is thought to form due to partial or complete obstruction of the normal cerebrospinal fluid (CSF) flow in the subarachnoid space. The cavity that forms often extends below and above the initial level as time passes.[1]

There are certain risk factors that can increase the predisposition to developing syringomyelia, including arachnoiditis, cord compression and/or a narrow spinal canal and kyphotic deformity. In SCI patients, a syrinx may be found incidentally on routine neuroimaging. However, if it is severe enough, it may manifest as worsening or new neurological symptoms. There have been some published case reports detailing holocord involvement of a syrinx, although this was in the significant minority.[2]

CASE PRESENTATION

A 57-year-old male was presented to the Physical Medicine and Rehabilitation (PM&R) Clinic after being referred by the Neurology Service due to rapid decline in the function of his left upper extremity. The patient's medical history was significant due to a motorcycle accident that happened approximately 30 years before, requiring a prolonged hospital stay. The patient was initially paralyzed below the waist, but was able to regain good strength of his legs and ambulate with bilateral Lofstrand crutches. The patient stated that he continued to have residual ankle weakness, which he reports that he did not receive braces for. The patient reported that he recently moved to the United States, and his prior care occurred in his native country. He reported that he has not had any neurological changes until a few months prior to his arrival in the clinic.

The patient stated that his recent symptoms began with pain starting in the left shoulder region. He was evaluated with a shoulder and cervical spine x-rays, which were negative for acute pathology. The patient reported that the pain progressed down his left hand, and eventually manifested as left upper extremity weakness, which limited his ability to manipulate the Lofstrand crutch. The patient also reported that he began to experience intermittent bowel incontinence. With progression of his symptoms, the patient was initially presented to the emergency department, and then he was referred to the neurology clinic. Neurology ordered an MRI of the cervical spine and an electromyography study to evaluate for cervical radiculopathy. The patient was also referred to the PM&R clinic, which began our initial involvement in the case.

During the PM&R clinic visit, the results of the recently completed MRI were discussed with the patient. The final report documented the findings of an expansive T2 hyperintense intramedullary spinal cord lesion from C1 inferiorly until at least the level of T12 (the limits of the film), consistent with a spinal cord syrinx. Physical examination at the time demonstrated a marked atrophy of the first dorsal interosseous muscle, the thenar and hypothenar eminences, as well as mild weakness on the

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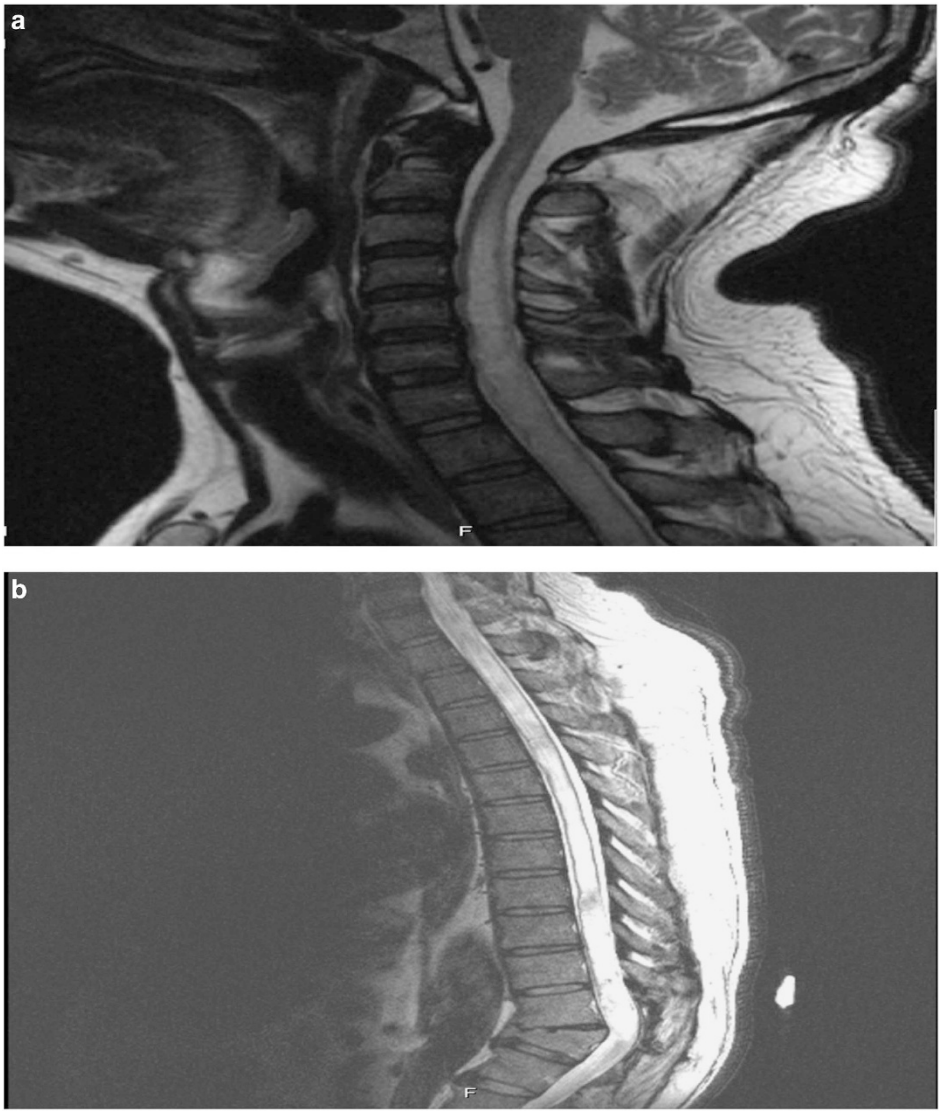

Figure 1. (**a**) T2 sagittal cervical spine. Marked dilatation of the central canal, consistent with a syrinx cavity. T2 hyperintense signal intermixed with a curvilinear hypointense signal. (**b**) T2 sagittal thoracolumbar spine. Marked dilatation of the central canal, consistent with a syrinx cavity. T2 hyperintense signal intermixed with a curvilinear hypointense signal.

patient's left elbow and wrist, and significant weakness on the hand. Additional findings confirmed the patient's bilateral ankle weakness, with only a trace contraction of the dorsiflexors and plantarflexors. These findings were discussed with the neurosurgery consultant, who recommended a clinic appointment in 1 week for the development of a treatment plan. The patient was asked to return to the PM&R clinic after neurosurgical evaluation.

Following the PM&R clinic visit, the patient was reevaluated by neurology, who ordered an MRI of the lumbar spine to evaluate the extent of the syrinx, which was found to be at the level of the conus medullaris (Figures 1a and b). Neurology was also in agreement with referral to neurosurgery. Upon assessment in the clinic, neurosurgery recommended the placement of a syringosubarachnoid shunt. Within 6 weeks from initial presentation at the PM&R clinic, the patient underwent a T6–T7 laminectomy for the placement of the syringosubarachnoid shunt (Figure 2). The patient's postoperative course was fairly unremarkable, and he was found to be a good candidate for acute inpatient rehabilitation. Upon admission to the inpatient rehabilitation unit, the patient's functional status was notable for requiring supervision for feeding and grooming, moderate assistance for lower body dressing, minimal assistance to transfer from sit to stand using a rolling walker and minimal assistance to ambulate with a rolling walker.

The patient underwent a 3-week acute inpatient rehabilitation course with intensive therapy consisting of 2 h of physical therapy a day and 1 h of occupational therapy a day, for 5 days per week. Physical therapy focused on a strengthening program with a focus on bed mobility, transfers and ambulation training using an assistive device. Occupational therapy focused on self-care, activities of daily living assessment and training, adaptive-equipment assessment, safety assessment and training and community reintegration. Psychology and social work services were also provided to the patient to assist him in safe discharge planning. Medical care was provided to the patient with close neurosurgical follow-up. The patient's surgical pain was managed with acetaminophen while his neuropathic pain was managed well with gabapentin. The patient was also provided with a left semisolid ankle foot orthosis (AFO) and a right-articulated AFO for bilateral foot drop. The patient's family was appropriately trained to assist with the patient's ongoing care. This multidisciplinary

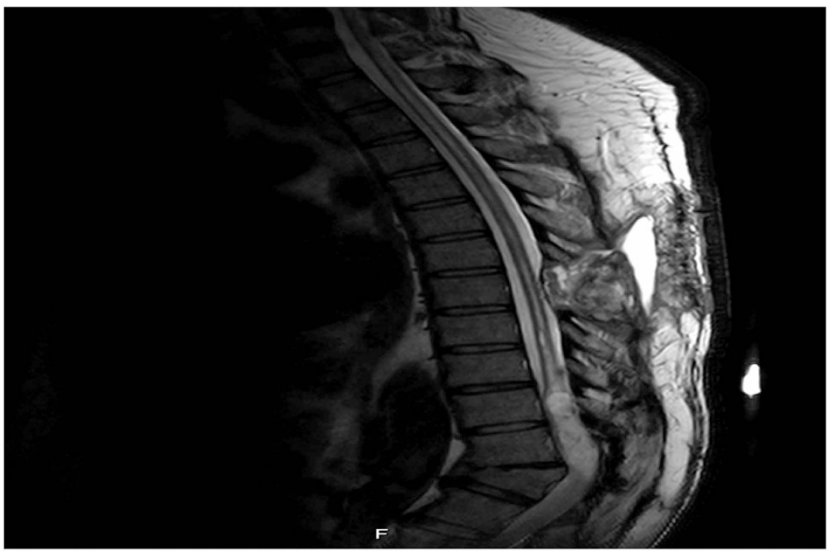

Figure 2. T2 sagittal thoracolumbar spine status—post syringosubarachnoid shunt. Note that the size of the syrinx cavity throughout the cervical and thoracic spine is markedly diminished.

approach during acute inpatient rehabilitation helped the patient to reach and surpass his rehabilitation goals, and he was successfully discharged home with home services and care from his family. On discharge, the patient's functional status improved to set up with upper and lower body dressing, supervision with toilet transfers, modified independence with squat pivot transfers, independence with donning and doffing bilateral AFOs and modified independence with ambulation with bilateral AFOs and a rolling walker.

DISCUSSION

This case was unique in that it involved a patient who developed a syrinx spanning the entire spinal cord, or holocord syringomyelia. The majority of the cases of holocord syringomyelia have been described in Arnold–Chiari malformations, which is the most common cause of syringomyelia overall.[3] Upon literature review, less than ten cases of a holocord syrinx after post-traumatic SCI have been previously described.[4,5] A study published in the Journal of Neurotrauma by Awai *et al.*[5] described several cases of holocord syringomyelia following thoracic and lumbar traumatic SCI. The patients in that study showed symptoms of Syringomyelia 19-34 years after their initial SCI. These patients were found to have holocord syringomyelia through MRI exams, as their only symptom at the time of presentation was mild sensory deficits. Our patient happened to present himself with worsening neurological symptoms that prompted further investigation, leading to the discovery of the holocord syrinx.

The patient in this study was treated with the placement of a syringosubarachnoid shunt. The postoperative course during acute inpatient rehabilitation was unremarkable, but the long-term effects of the procedure are not known as the patient has not returned for follow-up. A study carried out by Lee *et al.*[6] showed that arachnoiditis and/or duraplasty alone had slightly better outcomes for the treatment of post-traumatic syringomyelia, as opposed to shunt placement. However, this study did not specifically describe the treatments for post-traumatic holocord syringomyelia. Ghobrial *et al.*[7] described a case of post-traumatic holocord syringomyelia that was treated with arachnoiditis and shunt placement, which is in line with the treatment that was provided for this patient.[7]

In addition to the surgical treatment, this patient also underwent an intense acute inpatient rehabilitation program. Prior case descriptions of post-traumatic holocord syringomyelia failed to describe the role of rehabilitation in their patient's recovery. The order and placement of AFOs played a significant role in the patient's overall recovery and the ability to be discharged home within a reasonable amount of time.

CONCLUSION

SCI can cause a myriad of consequential conditions that affect a patient's functional mobility and activities of daily living. Syringomyelia can be a potentially serious sequela in a post-traumatic SCI patient, and the syrinx may not develop until years after initial injury. It is important to consider syringomyelia in this patient population, as the dysfunctional CSF mechanics may continue to expand the syrinx beyond the original level of injury. In rare occurrences, the syrinx can encompass the entire spinal cord, leading to worsening of the neurological dysfunction. The patient described in this case had holocord involvement, and showed that patients can make substantial recovery with shunt placement and PM&R involvement. Further studies need to be carried out looking at the long-term recovery in this specific patient population.

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COMPETING INTERESTS

The authors declare no conflict of interest.

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