Post-Traumatic Syringomyelia at Conus: Report of Two Cases

Abolfazl Rahimizadeh, Mohammad Saghri, Ava Rahimizadeh

Department of Neurosurgery, Pars Hospital, Tehran, Islamic Republic of Iran

ABSTRACT

Progressive post-traumatic cystic syringomyelia (PTS) is an increasingly recognized cause of morbidity following spinal cord injury. Post-traumatic development of syrinx at the conus is an uncommon event and our knowledge about this entity is confined to single case reports or small series of two to three cases. This pathology is usually associated with late kyphosis resulting from conservative treatment or inadequate decompression and stabilization of thoracolumbar spine fractures.

Herein, two cases of post-traumatic syringomyelia (PTS), both developing after conservative management of L1 vertebral body fracture and with late kyphosis are presented. In both instances, development of pain and new neurological symptoms pointed to the diagnosis, after 2 years in one and after 12 years in the other. Decompression of the canal and intradural arachnoidolysis followed by myelotomy in one and syringsosubarachnoid shunt in the other result in good outcome.

KEY WORDS: Late kyphosis, post-traumatic syringomyelia, spinal trauma, syrinx

INTRODUCTION

Syringomyelia is a relatively infrequent, but clinically serious and potentially disastrous complication of spinal cord injury (2,3,5,6,7,9,14,16-19,23,25,26,29,32-34,36,38,39,41,46-48,51,52,54-56,61,64). The incidence of PTS once was believed to occur in approximately 0.3 to 3.2% of all spinal injuries (38,55). However with increased availability of MRI, this pathology is now increasingly recognized (43). The chance of development of this pathology is increased whenever spinal cord decompression, restoration of alignment and stabilization is ignored (4,23,43,57,58).

PTS is mostly seen in the cervical and the dorsal spinal cord and usually extend several levels above and below to the site of injury (2,3,5,6,7,9,14,16-19,23,25,26,29,32-34,36,38,39,41,46-48,51,52,54-56,61,64). However development of PTS at the conus region is quite infrequent (1,8,26,40,49,52). The interval between the initial spinal injury and development of a syrinx at the conus is variable. It might develop from a few months to several years after trauma (1,8,26,40,49,52). Its development is usually subsequent to type A L1 or T12 fractures and usually as a consequence of ignorance or maltreatment (1,8,26,40,49,52).

Delayed onset of neuropathic pain and development of new neurological symptoms after an apparently stable period points to the diagnosis. Post-traumatic kyphosis and local canal stenosis frequently coexist with the syrinx at the time of diagnosis (14,8,23,26,40,43,49,52,57,58). This emphasizes the major role that these two factors play in the development of PTS. Taking these factors into consideration, the formulation of a single strategy in dealing with all these pathologies together becomes possible. This means that for achieving good outcome, one should reduce the spinal cord compromise, restore the kyphosis, release the arachnoid scarring, and perform detethering (4,23,32,33,43,57,58).

Subsequently, myelotomy with or without shunting procedure might be done on the preference of the surgeon despite the existence of controversies (19,25,26,38). Herein, two cases with untreated L1 burst fractures in whom development of new neurological symptoms was confirmed
to be due to PTS at the conus are presented. Decompression of the conus, correction of kyphosis and release of arachnoid scarring in both cases could reestablish faint pulsation of the cord. Therefore, myelotomy in one and myelotomy in combination of syringosubarachnoid shunt in the other resulted in collapse of the syrinx, normal pulsation of the cord with good postoperative outcome.

CASE 1

A 48-year-old woman was admitted because of gradual and progressive weakness of the feet and dysesthesia of the distal lower extremity and the buttocks in the last six months. She had a history of car accident two years before admission resulting in L1 fracture. This was conservatively treated with a cast. On admission, neurological examination revealed weakness of flexors of the feet. The patient could not stand on tiptoe; ankle jerk was diminished bilaterally. Sensation of the saddle area was also disturbed with difficulty in defeation where urination was not affected. Radiographs revealed L1 compression fracture with a kyphotic deformity. MRI revealed thoracolumbar kyphotic deformity, a syrinx extending from L1 to T12 with obstruction of the subarachnoid space in T-1 weighted images at the corresponding levels (Figure 1A).

Decompressive surgery was accomplished via subdiaphragmatic retroperitoneal corridor with partial corpectomy of L1 vertebra and removal of the offending compressive bones. Arthrodesis was done with a rib graft (Figure 1B). Later, posterior spinal pedicle screw fixation was performed from T11 to L3 with inclusion of L1 (Figure 1C). Subsequently, decompressive laminectomy of L1 was conducted, followed with the dural opening. Subarachnoid space at this level was obstructed internally because of severe adhesive arachnoiditis. These scars could be released partially. Therefore, the procedure was completed with small midline myelotomy. Postoperatively dysesthesia improved dramatically, and the progressive course of weakness was stopped and even showed improvement, although tiptoe standing did not change. Post-operative CT showed accurate decompression of the theca (Figure 1D). Two years postoperatively, the implants made of steel were removed, and one of the most caudal screw were pulled out (Figure 1E). MRI taken three years after hardware removal showed persistent collapse of the syrinx (Figure 1F).

CASE 2

A 53-year-old man was admitted because of progressive dysesthesia and weakness of the lower extremity and aggravation of previously disturbed micturition and defeation for ten months. The patient had a history of fall from the height twelve years before admission with L1 vertebral body burst fracture treated conservatively with a body-molded brace. The patient had severe paraparesis at the time of injury, which was gradually improved. Mild weakness of the lower extremity and disturbances of urination and defeation remained.

On admission, neurological examination revealed flaccid paraparesis with a score of 3/5 in the left and 4/5 in the right side. Decreased sensation in the saddle area was noted. Standing radiographs showed L1 vertebral body fracture with late kyphosis (Figure 2A,B). T1 and T2-weighted MRI revealed a multiloculated syringomyelia extending from L1 to T10 (Figure 2C,D). Reconstructed CT demonstrated kyphotic deformity and an intruded bony fragment at the upper end plate of L1 (Figure 2E).

The patient underwent surgery in the prone position. Thoracolumbar instrumentation from T11 to L3 was performed with exclusion of L1. Later, after decompressive L1 laminectomy and PSO of L1, the intracanal intruded bone was pushed back into the osteotomy defect with the aid of a boot-like pusher; subsequently, the deformity was corrected (Figure 2F). Now, the dura was opened in the midline where a thick arachnoid adhered to the dura and encircled the conus parenchyma. These adhesions were released with sharp dissector but with caution and only on the dorsal and lateral aspects of the cord where the scars anterior to the cord were left untouched. Therefore, syringosubarachnoid shunt was considered worthy with regard to the presence of the anterior-located adhesions. This was accomplished after making a small myelotomy at the posterior midline aspect of the cord above the area of adhesions. Subsequently, the proximal end and a shunt were introduced to the syrinx. The distal end of the catheter was inserted into the lumbar subarachnoid space. Simultaneously, the catheter was secured to the dura with a 3/0 silk suture. Then, the dura was closed but tacked to the paravertebral muscles. Postoperative course was uneventful. Three months after surgery, the patient showed improvement in muscle power and dysesthesia where MRI showed collapse of the syrinx (Figure 2G).

Reconstructed sagittal images showed 360 degree decompression and accuracy of the shunt placement (Figure 2H).
Figure 1: A) Showing L1 fracture with thoracolumbar kyphosis, note a syrinx at conus extending one level. B) Lateral plain radiograph after first stage anterior decompression and arthrodesis with rib graft. C) Lateral radiograph after 360 degree surgery. D) Demonstrating the accuracy of 360-degree decompression and stabilization. E) MRI after 5 years showing disappearance of syrinx. Note 360 degree decompression and the rib graft.
Figure 1F: Lateral radiograph 2-years after surgery, showing pullout of the most caudal stainless steel screw.

Figure 2: A,B) showing L1 Denis type B burst fracture with late kyphosis. C,D) T1 and T2 weighted MR sagittal images showing a syringomyelia at conus extending three level, note septation of the syrinx. E) Reconstructed sagittal CT slices, note kyphosis + a retropulsed fragment and canal compromise.
Post-traumatic Syringomyelia at Conus: Report of two Cases

Figure 2:
F) Post operative thoracolumbar radiographs after correction of kyphosis with PSO and instrumentation.
G) Post-operative T2-weighted sagittal image 2 months after surgery, demonstrating collapse of the syrinx.
H) Reconstructed sagittal CT images showing the accuracy of PSO and syringosubarachnoid shunt in place.
**DISCUSSION**

Late formation of syringomyelia after spinal cord injury is considered an infrequent pathology. Holmes in 1915 was the first who report PTS (27). This was paid less attention till the report of Barnett in 1966 (5). Since then, reports about this condition were published with increasing frequency. (2,3,5,6,7,9,14,16-19,23,25,26,29,32-34,36,38,39,41,46-48,51,52,54-56,61,65). In the last two decades, PTS was more commonly discovered because of the more frequent use of MRI (9,18,20,25,29,30,48,53). The cervical and upper half of the dorsal spine are the most common site for the formation of post-traumatic syrinx where the least frequent location for its development is the conus region (1,8,13,26,40,49,52). Moreover, in large series, details of PTS at the cervical and upper dorsal area are mostly described, where less attention is paid to the syrinx formation at the conus (2,3,5,6,7,9,14,16-19,23,25,26,29,32-34,36,38,39,41,46-48,51,52,54-56,61,65). With review of the clinical and imaging features of a few case reports about PTS at conus confirmed that all has developed subsequent to either by ignorance of the fracture or its conservative treatment (1,8,13,26,40,49,52).

Males outnumber females by far, reflecting the higher susceptibility of the former group to trauma. Pathogenesis of PTS is an interesting topic, and several theories have been proposed in its formation. The more accepted one is that cavity formation is partly due to liquefaction of the cord at the site of injury or resolution of hematomyelia and localized necrosis because of circulatory disturbances in the pia-arachnoid (9,10,11,15,21,24,28,62,65). Moreover, it is postulated that localized adhesive arachnoiditis at the site of trauma is responsible for misconduct of the normal pulsation of the CSF toward the spinal cord parenchyma with subsequent increased interstitial fluid, which ultimately results in the formation of a cystic cavity within the spinal cord and, ultimately, a post-traumatic syrinx. The cavitation is most frequently initiated at the level of the fracture in the narrow zone of the gray matter between the dorsal horns and posterior columns, which is relatively avascular. This zone is located between the dorsal and ventral arterial supply and is also relatively deficient in connective tissue. The cavity may subsequently break down and communicate with the central canal and forms a syrinx (9,10,11,15,21,24,28,62,65).

In the conus region, expansion of the syrinx occur in the rostral direction as a result of transmission of the distensile pressure pulses within the epidural veins because of coughing, sneezing, straining, or exercise (1,8,13,26,40,49,52). The mechanism of this event is described on the basis of slosh and suck phenomena (62,63).

Nowadays, attention is focused not only on syrinx formation but also on post-traumatic deformity and local canal stenosis with spinal cord compromise at the site of initial injury. Therefore, reduction and alignment in thoracolumbar injuries will eventually prevent PTS formation even in complete cord injuries (4,32,57,58). Another major factor responsible for the symptoms is cord tethering because of localized scarring (16,19,32,37,44). Therefore, in patients with thoracolumbar fractures, regular evaluation is not necessary only for early diagnosis of PTS but also for detection of post-traumatic kyphosis as well as Charcot spine, particularly in the patients with occult posterior ligamentous complex injuries. Pathologically, the syrinx is lined largely by flattened ependymal cells, thought to represent the remnants of central canal ependyma (45). The interval between the trauma, formation of PTS, and onset of new neurological symptoms is quite variable and might be from two months to more than 30 years after trauma (1-3,5-9,14,16-19,23,25,26,29,32-34,36,38,39,41,46-48,51,52,54-56,61,65).

Clinically, appearance of new neurological symptoms in a patient who has recovered dramatically from a thoracolumbar trauma is indicative of PTS, where appearance of minor changes in tone of the muscles, bladder, bowel, and erectile functions can be due to the formation of a syrinx in a patient with complete cord injury. Less frequently, these events might be due to cord tethering without formation of a syrinx.

By far, pain is the commonest initial symptom of PTS. It is usually a dull pain located at the site of initial injury. With formation of a syrinx at the conus, pain may radiate to the lower limbs or saddle region with intermittent dysesthetic, burning, and stabbing character. Pain might increase by straining, coughing, or sneezing. Conus syrinx usually extends rostral; the extension is mostly limited to 3 to 5 segments, but it might be extended occasionally as far as the upper cervical cord and even medulla (1,8,13,26,40,49). Dissociated sensory disturbances to pain and touch are seen at the saddle region and in the lower limbs. Gait disturbances attributed to increased motor weakness is the third most common feature of a syrinx and is usually associated with loss of deep tendon reflexes where the neuropathic joint is the least common feature of PTS. Moreover, formation of a syrinx at the conus may render the urinary bladder hyporeflexic,
which is presented with increased difficulty in voiding and defecation as the sole presenting feature or in combination with other clinical features of PTS at this certain location. Difficulty in defecation results from decreased sensitivity of the rectum to fecal impaction. Rarely, bowel dysfunction might be so severe that it necessitates a colostomy.

MRI is the best tool for demonstration of the syrinx and its size and extent (1,3,8,9,14,18,20,21,25,26,30,49). The intensity of the syrinx content is compatible with the intensity of the cerebrospinal fluid shown with low intensity at T1 and high intensity at T2-weighted images (3,9,18,20,21,25,26,30). However, demonstration of increased signal intensity in the areas surrounding the syrinx is due to gliosis. The circumferential gliotic bands giving a beaded appearance to the syrinx on the coronal or sagittal images are another common feature of PTS in MRI.

Periodic application of MRI plays a critical role in monitoring of syrinx progression. It is also of great value in finding the most appropriate site for myelotomy and shunt placement if desired.Assessment of surgical outcome can be accomplished with postoperative MRI. However, there is no direct correlation between the size of the syrinx and the intensity of the neurological deficits.

CT with bone window technique in axial and reformatted images can provide the necessary information about actual size of the canal, the intruded bony fragments and details of deformity, which eventually dictate the surgical strategy for restoration of alignment and extent of decompression (20). Surprisingly, the syrinx could be visualized in 9 out of 11 surgically proven PTS in delayed CT myelography, emphasizing on the necessity of its application in the pre-MRI era (20,22,49).

Surgery is recommended for all the symptomatic patients with PTS involving the conus, to minimize the risk of lower limb and sphincter problems secondary to rapid progression of the syrinx (1,8,13,26,49). In the past, it was believed that conservative treatment is acceptable if the disability is tolerable (3,5). To this should add the phenomenon of spontaneous regression of the syrinx, which has been rarely demonstrated. Nonetheless, the treatment should be individualized in each patient based on their history, symptoms, functional status, neurological examination, whole spine standing radiographs, dynamic X-ray, and analysis of MRI features (4,32,43,57,58).

Some of the previously described methods are seldom used now. Needle aspiration of the syrinx often has poor results because of reaccumulation of cyst content, which almost always occur. This is true for myelotomy alone that might be associated with high rate of failure because of its inability to maintain continued drainage of the syrinx, although those who have responded to myelotomy alone are not rare.

However, based on pathophysiological facts of PTS, which are localized arachnoid scarring, cord tethering, and CSF flow obstruction, the treatment should be focused on relieving the obstruction of the subarachnoid space at the level of injury (15,19,32,38). Around 1990 in Los angles and in Hanover, decompression of the subarachnoid space and arachnoid dissection has been performed as the first surgical option. Both these schools of thought focused on release of arachnoid scarring and duraplasty by making a pseudomeningocele finalized by suturing the dura to the muscles (32,38,39).

Great care had to be taken during dural opening to prevent injury to the spinal cord because the cord could be directly adherent to the overlying dura. Thorough release of arachnoid, which is completely encircling the spinal cord is recommended to be done with sharp dissection, and it should be stopped once it is regarded dangerous by the surgeon (19,32,38,47). With successful dissection, pulsation of the cord, which is already attenuated, might be replaced by normal pulsation. Notably, arachnoid dissection should be done only posteriorly and laterally, and one should avoid dissection anterior to the dentate ligament, particularly in a patient with incomplete neurology where disastrous consequences might be followed (19,32,38). Expansive duraplasty is the matter of controversy (32,39). Duraplasty seems unnecessary once myelotomy or diverting procedures are done, but it should be accomplished where the syrinx is not collapsed despite all procedures (39).

Does a decompressive surgery need to be accompanied with a shunting procedure or not has also remained a controversy. Willams and Klekampe, who were pioneers in decompressive surgery, combined their decompression with shunting early in their series. However, later, they stopped using a shunt unless the syrinx remained constant in size without neurological improvement postoperatively. Most believe that complete release seems unlikely whenever adhesions are so severe and therefore, if the syrinx did not collapse subsequent to arachnoid release and detethering, midline myelotomy alone and for placement of a catheter is indicated in the same stage (19,38,41,52). However,
this can be done in another stage where the inefficacy of a decompressive procedure becomes evident for the responsible surgeon postoperatively.

The myelotomy alone or as an entry for the shunt catheter insertion should be done in the areas not affected by arachnoiditis and usually should be accomplished in the midline. The proximal end of the shunt with multiple perforations should be placed in the entire length of the syrinx. CSF-diverting procedures should be followed with placement of the distal end in the subarachnoid space or in the peritoneal cavity known as syringosubarachnoid and syringoperitoneal shunts, respectively (1,19,41,50,54,56,64).

In the past, management of PTS was accomplished with syringoperitoneal or syringopleural shunt as the only solution, without decompression or arachnoid release with acceptable rate of early success (8,23,25,26,50,64). Later, good results were reported following syringo-subarachnoid shunt procedures (38,52,55,61). The advantages of syringo-subarachnoid shunt include not requiring an additional incision, lesser time of surgery, less blood loss, and fewer hospital stay. The CSF content of syrinx diverted to the subarachnoid space of the lower levels was known to be absorbed from the spinal root sleeves. Subsequent to these shunting methods, an alternative technique known as thecoperitoneal shunt was introduced (35,59,60). By this technique, CSF is diverted to the peritoneum from regions cephalad to the obliterated canal, which seems to be very successful at arresting the progressive cyst formation by the way of decreasing intraspinal pressures. Actually, shunting operations remained the treatment of choice at many institutions until the end of the past century (33,47,50,64). However, the patients treated with a shunt-only procedure, despite good short-term outcome, also had a high complication rate (46,50). The shunt-related complications, such as malfunction and infection, impede neurological improvement in the long-term, requiring several shunt revision surgeries in almost half of the patients (33,41,59,60),46,51. Interestingly, shunt blockage is less frequent in syringosubarachnoid compared with syringoperitoneal shunting procedures (33,59,60). Recently, lumoperitoneal shunting for diversion of CSF below the block was proposed on the basis of similar concept with success (42). Both of these procedures might be a good alternative for treatment of PTA in the patients with preserved motor function (32).

In fact, despite initial amelioration of neurological status obtained by shunt regardless of the method, there is the possibility of the shunt blockade, with re-expansion of the cyst and re-appearance of neurological deficits (46,51). Survey of the literature has shown that long-term collapse of the syrinx is seen in only 45% of the cases where in about 10% of the syrinx size remains unchanged. In the remaining 45%, the syrinx size, despite its initial collapse, re-expand to reach to its preoperative size because of blockage.

By far, laminectomy with release of the arachnoid scars and detethering, with or without shunting procedure, is the treatment of choice for PTS (19,32,33,38,39,47,51). However, once the surgeon is not satisfied of arachnoidolysis or when the cyst does not collapse shunting becomes necessary (19,37,38,47). But, management of PTS should not be considered without dealing with correction of kyphosis and restoration of spinal alignment (4,26,38,43,57). Actually, if one takes into account both of these issues, eventually, the chance of good recovery will be enhanced.

Generally, the results of surgery for PTS located at the conus seem favorable. Pain and motor deficits consistently show good response to operative treatment as compared with sensory dysfunction and reflex changes. Agrawal leasal, Lin, Moreover, decompressive surgery often improves induced bladder dysfunction, although it seldom returns to normal values. Eventually, in long-standing cases where degeneration at the site of the syrinx becomes permanent, the symptoms cannot be reversed by the surgery.

Surprisingly, it has been found that no correlation between the reduction in the size of the syrinx and neurological recovery exist; this means that complete drainage of the syrinx might be unnecessary for an optimal clinical result. Therefore, with mild-to-moderate collapse of the syrinx, a good outcome might be obtained.

In conclusion, early correction of kyphosis and restoration of alignment, as well as spinal cord decompression of the already compromised conus, soon after a thoracolumbar fracture, might eventually decrease the chance of PTS. Nonetheless, once it develops, its early detection and prompt appropriate surgery might avert or minimize the potentially devastating effects of this complication. The regular and frequent clinical follow-up of the patients who sustain thoracolumbar injuries and monitoring of images obtained on a yearly basis might result in the early detection of this complication.

Appearance of new neurological symptoms in a patient who had been either asymptomatic or stable for an
acceptable period can be indicative of syrinx formation. Decompressive laminectomy, arachnoidolysis, detethering, and correction of deformity are promising strategy for management of PTS. CSF-diverting procedures should be reserved for the situations where syrinx collapse does not occur with decompressive surgery (19,37,38,47).

REFERENCES


Manuscript submitted July 10, 2013. Accepted April 14, 2014.

Address correspondence to: Abolfazl Rahimizadeh, Pars Hospital, Neurosurgery Pars Hospital, Tehran, Islamic Republic of Iran
Phone: 912 322 61 49
email: a_rahimizadeh@hotmail.com