An Unusual Case of Syringohydromyelia Presenting with Neurogenic Bladder

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Abstract

We report the case of a 4-year-old boy who first presented with acute pyelonephritis at the age of 6 months. Diagnostic workup revealed high-grade bilateral vesicourethral reflux (VUR). At the age of 18 months, a bulking agent was used to treat bilateral VUR. Since the VUR persisted, an open bilateral Lich-Gregoir procedure was done at the age of 3 years. Immediately after surgery, he developed acute urinary retention with hydronephrosis that resolved with the placement of dwelling urinary catheter. After removal of the catheter urinary retention relapsed so placement of suprapubic urinary catheter was indicated since he did not have sensory loss. He was started with tamsulosin (α – 1-blocker) and prophylactic antibiotics. Urodynamics were performed and suggested bladder outlet obstruction. On the basis of previous urethroscopy and the absence of neurological sequelae, the differential diagnosis of Hinman syndrome was made. After removal of the suprapubic catheter, clean intermittent catheterization was started and α-blocker continued. However, magnetic resonance imaging of the brain and the spinal cord revealed syringohydromyelia extending from thoracic spine (Th5) to conus medullaris with 6 to 7 mm in diameter. Electromyoneurogram was normal. After a follow-up of 3 years, the hydronephrosis has resolved. The patient is on clean intermittent catheterization and has no urinary tract infections.

Keywords
► neurogenic bladder
► syringohydromyelia
► vesicourethral reflux

New Insights and the Importance for the Pediatric Surgeon

Syringohydromyelia is a very rare cause of neurogenic bladder in a boy without overt neurological sequelae. This is the first reported case in a child to date.
Introduction

Neurogenic bladder is a dysfunction of the urinary bladder due to disease of the central nervous system or peripheral nerves involved in the control of urination. The most common causes of neurogenic bladder in children are neurospinal dysraphisms such as spina bifida, sacral agenesis, tethered cord, and spinal cord injury. Up to a third of children with neurogenic bladder have vesicoureteral reflux (VUR). In case of neurogenic bladder, the presumed pathomechanism of VUR is a reflex secondary to elevated bladder pressures rather than due to a defective ureterovesical junction. The initial management involves clean intermittent catheterization (CIC) with or without prophylactic antibiotic therapy in combination with an anticholinergic agent. In children with VUR refractory to conservative measures, management includes surgery with ureteral implantation, bladder augmentation, or a combination of treatment methods. Urinary incontinence and bladder dysfunction are rarely described as the first manifestation of syringomyelia, a fluid-filled, gliosis-lined cavity within the spinal cord prevalent in 8.4 cases per 100,000 children. Hydromyelia refers to a fluid-filled cavity within the spinal cord lined by ependymal, which likely results from a developmentally nonobliterated central canal. The two terms are often interchanged. The aim of this article was to present an unusual case of syringomyelia that presented with neurogenic bladder without apparent neurological sequels.

Case Report

A 1.5-year-old boy was referred to our clinic for the endoscopic treatment of bilateral VUR (grade V on the left side and grade II/III on the right side). He was born after a third pregnancy and was healthy until the age of 6 months when he presented with an episode of acute pyelonephritis. Since spontaneous remission did not occur, instillation of bulking agent bilaterally was done at the age of 18 months. Cystoscopy revealed a bladder with trabeculation. Six months after endoscopic treatment, a contrast-enhanced voiding urosonography was done that filled, gliosis-lined cavity within the spinal cord and revealed hydromyelia extending from thoracic spine (Th5) to conus medullaris with 6 to 7 mm in diameter. There were no signs of Chiari 1 malformation on brain scans. Electromyoneurography (EMNG) of the lower extremities was normal. Neurosurgical consultation was done. After a follow-up of 3 years, the hydronephrosis has resolved. The patient is on CIC and has no urinary tract infections. The follow-up ultrasonography demonstrated the rightsided pathology, which can be managed expectantly. Singhal et al also concluded that syringomyelia often remains stable in patients receiving nonoperative treatment. On the basis of first urodynamic study, which was unremarkable, endoscopic antireflux surgery was first treatment of choice for our patient. This procedure has high success rate in primary VUR, while success rates in neurogenic bladder patients have been reported from 53 to 86%. Furthermore, this procedure is less effective in higher grades of reflux and success is generally transient rather than permanent; so patients require long-term follow-up. Since our patient had a persistence of VUR after using the bulking agent, we chose to perform ureteral implantation. At this point, we did not suspect that our patient could have neurogenic bladder since we did not question the validity of first urodynamics. With adequate bladder capacity (% estimated bladder capacity > 70%) and compliance (> 7 mL/cm H2O), high grades of reflux have been treated with ureteral implantation alone. Postoperative urinary retention after bilateral ureteral reimplantation is common, but is usually transient, what was not the case with our patient. The differential diagnosis of Hinman syndrome was made on the basis of normal neurological examination and MRI at that moment was not available. It is unusual for syringohydromyelia to present with neurogenic bladder only; it usually presents with back pain, brachial amyotrophy, dissociated sensory loss, and neurogenic arthropathies. Finally, Hinman syndrome, non-neurogenic neurogenic bladder, could still be a differential diagnosis if we consider IS as a benign condition, especially with other neurological sequelae absent and EMNG of the lower extremities normal.
Conclusion

Neurogenic bladder as the first and only manifestation of syringohydromyelia is rare and can mislead diagnostic workup and delay appropriate therapy. Proper neurologic examination, including MRI, should be done in patients with neurogenic bladder.

Conflict of Interest
None.

References
1 Dorsher PT, McIntosh PM. Neurogenic bladder. Adv Urol 2012; 2012:816274
7 Klekamp J. How should syringomyelia be defined and diagnosed? World Neurosurg 2018;111:e729–e745


