

Hinman Syndrome - Diagnosis and Management: A Rare Case Report

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Hinman syndrome or non-neurogenic neurogenic bladder is a rare clinical condition which is characterized by non-neurogenic urinary bladder dysfunction. It is a diagnosis of exclusion and patient presents with characteristics of a neurogenic bladder with external sphincter dyssynergia but without evidence of any neurologic alteration. The underlying pathology is not known. Bladder training and medical treatment have been recognized as effective management. However, when there is established damage to the upper urinary system or chronic renal failure, surgery is preferred over conservative treatment. Due to the low incidence of the disease, there is still no consensus for the most adequate treatment or management. We report the case of a pediatric patient presented with recurrent retention of urine with dysfunctional voiding and compromised renal functions. As patient already presented with upper urinary tract damage hence underwent successful surgical treatment for the same.

Keywords: Chronic renal failure, Hinman syndrome, Non-neurogenic neurogenic bladder

INTRODUCTION

Hinman syndrome, also known as non-neurogenic neurogenic bladder syndrome, is a rare entity characterized by non-neurogenic urinary bladder dysfunction. The majority of patients are children in early infancy or in preschool age.¹ Diagnosis of this clinical entity is made by exclusion and is assigned to those children that have clinical, radiologic, and urodynamic characteristics of a neurogenic bladder with external sphincter dyssynergia, but with no evidence of a neurologic alteration or disorder. Patients with Hinman syndrome generally presents with recurrent urinary tract infections, urinary incontinence, vesicoureteral reflux, hydronephrosis, and acute and chronic renal failure. Due to the low incidence of the disease, there is no consensus regarding the management and treatment. In 1973, Hinman and Baumann were the first to report a case series of 14 patients with non-neurogenic neurogenic bladder syndrome and treated those patients with bladder training.² Some clinicians have established the effectiveness of bladder retraining and medical treatment

for its management, while others have employed different treatments such as Botox application in the external sphincter with good results. However, when there is already damage to the upper urinary tract or risk for the same then more aggressive surgical treatment is preferred over conservative treatment in order to prevent chronic renal damage.³

CASE REPORT

A 5-year-old female child presented with complaints of dribbling of urine since - 1 year, lower abdominal distension with incomplete voiding since - 3 months and on and off fever since - 15 days. No history of trauma and constipation. The patient underwent cystoscopy for the same complaints 1 year back. No records of the procedure available. Development milestones were normal. On examination, child moderately built and nourished, well-oriented to time, place, person. Weight - 18 kg, height - 85 cm, gait - Normal, afebrile, vitals stable. A abdominal examination urinary bladder was palpable. External genitalia and spine were normal. The patient was catheterized with Foleys 10 Fr catheter and 300 ml of clear urine was drained. The patient was investigated, hemoglobin - 7.7 g%, urine culture and sensitivity-*Acinetobacter* species detected, serum creatinine - 4.9 mg/dl, X-ray kidneys-ureters-bladder (KUB) - Normal, ultrasonography KUB - Bilateral moderate hydronephrosis with thickened urinary bladder.

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Micturating cystourethrogram was normal. Urodynamic study done suggested of high detrusor pressure with detrusor over activity (Figure 1). Magnetic resonance imaging spine done was normal (Figure 2).

With this evaluation, the patient was diagnosed to have Hinman syndrome, i.e. non-neurogenic neurogenic bladder as the patient had functional bladder outlet obstruction without any neurological abnormalities and planned for surgical intervention. Patient underwent mitrofanoff procedure with augmentation ileocystoplasty.

Surgical Technique

Low midline incision given. Mobilization of the ascending colon along the line of Toldt done to gain mobilization of the appendix and its mesentery. Once the cecum has been mobilized the base of the appendix is amputated, leaving a small cuff of cecum with the appendix (Figure 3). Use of the cuff at the stoma may decrease the risk of stenosis. The cecum is closed in a fashion similar to an open

appendectomy. To perform the bladder augmentation, it was used 15 cm of ileum, 20 cm from the ileocecal valve that was opened longitudinally and reconfigured into a “U” shape (ileocystoplasty) with continent derivation (Mitrofanoff’s principle). The distal end of the appendix is tunneled and implanted into a posterolateral position within the bladder (Figure 4). The appendix brought up to reach the umbilicus without tension where stoma is made. Wound closed in layers. Post-operative period was uneventful. Sutures removed on post-operative day 8th. The patient was trained with self-clean intermittent catheterization through appendiceal stump, present at umbilicus. On follow-up, patient was asymptomatic, creatinine reduced to 1.5 mg/dl and ultrasound suggested normal kidneys with no hydroureteronephrosis.

DISCUSSION

Hinman syndrome or non-neurogenic neurogenic bladder, the most severe form of dysfunctional voiding, is a pure functional bladder outlet obstruction without any neurological abnormalities and can present in early infancy

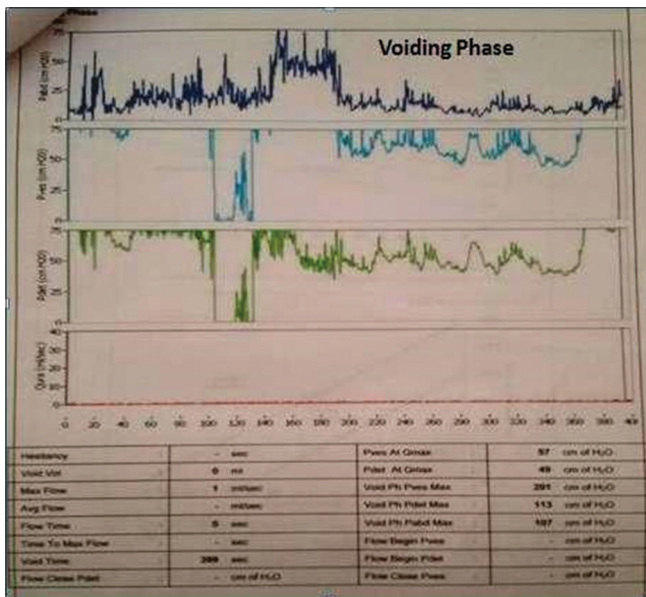


Figure 1: Urodynamic study - high detrusor pressure with detrusor over activity

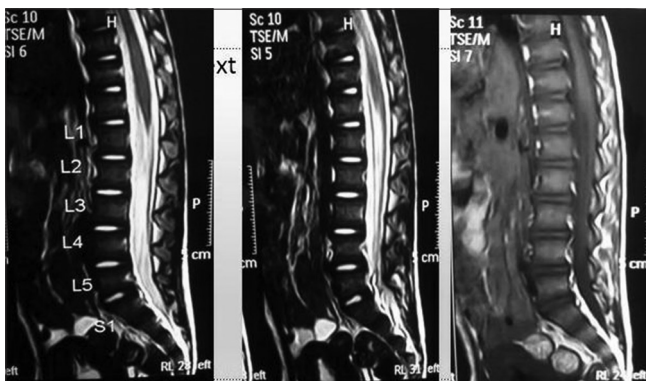


Figure 2: Magnetic resonance imaging lumbar spine – no neurological abnormality detected



Figure 3: Appendix mobilized with its mesentery

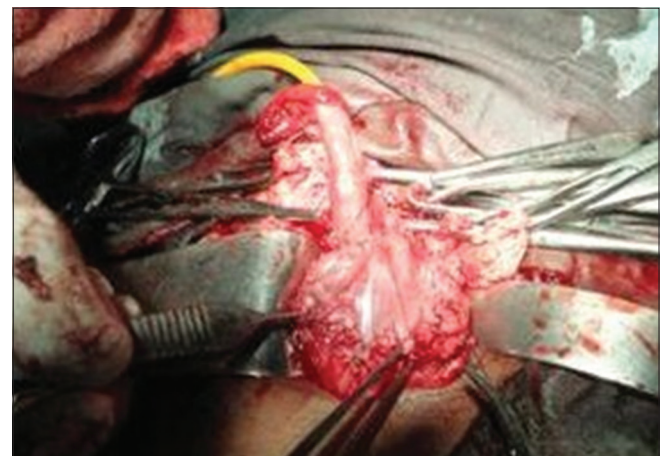


Figure 4: Appendix implanted in bladder with augmentation ileocystoplasty

or in older children around 12 years of age. The detrusor muscle maintains a long period of hyperactivity in relation to the infravesical obstruction, which ends up producing detrusor decompensation leading to upper urinary tract damage and renal failure at an early age. The majority of patients present with recurrent urinary tract infections, difficulty in micturition, urinary incontinence and the presence of residual urine as the most frequent symptoms. If early diagnosis is not established the detrusor muscle and upper urinary tract may become damaged, showing radiographic data of a thickened bladder, hydronephrosis, vesicoureteral reflux and loss of the renal parenchyma.⁴ Conservative treatment is recommended in patients with the first stage of the disease which includes bladder training, a strict bladder voiding schedule, psychologic support, anticholinergic medication, and clean intermittent catheterization, all of which are effective if strictly adhered to by both the patient and family members.⁵ If the conservative management fails in such patients than slightly more invasive treatment modalities are employed.⁶ Mokhless *et al.* reported on the application of Botox in the external urethral sphincter of 10 patients with Hinman syndrome. Results of this modality were good without any complications.⁷ Recently, some authors described the use of transcutaneous electrical bladder stimulation for treating Hinman syndrome. Rashid *et al.* reported that Hinman syndrome is a condition in which the bladder empties infrequently due to a lack of coordination between the sympathetic and parasympathetic activity causing a contraction in the external urethral sphincter muscle that leads to infravesical obstruction. They evaluated 24 patients with Hinman syndrome, all above the age of 12 years, and divided them into two groups. The first group was managed only with traditional treatment and the second with transcutaneous neurostimulation plus traditional treatment. Urinary symptoms were evaluated after 12 weeks of treatment, and there was a noticeable improvement in the patients that received neurostimulation. However, the follow-up period was very short, and the urinary symptoms did not completely disappear.⁸ Even with promising results, neuromodulation and Botox application must have long-term evaluations with controlled randomized studies in order to demonstrate their efficacy in children with Hinman syndrome. The prevention of progression into renal disease and protection of upper urinary tract are the principal aims of treating this condition. When initial treatment with bladder reeducation, psychologic support, clean intermittent catheterization, or some of the new techniques such as Botox application or transcutaneous neurostimulation have failed, and the upper urinary tract is at risk, more aggressive treatment needs to be undertaken to protect the renal function. It has been suggested that when there is established renal damage, conservative treatment

should be abandoned and the patient should be managed with surgical treatment to prevent further deterioration of renal functions. At present, there are no case series analyzing which is the best surgical technique for these types of patients. In our patient, we did mitrofanoff procedure with augmentation ileocystoplasty, and the later patient was trained in doing clean intermittent catheterization through the appendiceal stump. This procedure had good results on follow-up, however, its a major and complex surgical procedure and requires experience and expertise.

CONCLUSION

Hinman syndrome, or non-neurogenic neurogenic syndrome, is a urinary dysfunction with a lack of bladder-sphincter coordination. Hinman syndrome is a diagnosis of exclusion and underlying pathology not known. This produces damage to the upper urinary tract if it is not adequately diagnosed and treated. Conservative management is accepted as an initial treatment but when there are altered renal functions, surgical treatment is necessary in order to prevent chronic renal failure.

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