Neurogenic bladder in Hunter's syndrome

Kojiro Koyama, Yasuaki Moda, Atsushi Sone, Hiroyoshi Tanaka, Yousuke Hino

Abstract

We encountered a rare patient with Hunter's syndrome who exhibited urinary retention as a result of a neurogenic bladder, uninhibited detrusor contractions, and detrusor-sphincter dyssynergia. Neurological findings were consistent with cervical myelopathy and cervical MR imaging showed very narrow segments at the cord level C2-4. We speculate that this Hunter's syndrome patient has cervical myelopathy and that this neurological dysfunction causes the neurogenic bladder.

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The mucopolysaccharidoses (MPS) are inborn errors of metabolism and inherited lysosomal storage disorders which are classified into six distinct groups and several subtypes based on biochemical, clinical, and genetic factors. The pathological features of MPS vary. They include a chronic and progressive course, multi-system involvement, organomegaly, dysostosis multiplex, and abnormal facies. Hearing, vision, cardiovascular function, and joint mobility may be affected.

Although there have been many reports of Hunter's syndrome since Hunter's original description, previous reports have not described neurogenic bladder in association with this syndrome. In addition, there has been no previous description of a urodynamic study of this disorder.

A 19 year old Japanese male suffering from acute urinary retention was admitted to our department in July 1991. Before his admission, he had experienced difficulty in voiding, gait

disturbance, and weakness in his upper limbs. At 10 years of age, he was diagnosed as having Hunter's syndrome based upon a deficiency of lymphocytic iduronate sulphatase and increased urinary excretion of dermatan sulphate and heparan sulphate. Neurological findings were consistent with cervical myelopathy which indicated muscle weakness in both upper and lower extremities, incomplete paraplegia, and exaggerated deep tendon reflexes in all four limbs.

During the urodynamic study, the cystometrogram (CMG) showed uninhibited detrusor contractions and the sphincter electromyogram (EMG) showed enhanced sphincter activities, that is, detrusor-sphincter dyssynergia (DSD, figure). Neuroradiographic examination with MR imaging showed significantly narrow segments at the C2-4 level.

Based on the results of these neurological, urodynamic, and neuroradiographic investigations, we concluded that the DSD caused complete urinary retention and was induced by cervical myelopathy.

Patients with Hunter's syndrome usually have neurological system damage, with sensory and motor disturbance and an increase of deep tendon reflexes in both the upper and lower limbs as signs of cervical myelopathy.¹⁻³ This neurological dysfunction is presumably the result of cervical myelopathy, indicated by very narrow segments at the C2-4 level in a cervical MRI scan.

The CMG of a patient with neurogenic bladder owing to cervical myelopathy generally shows uninhibited detrusor contraction without synchronous external sphincter relaxation.^{4,5} In this case, the CMG showed uninhibited detrusor contraction and the

Department of
Urology, Kawasaki
Medical School, 577
Matsushima,
Kurashiki 701-01,
Okayama, Japan
K Koyama
Y Moda
A Sone
H Tanaka

Department of
Orthopaedics,
Kawasaki Medical
School, Kurashiki
701-01, Japan
Y Hino

Correspondence to
Dr Koyama.

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CMG shows uninhibited detrusor contraction without coordinated sphincter relaxation: detrusor-sphincter dyssynergia (DSD). FDV=first desire to void. UIC=uninhibited detrusor contraction.

EMG showed detrusor-sphincter dyssynergia. These findings are commonly seen in patients with neurogenic bladder caused by suprasacral spinal cord damage, especially cervical cord stenosis.

Furthermore, neurological and neuroradiographic findings in this patient supported the fact that he had suprasacral spinal cord damage.

Possible causes of a narrow cervical spinal canal are (1) cervical bony canal stenosis, (2) some degree of atlantoaxial subluxation or dislocation, and (3) dural or (4) ligamentous thickening.^{1-3,6} These abnormalities greatly narrow the effective lumen of the spinal canal throughout the entire cervical region. Concentric impingement on the cervical subarachnoid membranes has also been described.

Although no previous reports have described neurogenic bladder in Hunter's syn-

drome, we believe that the prevalence of Hunter's syndrome patients who suffer from bladder dysfunction is high. Since many of the organ dysfunctions associated with Hunter's syndrome are progressive, central nervous system damage occurs gradually. Under these conditions, neurogenic bladder or voiding dysfunction may be overlooked or disregarded.

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