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Urinary Incontinence in Children as a Sign of Neurological Disorders: Need for a High Index of Suspicion

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Syringomyelia; tethered cord; enuresis; void disorder

INTRODUCTION

Childhood enuresis is a common occurrence, affecting 7-20% of children[1, 2]. While more frequently cases of enuresis have ties to behavioral, psychiatric[3] and genetic[4] predispositions, less common causes, including neurologic conditions, should not be overlooked. Given the association of neurologic problems with enuresis, the importance of thorough neurologic evaluation cannot be underestimated, as findings could suggest a neurologic etiology, particularly in cases refractory to traditional management.

We present three pediatric cases of enuresis related to neurologic disorders in order to illustrate the importance of a high index of suspicion for neurological causes and a thorough neurologic history and examination in the evaluation of enuresis.

In the cases presented, while all underlying diagnoses were promptly recognized by specialists, symptoms and signs were overlooked during general examination by nonspecialist.

PATIENTS PRESENTATION

Case 1 – Benign Rolandic Epilepsy (BRE)

An 8-year-old female presented with persistent bedwetting and occasional daytime incontinence. Her past medical history was notable only for vesicoureteral reflux, chronic otitis media with tympanostomy, and constipation. Because she would wait “until the last minute” to urinate, behavioral measures first implemented included fluid restriction before bedtime. Trials of antibiotics and antispasmodics were discontinued after a voiding cystourethrogram showed an insignificant amount of reflux. Past physical examination by

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the family doctor showed normal external genitalia and positive anal reflex with good tone, and unremarkable neurological examination.

At age 8, to the persistence of her urinary issues, she was started on imipramine, without benefit. Around that time she presented to our clinic following a brief episode of right sided facial twitching during sleep with deviation of the mouth to the right. The mother, who witnessed the episode, reported 30-seconds duration and no incontinence. When asked regarding occurrence of similar symptoms, the patient recounted an episode six months prior when after a bath she had similar lip twitching that lasted for 10 to 15 seconds. There were no headaches, bowel or bladder incontinence during or following the episode and no personal or family history of seizures. EEG showed multifocal epileptiform discharges more prominent in the fronto-parietal area, consistent with BRE.

The child was started on oxcarbazepine; a 3 months follow-up revealed very rare nighttime incontinence.

Case 2 – Cervical Syringomelia

A 13 year old girl with multi-year history of bladder problems with bed wetting, daytime incontinence, and urgency presented with a few month history of progressive hand atrophy with diminishing grip leading to difficulty opening jars and writing. Her parents noticed thinning of her shoulders with prominent scapulae. Over this time she was active in cheerleading and participated in tumbling classes. Two years prior to evaluation she sustained a fall from a pyramid during cheerleading exercises, suffering a trauma to her neck.

Physical exam showed a small girl with normal cognition. Her neck was non-tender with full range of motion. On musculoskeletal examination, moderate-to-severe atrophy of the intrinsic hand muscles was evident along with some atrophy of the shoulder muscles with scapular winging. Grip was weak with strength 3/5, and she lacked full finger extension. Lower extremity strength and gait were normal with adequate tandem walk and normal reflexes. Plantar responses were normal. MRI revealed a large cervicothoracic syrinx extending from C1 into the low thoracic region with associated Chiari I malformation (figure 1). Neurosurgical evaluation prompted Chiari decompression with C1 and C2 laminectomy and subarachnoid shunt placement. Although a post-surgical cervical spine MRI (figure 1) showed radiographic improvement of the structural abnormalities, the patient did not regain strength and muscle bulk in her hands.

Case 3 – Tethered cord

A four year old female presented with complaint of chronic headaches exacerbated by activity, including laughing, crying, and running, without associated back pain.

Developmentally, she began walking at appropriate age, though her gait was mildly abnormal, with a pace slow for age and with difficulty climbing stairs. The remainder of her development was unremarkable; toilet training occurred at age three, but she continued to have ‘accidents’ of urinary incontinence about once per month, as well as occasional nocturnal enuresis. The more recent onset of severe urinary urgency and frequency

associated with decreased stream and dribbling of urine led to evaluation by nephrology. A renal ultrasound and voiding cystourethrogram showed a normal bladder without signs of vesico-ureteral reflux, and a diagnosis of dysfunctional voiding syndrome was made.

Neurosurgical consultation was obtained and on physical examination she was noted to have a broad-based gait, mild difficulties with tandem walk, and a birthmark at the L5 level. Brain MRI showed a Chiari I malformation with tonsillary protrusion of 13 mm without hydrocephalus. Spine MRI revealed a conus medullaris ending at L2 with a small syrinx; the filum terminale appeared adhesive to the dorsal dura at L4-L5 (figure 2). Laminectomy at L4-L5 for tethered cord release and craniectomy with C1, C2 and C3 laminectomy for Chiari decompression were performed. In the month following surgical intervention, the patient's gait improved, but her headaches worsened

DISCUSSION

The significance of the cases presented lies in understanding that the diagnosis was reached only because of a high index of suspicion leading to more accurate neurologic history and examination. Diagnosing BRE in Case 1 was prompted by the classical nocturnal symptoms but the possibility of nocturnal seizure was never considered during multiple evaluations for persistent, nocturnal urinary incontinence. Although no incontinence was reported with the witnessed seizures, the substantial improvement of enuresis with antiepileptic medication suggests a relation between the two events. Because of the relatively common occurrence of BRE in school-age children, the possibility of nocturnal seizures should be considered in cases of persistent incontinence in this age group. Care should be taken to ask about specific symptoms like nocturnal twitching, staring spells, or episodes of altered mental status in the morning that could suggest epilepsy as cause of incontinence.

The identification of a cervical syrinx in Case 2 was driven by an accurate neurologic examination with the identification of clear findings localizing the problem at the cervical spine level. In this patient the long-lasting urinary incontinence was dismissed as common childhood developmental condition. The importance of her head trauma –which is known to precipitate worsening of cervical syrinx in the setting of Chiari I malformation - was also underestimated.

The diagnosis of a Chiari I malformation and tethered cord in Case 3 as a cause for the urinary incontinence relied on the persistent headaches and the subtle gait-related findings. While incontinence occurs in up to 20% of four-year-old patients[1], it was again the combination of multiple neurologic complaints: urinary incontinence, chronic headaches, and mild abnormalities on exam that suggested an underlying neurologic cause.

While purely neurologic causes only account for a fraction of childhood enuresis, the importance of a thorough history and neurologic examination cannot be underestimated. General practitioners should be vigilant regarding signs and symptoms suggesting underlying neurologic problems as etiology to common childhood diseases. These cases illustrate that performing a thorough neurological history and examination in patients with enuresis can be greatly beneficial as part of the initial evaluation. Furthermore, as neurologic

signs and symptoms may be indicative of other rare childhood diseases, neurologic examination at every well-child visit should never be overlooked.

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Figure 1.

Cervical and thoracic spine MRI. A: Extensive holocord syringomyelia with Chiari 1 malformation at the craniocervical junction (ectopic cerebellar tonsils extending 1.4 cm below the foramen magnum). B: after surgery, the syrinx appears smaller in comparison to pre-surgical evaluation. A syringe-subarachnoid shunt is visible. 25×22mm (300 × 300 DPI)

Figure 2.

[A] Brain MRI: T1 image demonstrating Chiari malformation with 13 mm tonsillary protrusion, but no hydrocephalus; [B] T2 lumbo-sacral spine MRI: conus medullaris ending at the level of L2 and filum terminale adhesive to the dorsal dura at L4-L5. 90×48mm (300 × 300 DPI)