Urological Problems in Tethered Cord Syndrome

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Tethered cord syndrome is infrequently seen in out-patient urological department as urological problems do not present as the initial signs or symptoms for this syndrome. The symptom of voiding dysfunction is frequently confused with other causes of urinary incontinence. Due to long-term complications which can result in hemodialysis, early diagnosis of tethered cord syndrome is important. The aim of this study was to investigate the clinical manifestation and severity of tethered cord syndrome. From January 1995 through April 1999, we retrospectively collected the medical records of 5 patients with tethered cord syndrome who had an average age of 30 yrs. They all presented with urological symptoms as the initial call for help. Four patients had predominant symptoms of lower abdominal fullness and pain. Detrusor hyperreflexia with detrusor-sphincteric dyssynergia were seen in 4 patients. One patient presented with detrusor hyporeflexia and poor compliance as seen on the cystometrogram. All patients with tethered cord syndromes were diagnosed with magnetic resonance imaging (MRI). Untethering surgeries with removal of lipoma were performed in 4 patients. One patient developed hemodialysis during the 1-yr follow-up period. Two patients showed persistent chronic renal insufficiency. Only 1 patient maintains normal renal function because of early diagnosis and early treatment. This result indicates that prognosis is poor when the diagnosis is delayed. We suggest early diagnosis and early treatment as the preferred way to resolve the problem of progressively worsening renal function. Because only a small number of patients were available, more studies of tethered cord syndrome would be helpful to clarify the clinical presentations of urological problems. ***(J Urol R.O.C., 11:6-11, 2000)***

Key words: *tethered cord syndrome, neurovesical dysfunction, detrusor hyperreflexia, intraspinal lipoma.*

INTRODUCTION

Tethered cord syndrome is diagnosed in patients with neurogenic sequalae and urological problems of neurovesical dysfunctions, [1] which are caused by mechanical stretching and ischemic injury to the spinal cord or nerve roots due to differential growth rates between the bony vertebrate and neural elements.[2] Myelomeningocele and occult spinal dysraphism are associated with tethering of the spinal cord.[3] Spinal dysraphism encompasses a wide range of developmental defects of the spinal column, including open defect of the myelomeningocele and closed occult dysraphic states, such as lipomyelomeningocele, diastematomyelia, thickened film terminale, dermoid sinus, and lipoma of the terminal spinal cord.[4] Tethering may also be a secondary phenomenon after surgery or infections of the spinal cord.[5] Diagnosis of occult spinal dysraphism is commonly suggested by a host of abnormal physical findings, including a patch of hairs, subcutaneous lipoma, dimple or dermal sinus, or increased skin pigmentation in more than 75% of patients.[4] However, the urological symptoms may be occult and mimic other causes of incontinence. That could delay the time of diagnosis.

Orthopedic or neurological symptoms of tethered cord syndrome may include lower extremity weakness, back pain, worsening deformities, or scoliosis.[6] The urological symptoms are varied and include voiding dysfunctions, such as urinary incontinence, frequency, urgency, urinary retention, urinary tract infection, hydronephrosis, or vesicoureteral reflux.[7] These symptoms may worsen if the diagnosis is delayed which could become end-stage renal disease and finally long-term hemodialysis. Thus, early diagnosis of tethered cord syndrome is crucial in the prevention of renal

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function deteroriation. The aim of this study is to define the early urological signs of tethered cord syndrome.

PATIENTS AND METHODS

We retrospectively collected the medical records of 5 patients with tethered cord syndrome from January 1995 through April 1998 who were diagnosed in this hospital. All of them had visited our urological out-patient department for the initial diagnosis. There were 3 men and 2 women with an average of age 30 of yr (ranges 22 to 42 yrs). All of them had symptoms of vesical dysfunction. Of the 5 patients, 4 were diagnosed with tethered cord syndrome as an adults and 1 had had previous spinal surgery due to myelomenigocele at the age of 5 yrs. The presenting clinical signs and symptoms were initialized in the urological out-patient department. The urological symptoms were recorded during hospitalization. Tethered cord syndrome was diagnosed using magnetic resonance imaging (MRI) in all patients. All patients underwent complete neurological and urological evaluations. Four of them received pre-operative or post-operative urodynamic studies.

RESULTS

The most common presentations of urinary symptoms was lower abdominal pain and fullness in 4 patients. The average duration from symptom onset to diagnosis was 14 mo on average (range, 8 mo to 2 yr). Frequency and dysuria were complaints in 3 patients. Two patients developed acute urinary tract infection and presented with fever, frequency, scrotal swellings and pyuria. In addition, they had had symptoms of frequency, incontinences and nocturia for 1 yr. Urodynamic studies of cystometrography (CMG) and external sphincteric electromyography (EMG) revealed detrusor hyperreflexia with detrusor-spincteric dyssynergia (DSD) in 4 patients (Fig.1). Detrusor hyporeflexia and decreased sensation was found in 1 patient (Fig.2). Hydronephrosis of bilateral kidneys with reflux was seen in the patients with detrusor hyperreflexia. One patient had a history of myelomeningocele and had undergone surgical correction when she was 5 yr old, but she had had no regular follow up after surgery, and lower abdominal fullness had lasted for 1 yr until the diagnosis at the age of 33 yr old. One patient had a history of spinal trauma when he was 10 yr old with a sequala of lumbar scoliosis. Only 1 patient had serum renal function test results within the reference range; the others revealed chronic renal insufficiency with an average blood urea nitrogen of 50 mg/dl and creatinine of 5 mg/dl. All anal tone examination results showed loose and poor bulbocavernosal reflex.

Abnormal findings along the spinal column were found by MRI in all patients with tethered cord syndrome. MRI revealed intraspinal lipoma between L5 and whole sacrum in 4 patients (Fig.3A). Spinal dysraphism and dural ectasis were found in 4 patients (Fig. 3B). Three patients had spinal bifida as shown on MRI (Fig.3C). Myelomeningocele presented in 1 patient. Surgical neurolysis was performed in 4 patients. The procedure including laminectomy, duraplasty, and laminoplasty in 3 patients. All operative patients had tumors in their spinal canals which were excised, and pathological tests revealed lipoma. Postoperative clean intermittent self-catheterization (CISC) was used in 3 patients. One patient required permanent hemodialysis in spite of neurosurgical treatment. Only 1 patient has normal renal function and improved voiding symptoms after surgery. The symptoms remained at least 9 mo in the patients. The remaining patients revealed no further worsening of renal function during the 1-y follow-up period. The follow-up cystometrography revealed stable after-surgical intervention in those patients with detrusor hyperreflexia. The clinical correlations of general data, symptoms, treatment, and outcome of patients are listed in Table 1.

DISCUSSION

Tethered cord syndrome is infrequently seen in patients when visit our-patient urological department for initial examination. Only 10% of patients reported having urological problems as initial symptoms.[7] The occult symptoms may delay the time of diagnosis with a serious complication such as permanent hemodialysis

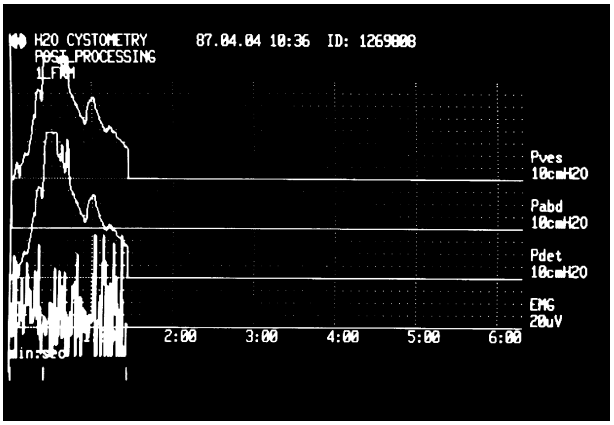

Fig.1 Detrusor hyperreflexia with detrusor sphincteric dyssynergia (DSD) showing on the cystometrograph of a patient with tethered cord syndrome.

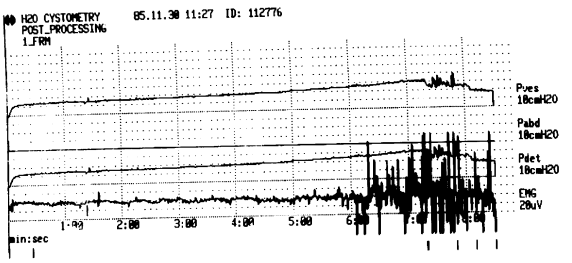

Fig.2 Urodynamic study of cystometrography with electromyelography showing detrusor hyporeflexia with DSD in patients with tethered cord syndrome.

Fig.3A Spin echo T1-weighted image (TR/TE = 560/25 ms, thickness 4 mm) on sagittal section showing tethered cord with low position of the conus medullaris at the L5 level (long arrow). Note also the adjacent lipoma (short arrow).

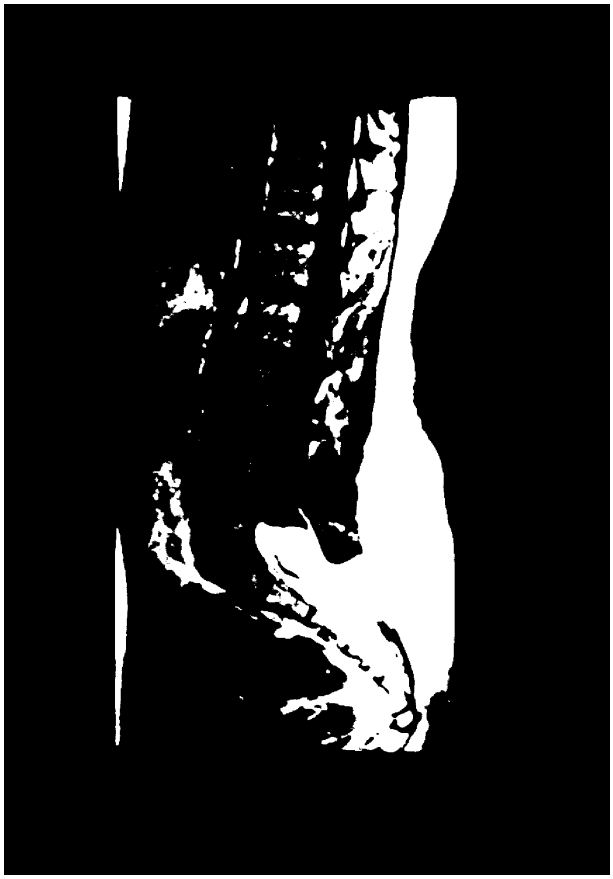

Fig.3B Same pulse sequence but one section left lateral to 1A. A huge bright-signal lipoma is protruding from the spinal defect anteriorly into the spinal canal.

Fig.3C Gradient echo T2*-weighted image (TR/TE/ flip angle = 647/27 ms/20 degrees) on axial section clearly demonstrating the lipoma (short arrow) extending from the back via the spinal bifida (long arrow) into the spinal canal. Note that the signal intensity of both the bone marrow and the lipoma appear as intermediate signals at this pulse sequence.

due to a progressively worsening renal function. It can also lead to irreversible neurological complications such as lower limb abnormalities.[8] Due to the variety of symptoms and poor outcome of patients without definite treatment, early identification of the syndrome is important. The aim of this study was to investigate the presentation of urological symptoms of tethered cord syndrome. This evidence may lead to improved early diagnosis of tethered cord syndrome

The result show that MRI can be used as a reliable diagnostic tool for tethered cord syndrome. Myelomeningocele, dural ectasis, spinal dysraphism and an occasional tumor mass were well demonstrated on MRI. MRI not only provided a definite diagnosis but also differentiated various lesions such as hydromyelia, diastematomyelia, arachnoid cysts, dermoid cysts, and tumors.[9]. Since patients with occult spinal dysraphism may present with dermal sinus, hair patches, hemangiomas or soft tissue tumors, it is important to perform MRI for early diagnosis of tethered cord syndrome.[4]

There was no obvious presentation of urine incon-

Table 1. Clinical data of patients with tethered cord syndrome

P't. n	Age (yr)	Sex	Duration of symptoms	Urodynamic findings	MRI findings	Operation	(BUN/Cr.)	Outcome
1	30	F	9 mo	Hyporeflexia, decreased compliance	Lipomeninogocele, lipoma, spina bifida	Laminectomy, tumor excision	18/0.9	Improvement incontinence
2	42	M	11 mo	Hyperreflexia, DSD	Myelomeningocele, spinal dysraphism, dural ectasis	Nil	50/5.0	CISC, stationary
3	26	M	18 mo	Hyperreflexia, DSD	Lipomeningocele, spinal dysraphism, dural ectasis	Laminectomy, tumor excision	80/6.0	Hemodialysis
4	33	F	12 mo	Hyperreflexia, DSD	Lipomeningocele, dural ectasis, spina bifida, spinal dysraphism	Laminectomy, tumor excision	45/4.1	CISC, stationary
5	22	M	20 mo	Hyperreflexia, DSD	Lipomeningocele, dural ectasis, spina bifida, spinal dysraphism	Laminectomy, tumor excision	40/2.0	CISC, stationary

DSD: detrusor sphincteric dyssynergia, CISC: clean intermittent self-catheterizations
BUN: blood urea nitrogen, Cr: creatinine.

tinence, which was only found in 2 patients. This may be due to adults tending to present different signs and symptoms than children with tethered cord syndrome. The symptoms varied from irritable voiding symptoms, recurrent infection, incontinence, to abdominal fullness due to urine retention. Most of the urodynamic study findings showed hyperreflexia and detrusor-spincteric dyssynergia in 4 patients (80%). One patient had decreased sensation, decreased compliances and hyporeflexia shown on cystometrography. These are compatible with the results in the report by Giddens et al., who reported a 72% incidence of hyporeflexia in a 21-patient study.[10] However, detrusor-spincteric dyssynergia (DSD) and findings highly associated with detrusor hyperreflexia were not seen in their report. Because of the involvement may above the S2-4 nerve root in patients with tethered cord syndrome, DSD was common in our study.

Lumbosacral lipoma can be responsible for progressive neurological and neurovesical defects. Tethering of the spinal cord was noted in 4 of our patients. Surgery for untethering these structures may prevent late deterioration. Pierre-Kahn et al. routinely and systematically performed an operation even in the absence of neurological defects.[11] Because lipoma may also be associated with various other malformations, either intra- or extraspinal, untethering surgery may help decompress and spare functional neural tissue and prevent retethering of the spinal cord. In our patients, the operations included untethering, removal of tumors, duroplasty, laminectomy, and neurolysis. The result shows stability in cystometrography. The surgery may be not only for the tumors but also for associated anomalies. Prophylactic release of the spinal cord can prevent long-term disabilities associated with this anomaly.[1] Although it delayed the release of our patients, the effects reversed the loss of neurological function and thus prevented deterioration of renal function.

Although repair surgery had been performed in a patient with myelomeningocele in her childhood, secondary tethering of the spinal cord still bothered her. Surgical closure of the myelomeningocele was primarily carried out to prevent serious infections of the central nervous system. Characteristic presentations of leg weakness and urinary incontinence are symptoms of possible long-term complications of surgery for tethered cord syndrome. The outcome seems to become progressively worse if no treatment is done. Surgical treatment for untethering is suggested during this secondary deterioration after myelomeningocele repair.[9] Long-term follow up should be carried out by a team of neurosurgeons and urologists for those patients with a history of myelomeningocele repair. Since lower abdominal fullness was the major symptom in this study, it is stressed that young patients with voiding symptoms associated with lower abdominal fullness should be carefully examined. Early suggestions of tethered cord syndrome can be made during meticulous physical examination including digital anal tone examination and neurological survey.

Tethered cord syndrome is a progressively worsening disorder. It may cause irreversible changes of neurological and urological complications if the diagnosis is delayed. The number of patients surveyed was quite small in this study. However, the results suggest that early diagnosis of tethered cord syndrome is important. Since surgical intervention did not completely cure this condition during adulthood, it would be helpful to study ways to detect this disease early during childhood. Long-term follow up of patients and more patient studies are needed to explore the clinical course of tethered cord syndrome.

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REFERENCES

1. McLone DG, La Marca F: The tethered cord: diagnosis, significance, and management. Semin Pedatr Neurol 1997;4:192-208.
2. Yamada S, Zincke DE, Sanders D: Pathophysiology of "tethered cord syndrome". J Neurosurg 1981;54: 494-503.
3. Cornette L, Verpoorten C, Lagae L et al: Tethered cord syndrome in occult spinal dysraphism: timing and outcome of surgical release. Neurology 1998;50:1761-1765.
4. Soonawala N, Overweg-Plandsoen WC, Brouwer OF: Early clinical signs and symptoms in occult spinal dysraphism: a retrospective case study of 47 patients. Clin Neurol Neurosurg 1999;101:11-14.
5. Mandell J, Bauer SB, Hallett M et al: Occult spinal dysraphism: a rare but detectable cause of voiding dysfunction. Urol Clin N Am 1980;7:349-356.
6. Pang D, Wilberger JE: Tethered cord syndrome in adults. J Neurosurg 1982;57:32-47.
7. Gross AJ, Michael T, Dodeman F, Weigel K, Huland H: Urological findings in patients with neurosurgically treated spinal cord. J Urol 1993;149:1510-1511.
8. Johnston LB, Borzyskowski M: Bladder dysfunction and neurological disability at presentation in closed spina bifida. Arch Dis Child 1998;79:33-38.
9. Jeelani NO, Jaspan T, Punt JAG: Lesson of the week: tethered cord syndrome after myelomeningocele repair. BMJ 1999;318:516-517.
10. Giddens JL, Radomski SB, Hirshberg ED, Hassouna M, Fehlings M: Urodynamic findings in adults with the tethered cord syndrome. J Urol 1999;161:1249-1254.
11. Pierre-Kahn A, Zerah M, Renier D et al: Congenital lumbosacral lipomas. Childs Nerv Syst 1997;13:298-334.