

Clinical course and evaluation of meningocele lesion in adulthood: a case report

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ABSTRACT

The progressive course of spinal dysraphism has not yet been understood. Adult patients with spinal dysraphisms may give some evidences about the clinical progression of the malformations. In the present study we discussed a 48 years old patient with meningocele who has developed adult onset of impairment of sacral functions. The radiological, electrophysiological and urodynamic evaluations and pathophysiology of the adult onset of symptoms are also discussed. *Neuroanatomy; 2005; 4: 52–54.*

Key words [meningocele] [adulthood] [surgery] [spinal dysraphism] [neurosurgery]

Introduction

Surgical management of spinal dysraphism is an important area of neurosurgery. Spinal dysraphism defines incomplete fusion the neuronal arch, varying from the occult to more severe open neural tube defects. Meningocele, the simplest form of open neural tube defects, is characterized by cystic dilatation of meninges, which contain cerebrospinal fluid without any neuronal tissue. The natural course of meningocele has not entirely explored. Adult patients with spinal dysraphism may give some evidences about the clinical progression of malformations. Small series or few cases have been reported on dyplastic malformations in adults [1–5].

In this report, we present a 48 years-old patient with meningocele who has developed urinary and fecal incontinence in adulthood. The clinical course of this congenital malformation and the pathophysiology of the adult onset of symptoms are discussed.

Case Report

48 years old male ambulance driver presented with low back pain without radiating to legs for nearly 8 years and a 5 years history of progressive loss of sensation of bladder fullness, loss of ability to develop erection, decreased penile sensation and constipation. From puberty until age of 43, he had no difficulty with bladder function. For the past 5 years, he has realized increased effort in initiating urination and decreased force of urination. Over the past 5 years, he experienced decreased firmness and duration

of erection. His difficulty with bowel movements was one of chronic constipation, increasing over 5 years. He did not have any systemic disease.

During the inspection, a dorsal midline cystic lesion, measuring 4x3 cm, covered with skin, was noticed (Fig. 1). The lesion was tender to touch. The neurological examination of the patient revealed intact strength in all muscle groups and no sensory abnormality was evident. Straight leg raising test were negative bilaterally. Deep tendon reflexes were diminished and no fasciculation accompanied. There was decreased touch sensation over the perianal area and over the shaft of penis corresponding to S3-S5 spinal cord segment. The patient had a very weak anal sphincter although it was capable of voluntary contraction. The anal wink reflex and the bulbocavernous reflex were markedly decreased.

Urological evaluation with abdominal and urogenital ultrasonography showed no abnormality. The urodynamic evaluation with pressure flow studies and sphincter electromyographic studies demonstrated a flaccid neurogenic bladder with evidence of internal sphincter denervation.

The blood chemistry was normal including blood glucose and prostate specific antigen levels.

X-ray images of lumbosacral region demonstrated L5 spina bifida. Sagittal T1-weighted magnetic resonance (MR) imaging of lumbosacral region revealed a myelocele sac originated from L5 spina bifida. It was also observed



Figure 1. Patient with lumbosacral meningocele.

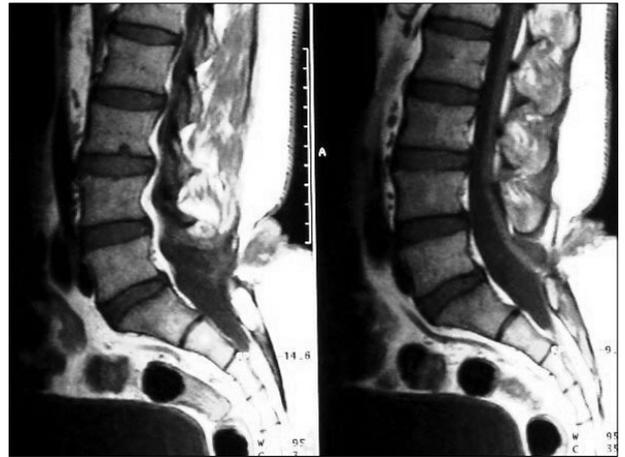


Figure 2. Sagittal T1-weighted MR images demonstrated myelocoele sac originated from L5 spina bifida. Notice that conus medullaris reached L5 level and extension of the filum terminale terminated within the meningocele.



Figure 3. Sagittal T2-weighted MR images demonstrated tethering of spinal cord with no hydromyelia nor myelomalacia.

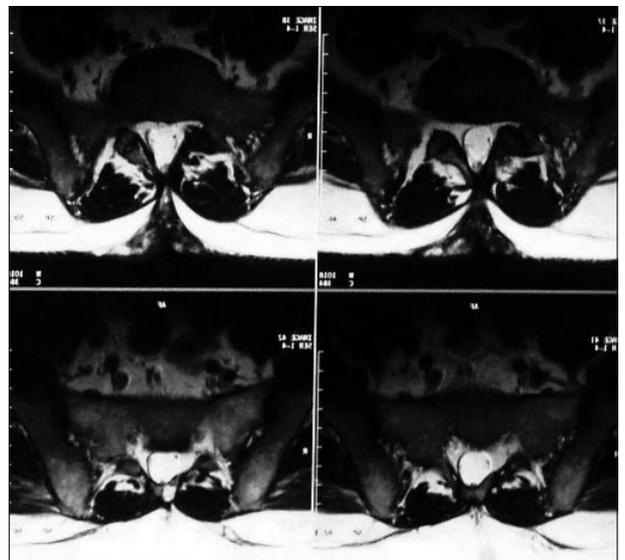


Figure 4. Axial T2-weighted MR images demonstrated associated lipoma and no neurological tissue involved in the meningocele cavity.

that conus medullaris reached L5 level and extension of the filum terminale terminated within the meningocele, causing tethering of the spinal cord (Fig. 2). Sagittal T2-weighted MR images demonstrated tethering of spinal cord with no hydromyelia nor myelomalacia (Fig. 3). With T1-weighted axial MR images an associated lipoma was detected and no neurological tissue involved in meningocele cavity (Fig. 4). Somatosensory evoked potential (SSEP) was in normal range.

Since the patient had progressive neurological symptoms, surgical treatment was suggested. The patient did not accept the operation and he has been under control of both neurosurgery and urology departments. He has been followed up with intermittent urinary catheterization.

Discussion

Any meningocele patient has deteriorating clinical symptoms such as progressive orthopedic deformities, lower extremity weakness or urinary and fecal incontinence associated with low back pain, should be considered as tethered cord [4]. The degree of traction of the conus is thought to determine the age of onset of symptoms [3–4]. In cases of marked tethering and severe stretching of the conus, neurological symptoms appear in infancy or early childhood [6]. Minimal tethering may remain subclinical until adulthood [3]. 29% of patients with symptomatic tethered cord, have been found to be older than 35 years [7]. The mechanism of late onset of symptoms has not yet been well understood, however the mechanism is explained by the cumulative effect of repeated cord traction by various postures [3–5, 8]. Yamada et al. stated that neurological dysfunction in patients with tethered cord correlates with mitochondrial anoxia within the conus [9]. The narrowing of the spinal canal by lumbar spinal stenosis and disc prolapsus and resultant increased tension in spinal cord may also precipitate symptoms [7–9]. It has been suggested that the longitudinal stress within the spinal cord may be transmitted more distally along the lateral columns as they are fixed by dentate ligaments [3–5, 8]. Direct trauma to the lumbosacral region may precipitate the symptoms causing deformation of the marginally functioning neuronal elements within the stretched cord [3–5].

In our patient, conus situated at L5 and extension of the filum terminale terminated in meningocele at the level of S1. The precipitating factors in the appearance of

symptoms that are the momentary stretching of the tight conus with sudden flexion of the neck and hip, may be related to his occupation since driving often predispose to these conditions.

MR is a useful technique for evaluation of patients with spinal dysraphisms. Direct X-ray and computed tomography may give information about the associated bone defects, while MR is a superior diagnostic tool for verifying the cystic lesion in the sagittal plane, its relation with the spinal cord and associated spinal cord anomalies [7]. In the present case, direct X-ray shown L5 spina bifida and MRI studies demonstrated low lying conus associated with lumbosacral meningocele.

SSEP was reported as more sensitive than clinical testing for detection of neurological deficits in patients with spinal cord lesion [10]. Abnormal SSEP is a clear indication of spinal pathology. However, not all spinal pathologies are associated with abnormalities in SSEP [4, 10]. In the reported patient, SSEP was found to be within normal range.

Pre-operative urodynamic investigation is strongly recommended, especially if the patient seems continent [2]. In our patient, urodynamic evaluation demonstrated non-functioning internal sphincter and detrusor hyporeflexia indicating flaccid neurogenic bladder. It was demonstrated that tethering of the conus usually causes mixed abnormalities of parasympathetic, sympathetic and somatic pathways [2]. Sympathetic innervation was often impaired first, resulting in non-functioning internal urethral sphincter, which characteristically causes post-voiding dripping and stress incontinence [2].

Although conflictions are present about the surgical indication of asymptomatic patients, it has been suggested that, each adult or child should be operated as soon as symptoms appeared or progressed because in the majority of patients only stabilization of the disease is achieved [1, 4–5]. Because of the progressive neurological symptoms, the patient was suggested to surgical treatment. Since the patient did not accept the operation, we could not comment on the results of surgical intervention. This patient may give some evidences about the relation between the untreated intradural abnormalities and progressive neurological deficits. Further investigations are necessary on the natural history of spinal dysraphism.

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