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Huey-Wen Liang

Tyng-Guey Wang

Yen-Ho Wang

Tiffany T. F. Shih

Jin-Shin Lai

See next page for additional authors

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Cervical myelopathy secondary to cervical disc protrusion in patient with dystonic cerebral palsy--A casereport

Authors

Huey-Wen Liang, Tyng-Guey Wang, Yen-Ho Wang, Tiffany T. F. Shih, Jin-Shin Lai, and I-Nan Lien

Cervical Myelopathy Secondary to Cervical Disc Protrusion in Patient with Dystonic Cerebral Palsy --A Case Report

Huey-Wen Liang, Tyng-Guey Wang, Yen-Ho Wang,

Tiffany T. F. Shih*, Jin-Shin Lai, I-Nan Lien

Cervical myelopathy had been noted in patients with involuntary neck movement, especially in individuals with athetoid-dystonic cerebral palsy(CP). The excessive and repeated neck motions were considered to be responsible for the premature spondylosis, which might be the cause of myelopathy. The exact pathophysiology and managements were still controversial. Here we present a rare case who was a male patient with long-lasting dystonic neck movement developing cervical myelopathy due to protrusion of disc in his relatively young age. Magnetic resonance imaging (MRI) showed the protrusion of cervical disc at the level of C3-4, which was correlated well with his clinical symptoms. Anterior diskectomy and bony fusion were done for decompression. Halo-vest immobilization was applied to prevent the instability coming from dystonic neck movement after surgery. There was partial neurologic recovery after surgery. It is a rare clinical condition and may be easily overlooked. Therefore we recommended that MRI should be done as early as possible for cases with similar presentations, in order to establish an early correct diagnosis and early intervention.

Key words : cervical myelopathy, intervertebral disc, cerebral palsy

INTRODUCTION

Cerebral palsy(CP), being defined as " non-progressive motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development" [1], is a frequent cause of disabilities requiring rehabilitation. In patients with spastic or athetoid-dystonic cerebral palsy, progressive neurological deterioration in their adulthood had been reported[2-9]. Cervical radiculomyelopathy secondary to premature spondylosis, disc protrusion, ligament hypertrophy, instability and malposture of the cervical spine was considered as possible cause[7,10]. Excessive neck motion, which is common in these two groups of patients, may be responsible for these pathologic changes. The diagnosis of cervical myelopathy and defining its etiology becomes complicated in this group of patients

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Departments of Physical Medicine and Rehabilitation and of Image Medicine*
National Taiwan University Hospital, Taipei, Taiwan, R.O.C.
Correspondence and reprint requests to Dr. Huey-Wen Liang, Department of Physical Medicine and Rehabilitation, National Taiwan University Hospital, No.7 Chung-Shan South Road, Taipei, Taiwan, R.O.C.
Tel.: 02-3970800 ext 7182

because of their pre-existing physical and mental disabilities. It is still controversial if certain management is beneficial for CP patients with cervical radiculomyelopathy.

To our knowledge, all cases being reported were having marked spondylosis as the major roentgenographic findings and disc protrusion was rarely mentioned. We present a dystonic CP patient with cervical myelopathy diagnosed by magnetic resonance imaging (MRI), who had a protruding intervertebral disc at C3-4 level without definite spondylotic change. Problems being encountered during the post-operative rehabilitation period will be discussed.

CASE REPORT

A 24 year-old male was a case of cerebral palsy diagnosed by the history of delayed development milestone, abnormal posture and muscle tone. There was also accompanied by repeated dystonic neck movement. Impaired mentality, slight dysarthria, and clumsy movement had been noted since his childhood. He was ambulatory and independent in self-care. His condition had been stable in the past years.

One year before his admission, he suffered from neck pain, followed with slowly progressive weakness and numbness over four limbs in six months. Easy falling became a problem and one episode of urinary retention occurred two months before his admission. At the same time, he had difficulties in ambulation .

Neurologic examination on admission showed a dysarthric young man with a hypertrophic sternocleidomastoid muscle and repeatedly dystonic neck movement. Impaired muscle strength was recorded over all four limbs, which were 4/5 over bilateral upper limbs and right leg, and 3/5 over left leg. Muscle atrophy and fasciculation were noted over bilateral shoulder girdles and biceps muscles. There was mild hypesthesia to pinprick and light touch below the distribution of C4 dermatome. Deep tendon reflexes were hyperactive throughout and Babinski's sign was present bilaterally. Transient ankle clonus could be elicited.

Radiography of cervical spine showed an abnormally straight alignment. (Fig. 1)The osteophyte formation was not prominent. The intervertebral disc spaces were relatively preserved. No bony fracture was noted. MRI examination was done, which showed a protruding intervertebral disc at the level of C3-4 with compression to spinal cord.(Fig. 2) From the T1 and T2 axial section through C3-4, the dural sac was flat in A-P diameter. the spinal cord was compressed by the protruding disc, especially over right side. The signal intensity of intervertebral disc showed no evidence of degeneration.

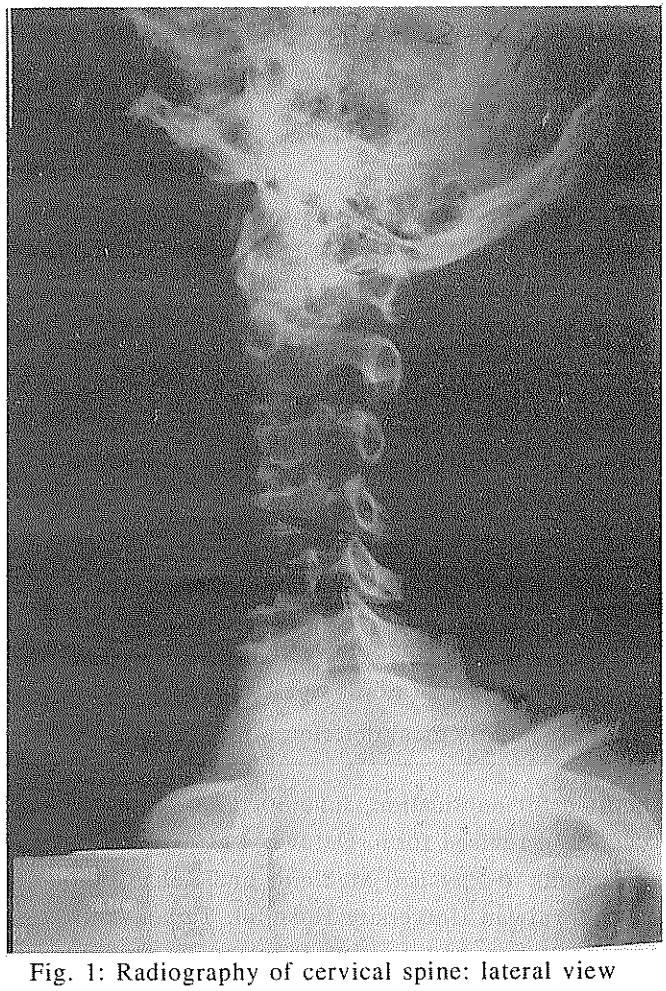


Fig. 1: Radiography of cervical spine: lateral view in neutral position

Anterior diskectomy of C3-4 and bony fusion were performed with halo-vest immobilization. Following surgery, his muscle strength generally gained improvement in about one grade by manual muscle test. The spasticity was still prominent. He received a two-month rehabilitation program, which included progressive resistance exercises to strengthen his muscle power, ADL training to restore his ability of self-care, ambulation training, and other functional training programs. When he was able to stand, he still had poor performance on bed mobility because of the interference of halo-apparatus. Ambulation was possible only with bilateral long leg braces and walker when he was discharged three months later. He still needed some assistance in doing daily activities.

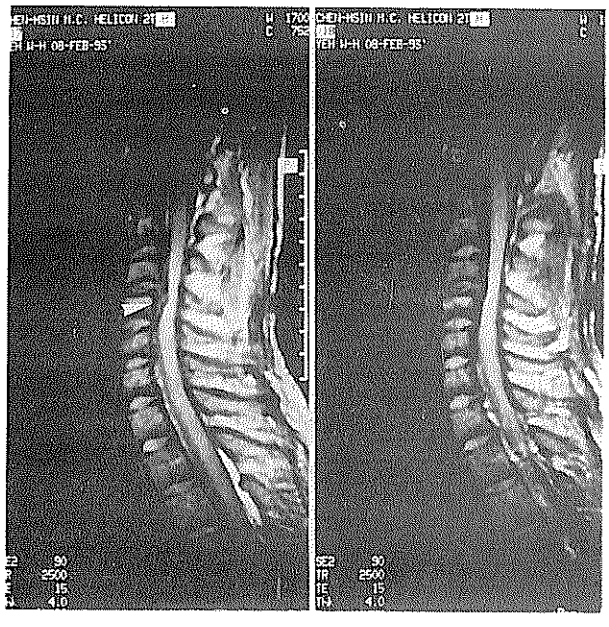
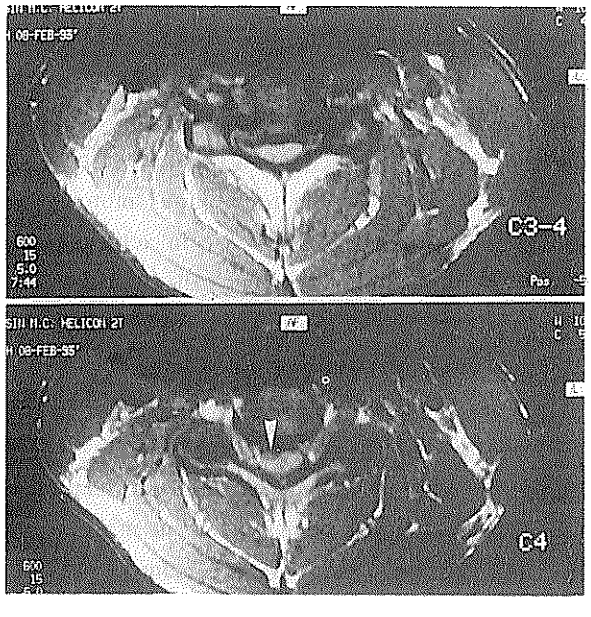


Fig. 2: MRI

A)axial view: disc protrusion at the level of C3-4 (arrow).



B)sagittal view: a flat dural sac at the level of C3-4 with compression at the right aspect (arrow).

DISCUSSION

Since Freiman and Luwisch[11] presented two cases of dystonia with muscular atrophy, termed "dystonic atrophy" in 1956, several reports had been made in attempt to clarify the relationship of cervical myelopathy and long-lasting abnormal neck motion, especially in patients with athetoid-dystonic and spastic cerebral palsy.[2,3,8] Despite of limited experience in the past decade, a specific pattern of clinical features had been presented. The age of onset in CP with cervical myelopathy is younger than that of non-dystonic population.[4,5] The spondylotic change at the upper and middle cervical disc level, including C3-4, C4-5 and/or C5-6, instead of C5-6 and C6-7, are the most common sites of lesions, which is different from cases with cervical spondylosis due to aging.[9,13] The diagnosis was made by myelography or computed tomography but was usually delayed while patients had lost their previously acquired functional skills, including ambulation[10]. Similarly, our case also had long-lasting history of dystonic neck motion and the symptoms of myelopathy occurred at his relatively young age. He was diagnosed cervical myelopathy one year after the onset of weakness in four limbs and neck pain.

In most of the previous reports, cervical spondylosis was believed to be the dominant cause of cervical radiculomyelopathy in cerebral palsy patients , and the constant neck movement played a major role in its occurrence.[5,7,8]. The other associated

roentgenographic findings in these patients reported previously before including spinal canal stenosis, malposture of cervical spine, disc protrusion, and hypertrophic ligament. However, some researches considered that prolonged athetoid movement of neck was not responsible for the premature spondylosis[2], or the severity of spondylosis had no proportional relationship with myelopathy[3]. Ebara et al[12] analyzed the static and dynamic X-ray of cervical spine in fifty-seven patients with athetoid type CP and found increased frequency of instability and malposture at C3-4 and C4-5 levels of the cervical spine.

Cervical disc protrusion is a rare condition in a patient without history of trauma. Odom et al [13] classified cervical disc lesions into four categories : unilateral soft disk protrusion with nerve root compression, foraminal spur (or hard disc) with nerve root compression, medial soft disk protrusion with spinal cord compression and cervical spondylosis with spinal cord compression. Degeneration was an important contributing factor. In his study, soft disk protrusion occurred in a much less frequency (14 in 246). In patients with involuntary neck movement complicated with radiculomyelopathy, disk protrusion in the myelogram was mentioned but the significance had not been clarified.[8,10] Nevertheless, it had been proved that prolonged heavy activity can make the posterior annulus vulnerable, and consequently a complete fissure or prolapse in cadaveric lumbar intervertebral discs[14]. No similar study had been reported on cervical spine. Dystonic motion is an involuntary movement with the combination of abnormally excessive flexion, rotation and extension. Repeated flexion may put large stress on intervertebral disc and presents as loss of normal physiologic curve. The result of long-lasting malalignment may elicit disc protrusion. In our patient, he had severe dystonic motion for more than twenty years. He had no history of acute or occupational trauma. It is rational to assume that the excessive neck movement may be related to the mechanism of disk protrusion in such a young person.

Delayed in the diagnosis of cervical myelopathy was common in these patients with underlying impairment of motor function and mentality. In addition to a classic neurologic examination, regular assessment of mentality and functional status was advised by Reese et al [10]. The alertness of both caregiver and physician is important in early detection of any deterioration of motor function. MRI has been used widely in the evaluation of spinal lesion in recent days. It is non-invasive with the advantages of good resolution, better visualization of soft tissue and intraspinal lesions, and unnecessity of usage of contrast medium use. In patients with dystonia, sedation is indicated to prevent involuntary movement during examination.

The results of surgery were widely variable with different procedures. Levine et al [3] performed extensive laminectomy from C2 to T1 in one patient but no neurological recovery was observed. Neither did the two patients reported by Kidron et al [8]. Decompressive cervical spine surgeries were ineffective in their reported cases. Delayed operation was believed to be responsible for the poor results. All four cases reported by Hirose and Kadoya [4] regained the ability to ambulation with or without assistant device and they suggested early anterior decompression with interbody fusion to be the treatment of choice. In Fuji's report[7], six in ten of his patients had good to excellent post-operative results (improving to previous motor function or with a minimum decrease in activity). Our patient had some recovery but still unable to resume prior motor function post-operatively. The unsatisfactory result may be possibly due to irreversible neural damage and delayed surgery.

Post-operative instability is of major concern, especially in patients receiving multiple laminectomies. The continuous neck movement make immobilization more difficult. Management of post-operative instability including internal fixation, external immobilization and pharmacologic therapy to reduce involuntary motion had all been tried with

unsatisfactory results. Wang and Chen [6] used Trihexyphenidyl HCl (Artane) for the dystonic movement but in vain. Hirose et al [4] performed microsurgical anterior operation in four patients and three of them had anterior bony fusion. He used a neck collar for immobilization constantly for one month and for another one month in daytime. He claimed that a good bony fusion was achieved, except pseudarthrosis in one patient. Nishihara et al[5] considered that laminectomy alone was contraindicated in patients with potential instability after operation, and anterior fusion combined with halo-cast could achieve good bony fusion. In order to shorten the duration of external immobilization, Fuji et al[7] recommended interspinous wiring. In our cases, anterior bony fusion and halo-vest immobilization for three months were able to achieve the stability. Halo-apparatus can provide good immobilization of the cervical spine, especially in the high cervical region, but it does interfere with our rehabilitation programs. Skull screw loosening, the common complication of halo-vest, occurred in our patient during hospitalization period.

In conclusion, cervical myelopathy is a relatively rare condition reported in CP patients. Premature spondylosis is not the only pathologic changes but possibility of cervical disc protrusion should be kept in mind if there is no significant bony lesions shown in plain X-ray. MRI is a superior diagnostic tool in patients with cervical myelopathy. It deserves our attention because early diagnosis and treatment can minimize the permanent neurologic sequelae.

REFERENCE

1. Mutch L, Alberman E, Hagberg B, et al.: Cerebral palsy epidemiology: where are we now and where are we going? Dev Med Child Neurol 1992;34:547-51.
2. Anderson WW , Wise BL, Itabashi HH, et al.: Cervical spondylosis in patients with athetosis. Neurology (Minneap.) 1962;12: 410-2.
3 .Levine RA, Rosenbarum AE, Waltz JM, et al.: Cervical spondylosis and dyskinesias. Neurology 1970;20:1194-9.
4. Hirose G and Kadoya S: Cervical spondylotic radiculo-myelopathy in patients with athetoid-dystonic cerebral palsy: clinical evaluation and surgical treatment. J Neurol Neurosur Psy 1984;47, 775-780.
5. Nishihara N, Tanabe G, Nakahara S, et al.: Surgical treatment of cervical spondylotic myelopathy complicating athetoid cerebral palsy. J Bone Joint Sur 1984;66B: 504-8.
6. Wang PY, Chen RC: Cervical spondylotic radiculomyelopathy caused by athetoid-dystonic cerebral palsy, clinical evaluation of 2 cases. J Formo Med Assoc 1985;84: 986-94.
7. Fuji, T, Ronenobu K, Fujiwara K, et al.: Cervical radiculopathy or myelopathy secondary to athetoid cerebral palsy. J Bone Joint Surg 1987;69A(6): 815-21.
8 .Kidron D, Israel S, Melamed E: Late-onset progressive radiculomyelopathy in patients with cervical athetoid-dystonic cerebral palsy. Eur Neuro 1987; 27: 164-6 .
9. El-Mallakh RS, Rao K, Barwick M: Cervical myelopathy secondary to movement disorders: case report. Neurosurgery 1989;24: 902-5.
10.Reese M E, Msall M E, Owen S et al.: Acquired cervical spine impairment in young adults with cerebral palsy, Dev Med Child Neurol ,1991; 33: 153- 66.
11.Freiman IS, Luwisch J: Dystonic amyotrophy, dystonia and muscular atrophy. Neurology (Minneap) 1956;6: 108-14.
12.Ebara S, Harada T, Yamazaki Y, et al.: Unstable cervical spine in athetoid cerebral palsy. Spine 1989;14(11) 1154-9.
13.Odom GL, Finney W, Woodhall B, et al.: Cervical disk lesions. JAMA1958; 166(1) : 23- 8.
14.Adams MA, Hutton WC: The effect of fatigue on the lumbar intervertebral disc. J Bone Joint Surgery. 1983; 65B(2) 199-203.

張力異常型腦性麻痺患者伴隨頸部不正常動作之頸髓病變一病例報告

梁蕙雯 王亭貴 王顏和 施庭芳* 賴金鑫 連倚南

頸部有不自主運動的患者，發生頸椎神經根或脊髓壓迫病變過去僅有少數報告，主要發生在舞蹈型-張力異常型的腦性麻痺患者。一般認為，長期過度的頸部動作易使頸椎提早退化，是造成神經壓迫的主要原因，但真正的致病機轉及其治療原則，則尚無定論。這裡，我們報告例罕見的頸部椎間盤凸出導致頸髓病變的病例。該病人是一位24歲的腦性麻痺男性患者，自幼即有頸部的張力異常動作，但日常生活可獨立。其臨床表徵包括頸部疼痛及漸進性的四肢無力，經核磁共振造影檢查發現在頸椎第三、四節之間有椎間盤凸出並壓迫頸髓的情形。由於放射線檢查並無頸椎退化現象，故我們認為反覆性頸部不自主運動可能造成其椎間盤凸出，而致頸髓病變，不一定源自傳統所認定之脊椎退化。患者接受頸椎間板切除術以解除脊髓壓迫症狀，手術後，有部份的功能復原。經過復健治療，雖可以穿戴兩側長腿支架，持助行器行走，但日常生活仍需協助。由於這種病例在臨床上並不多見，極易誤診而延誤治療導致病情惡化，因此建議臨床醫師如逢類似情形之病例應及早作核磁共振造影檢查以利早期作適當處置。

國立臺灣大學附設醫院 復健部，影像醫學部*
抽印本索取地址： 梁蕙雯， 國立臺灣大學附設醫院 復健部，北市中山南路七號
電話：02-3970800 轉7182