Transverse Myelitis: A Case Report

Transvers Myelit: Bir Olgu Sunumu

Cemile Sevgi Polat, Asuman Doğan, Belma Füsun Köseoğlu, Mahir Kemal Taşkın

Ankara Physical Therapy and Rehabilitation Education and Research Hospital, Department of Physical Medicine and Rehabilitation, Ankara, Turkey

ABSTRACT

Transverse myelitis (TM) is one of the etiologic factors of non-travmatic spinal cord injury and characterized by focal inflammation within the spinal cord. One of the most important differential diagnosis is Guillain-Barre syndrome (GBS) with its variants. Our patient was referred from a children's hospital with the diagnosis of GBS , but detailed neurological examination made us have doubt on the diagnosis and we considered TM as diagnosis. Correct diagnosis is important for treatment, rehabilitation and prognosis. Rehabilitation is an important part of TM treatment and the functional gains of these individuals can be increased by rehabilitation efforts.

Keywords: Transverse myelitis, misdiagnose, Guillain-Barre Syndrome, rehabilitation

ÖZET

Transvers myelit (TM) non-travmatik spinal kord hasarı nedenleri arasındadır ve spinal kordda lokal inflamasyonla karakterizedir. En önemli ayırıcı tanılardan biri de Guillain-Barre sendromu (GBS) ve varyantlarıdır. Bizim hastamız çocuk hastanesinden GBS tanısı sevk edildi, fakat detaylı nörolojik muayenesi tanıda şüphe duymamıza sebep oldu ve tanı olarak TM düşünüldü. Doğru tanı tedavi, rehabilitasyon ve prognoz açısından önemlidir. TM'de rehabilitasyon tedavinin önemli bir parçasıdır ve olguların fonksiyonel kazanımlarını arttırmaktadır.

Anahtar sözcükler: Transvers myelit, yanlış tanı , Guillain-Barre Sendromu , rehabilitasyon

Corresponding Author
Yazışma Adresi

Cemile Sevgi Polat

Ankara Physical Therapy and Rehabilitation Education and Research Hospital, Department of Physical Medicine and Rehabilitation, Ankara, Turkey

E-mail: drsevgipolat@hotmail.com

Received/Geliş Tarihi: 22.04.2015
Accepted/Kabul Tarihi: 01.12.2015

Introduction

Transvers myelitis is an acute inflammatory disease of the spinal cord, characterized by acute onset of motor, sensory, and autonomic dysfunction. The causes of transverse myelitis are diverse; etiologies implicated include demyelinating conditions, colllagen vascular disease, and parainfectious causes, however, despite extensive diagnostic work-up many cases are considered idiopathic (1, 2). TM affects individuals of all ages with bimodal peaks between the ages of 10 and 19 years and 30 and 39 years (3). There is no sex or familial predisposition to TM (4). Approximately 20% of cases of transverse myelitis occur in children (1). A spinal Magnetic Resonance Imaging (MRI) and lumbar puncture often show evidence of acute inflammation (5). The most important differential diagnosis are Multiple Sclerosis (MS) and Guillain-Barre Syndrome (GBS) with its variants (6). The most common long-term complications of transverse myelitis are urinary, motor, or sensory dysfunction (1). The long-term management should focus on neurorehabilitation and a multidisciplinary approach aimed at managing the various complications of spinal cord damage. Here, we report the case of a 14-year-old patient with TM who was misdiagnosed as guillain-barre syndrome and also discussed in view of the literature.

Case Report

14 -year-old male patient was referred from The Children's Hospital with the diagnosis of GBS to Ankara Physical Medicine and Rehabilitation Training and Research Hospital, for coming into a rehabilitation program. The patient was interviewed and underwent

a complete medical assessment that included medical history, neurological examination, and routine laboratory measurements. Twenty-eight days before admission to the our hospital, he had sudden onset of numbness and weakness in the both arms and the legs, then severe respiratory distress occured within a few hours. Therefore, he was monitored in the intensive care unit (ICU) of the children's hospital and had cardiac arrest 3 times during this period. He had a gradient of motor and sensory loss involving the lower extremities greater than the upper extremities. There was no significant finding in the cerebro-spinal fluid analysis, there were normal cranial and spinal cord imaging by MRI and there were no signs of demyelinisation, but in the amplitude decrease, and axonal injury findings detected at the needle EMG study. Result of the first examination in rehabilitation center was revealed that he was bedridden, conscious, well orientated, with 36.5 fever and 120/80 blood pressure. The cranial nerves were intact, had no sitting balance and was partially depended in-bed activities such as righting and rotation. There was no sensation and sphincter control in the bladder and there was sensation but no sphincter control in the rectum. Muscle strength was 4/5 at right upper limb, 4/5 at proximal of left upper limb and 3/5 at the distal part, 2/5 at proximal of the right lower limb, 4/5 at the distal part, 1/5 at the proximal of the left lower limb, 2/5 at the distal part. Right upper and lower limb spasticity was recorded as score of 1, and left lower limb spasticity as score of 2, according to the Ashworth Scale. There was loss of sensation at the level of T4. Deep tendon reflexes (DTRs) were hyperactive and pathological reflexes such as babinski, clonus and hoffman tests were positive bilaterally. Although the patient's history such as muscle weakness starting from the lower limbs and ascending to the arms, face and respiratory muscles that makes us consider GBS diagnosis, existence of the upper motor neuron involvement findings made us have doubt on the diagnosis. Therefore, cranial and spinal MRI were scheduled again. Cranial MRI was reported as normal but there was a T2A hyperintensity at the cervical spinal cord white matter areas, especially more prominent at the level of C3-C4, on the cervical MRI. EMG study was performed again in our hospital. Nerve conduction studies were normal, and there was no sign of demyelinisation. With the current clinical and radiological findings, transvers myelitis was considered as diagnosis. Considering the current condition and problems of the patient, rehabilitation program was scheduled. Mild restrictive ventilation defect and decreased endurance of the respiratory muscles were found by pulmonary function test. Pulmonary rehabilitation program was started according to this results. The patient had detrussor overactivity, and difficulty on storing and emptying bladder according to the urodynamic analysis. He was considered appropiate for the permanent urinary catheter and started on oxybutinin 10 mg for a day and urodynamic control was planned 3 months later. Postural and active-assistive trunk exercises, sitting balance training were given to the patient who had difficulty at maintaining sitting balance at the acute period. Dorsolumbar corset with steel underwire were also applied to support sitting balance. Passive, active assistive and active ROM exercises were performed for all limbs according to the result of the muscle testing. Progressive resistive exercise therapy was applied to the muscles at and above 3/5 muscle strength. Static functional electrical stimulation was given to the left upper limb and left lower limb muscles at and below 2/5 muscle strength. Volar wrist-hand stabilizer (resting splint) for the left hand and plastic knee ankle foot orthoses for the both lower limbs were given to control the aligment of the joints of the limbs. Hotpack and strecthing exercises were performed to the left gastrocnemius muscle group because of going to the equinus deformity. Transfer exercises from bed to the wheelchair, and also verticalisation and walking exercises in the parallel bars were administred to the patient after he had trunk and sitting control. Occupational therapy was also added. Patient's muscle strength improved with the rehabilitation program. Muscle strength levels increased to 5/5 at right upper limb and 5/5 left upper limb at the proximal part, and 4/5 at the distal part. Muscle strength was also determined as improved to 4/5 at right lower limb proximal and 5/5 at the distal part, and 4/5 at left lower limb proximal and 3/5 at the distal part. Short walking orthoses were prescribed to assist dorsiflexor weakness of the ankle. At the end of the rehabilitation program, patient was independently mobile with one cane and short walking orthesis.

Discussion

The onset of TM is characterized by acute or subacute development of neurologic signs and symptoms consistent with motor, sensory and/or autonomic dysfunction. Motor symptoms include a rapidly progressing paraparesis that can involve the upper extremities, with initial flaccidity followed by spasticity (7,8). Most patients have a sensory level. The most common sensory level in adults is the mid-thoracic region, though children may have a higher frequency of cervical spinal cord involvement and a cervical sensory level (9). Typical sensory symptoms are pain, dysesthesia, and paresthesia, although paresthesia are uncommon in children. Autonomic symptoms include increased urinary urgency, bowel or bladder incontinence, difficulty or inability to void, incomplete evacuation or bowel constipation, and sexual dysfunction (7). According to our patient's anamnesis, his symptoms developed acutely, weakness started with the legs and rapidly involved the arms. Spasticity was obtained when

we examined the patient. Spinal cord involvement was in the cervical region but sensory level was at the thoracic region, bladder dysfunction was also present.

We suggested that our patient was misdiagnosed as GBS in the children hospital. TM is often misdiagnosed as GBS, because both conditions may present with rapidly progressive sensory and motor loss involving principally the lower extremities. Therefore, GBS is mentioned in the differantional diagnosis of TM in the literatures (1,6,10-12). Several clinical features may be used to discriminate patients with GBS from those with TM. Patients with GBS often have both upper and lower extremity involvement, though the lower extremity involvement is usually more severe. Patients with TM may have only lower extremity involvement if the myelopathy is thoracic, or equivalent upper and lower extremity involvement if the myelopathy is cervical. A sensory level is often definable in patients with acute TM but is never present in GBS. Pathologically brisk deep tendon reflexes are supportive of TM. However, patients with fulminant cases of TM that includes significant destruction of spinal cord gray matter may present with hypotonia and have decreased or absent DTRs. Autonomic involvement differs between patients with GBS and TM. Patients with TM are more likely to have urinary or bowel urgency or retention, while those with GBS are more likely to have cardiovascular instability. Our patients had both urinary and bowel dysfunction, and cardiovascular instability during the acute period. An MRI of the spinal cord may show an area of inflammation in TM but not in GBS. Although cerebral spinal fluid findings in TM are not consistent and an elevated cell count may be absent, there is usually a moderate lymphocytic pleocytosis and elevated protein level. This is in contrast to the albumino-cytologic dissociation of the cerebro-spinal fluid seen in GBS. Electrodiagnostic studies may show conduction block or slowed conduction in peripheral nerves in GBS and are usually normal in TM (6,10,11).

Our case was hospitalized with the diagnosis of GBS. A detailed clinical history and physical examination are critically important. Although the case made us think GBS clinically because of patient's history (rapid progressive sensory and motor loss especially in legs and had cardiovascular instability in childrens hospital), presence of hyperactive DTRs, spasticity, ongoing urinary bladder symptoms, and contradiction of EMG findings and CSF analysis results caused us reconsider the diagnosis. Respiratory distress and cardiovascular instability in addition to the rapid progressive sensory and motor loss of the limbs at acute stage, and also normal spinal cord imaging by MRI were considered as patient had GBS in the children's hospital. Magnetic resonance imaging is the modality of choice for TM diagnosis; it shows signal abnormalities, usually T2 hyperintensity, focal or extensive, gadolinium enhancement and sometimes cord swelling. Despite its high sensitivity, about 40% of acute transverse myelopathies remain undemonstrated (13). At the acute stage in the children's hospital, because of the flask type muscle weakness and no DTRs (as pyramidal signs expected came up at the end of second week), cardiovascular instability , and respiratory distress may caused missing the diagnosis as described in the literatures (9,11, 14). Rehabilitation is an important part of the treatment in TM and increases the potential of the patient's ambulation. Many patients withTM will require rehabilitative care to prevent secondary complications of immobility and to improve their functional skills. TM is similar to spinal cord injury by the the terms of complications, clinical features, healing, rehabilitation program scheduling and the observation. But the observation of the TM cases is of a particular importance in the terms of recurrence and transformation to the MS (15). The majority of patients with TM experience monophasic disease. Recurrence has been reported in approximately 25 to 33 percent of patients with idiopathic TM (16,17). Patients presenting with TM have a generally cited risk of multiple sclerosis of only 5 to 10 percent (18,19). A small percentage of children diagnosed with TM later are diagnosed with other demyelinating diseases, especially neuromyelitis optica, or multiple sclerosis. The most common long-term complications of TM are urinary, motor, or sensory dysfunction (1). Regular evaluations by medical specialists for urodynamic studies are recommended to prevent potentially serious complications (11).

Rehabilitation is an important part of TM treatment. The therapeutic goal is to improve the function of the patient in performing activities of daily living. Correct diagnosis is important for appropriate rehabilitation. Therefore a physiatrist must have a well knowledge of the neurological examination as well as a neurologist. The main purpose of the discussing this case is getting the correct diagnosis of the case and making the patient independent at the daily living activities at a short time without complication thanks to a comprehensive rehabilitation program .

References

1. Wolf VL, Lupo PJ, Lotze TE, Pediatric acute transverse myelitis overview and differential diagnosis. J Child Neurol. 2012;27(11): 1426-36
2. Goh C, Desmond PM, Phal PM. MRI in transverse myelitis. J Magn Reson Imaging. 2014; 40(6):1267-79.
3. Proposed diagnostic criteria and nosology of acute transverse myelitis. Transverse Myelitis Consortium Working Group. Neurology 2002;59:499-505

4. Scott TF , Frohman EM, De Seze J, Gronseth GS, Weinshenker BG; Therapeutics and Technology Assessment Subcommittee of American Academy of Neurology Evidence based guideline: clinical evaluation and treatment of transverse myelitis: report of the Therapeuticsand Technology Assessment Subcommittee of the American Academy of Neurology. Neurology. 2011 ;77(24):2128-34
5. Beh SC, Greenberg BM, Frohman T, Frohman EM. Transverse myelitis. Neurol Clin 2013;31:79-138
6. Knebusch M, Strassburg HM, Reiners K. Acute transverse myelitis in childhood: nine cases and review of the literature. Dev Med Child Neurol 1998;40(9):631-9.
7. Krishnan C, Kaplin Al, Pardo CA, Kerr DA, Keswani SC.Demyelinating disorders: update on transverse myelitis. Curr Neurol Neurosci Rep 2006;6(3):236-43
8. Kaplin Al, Krishnan C, Deshpande DM, Pardo CA, Kerr DA.Diagnosis and management of acute myelopathies. Neurologist 2005; 11(1):2-18
9. Pidcock FS, Krishnan C, Crawford TO, Salorio CF, Trovato M, Kerr DA. Acute transverse myelitis in childhood: center-based analysis of 47 children Neurology. 2007 May 1;68(18):1474-80.
10. Chitra Krishnan, Benjamin Greenberg, Douglas Kerr. Transverse myelitis.2014 UpToDate.
11. Krishnan C, Kaplin Al, Deshpande DM, Pardo CA, Kerr DA. Transvers Myelitis: Pathogenesis, Diagnosis and Treatment. Front Biosci. 2004;9:1483-99.
12. Pope JV, Edlow J A. Avoiding Misdiagnosis in Patients with Neurological Emergencies Emerg Med Int. 2012;2012:949275.
13. Scotti G, Gerevini S. Diagnosis and differential diagnosis of acute transverse myelopathy. The role of neuroradiological investigations and review of the literature. Neurol Sci. 2001;22 Suppl2:S69-73
14. Canpolat M, Gümüş H , Per H, Poyrazoğlu H.G, Yıkılmaz A, Çoşkun A, Kumandaş S, Çocukluk Çağında Akut Transvers Myelit. J Clin Anal Med 2014;5(4): 294-9
15. Dalyan M, Kaya A, Akyüz M, Keykubatlı A, Çakcı A.Transvers myelit: 32 0lgunun retrospektif değerlendirmesi. Fiziksel Tıp 1999; 2 (1): 49-52
16. Seifert T, Enzinger C, Ropele S, Storch MK, Strasser-Fuchs S, Fazekas F. Relapsing acute transverse myelitis: a specific entity. Eur J Neurol 2005; 12(9):681-4
17. Borchers AT, Gershwin ME. Transverse myelitis. Autoimmun Rev 2012;11(3):231-48
18. Chen L, Li J, Guo Z, Liao S, Jiang L. Prognostic indicators of acute transverse myelitis in 39 children. Pediatr Neurol 2013;49(6):397-400
19. Bruna J, Martínez-Yélamos S, Martínez-Yélamos A, Rubio F, Arbizu T. Idiopathic acute transverse myelitis: a clinical study and prognostic markers in 45 cases. Mult Scler 2006;12(2):169-73