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Vocal Cord Paralysis in Multifocal Motor Neuropathy: A Case Report

Multifokal Motor Nöropatide Vokal Kord Paralizisi: Olgu Sunumu

Hatice Balaban[1], Özlem Kayım Yıldız[1], İlteriş Ahmet Şentürk[1], Emine Elif Altuntaş[2], İsmail Önder Uysal[2], Ertuğrul Bolayır[1], Suat Topaktaş[1]

[1]*Cumhuriyet University Faculty of Medicine, Department of Neurology, Sivas, Turkey*

[2]*Cumhuriyet University Faculty of Medicine, Department of Otorhinolaryngology, Sivas, Turkey*

Summary

Multifocal motor neuropathy is characterized by a slow, progressive, asymmetrical weakness of the extremities without sensory loss. The disease can also cause vocal cord paralysis as it runs its course. A 34-year-old man developed progressive weakness in his hand muscles for one year. Dysphonia was also present for a three week period. Neurological examination revealed there was moderate weakness and mild muscle wasting in the distal muscles of the upper limbs, predominantly in the left hand. Left laryngeal nerve palsy was also discovered during his otolaryngological examination. Nerve conduction studies revealed multiple sites of conduction block without sensory abnormalities consisting of selective involvement of motor fibers. The patient received intravenous immunoglobulin treatment after a diagnosis of multifocal motor neuropathy. Multifocal motor neuropathy with conduction block is rarely associated with vocal cord paralysis. This condition should be kept in mind in cases of laryngeal nerve palsy with an asymmetric motor neuropathy, as patients might benefit from intravenous immunoglobulin treatment. *(Turkish Journal of Neurology 2012; 18:114-7)*

Key Words: Multifocal motor neuropathy, vocal cord paralysis, diagnosis

Özet

Multifokal motor nöropati duyu bozukluğunun eşlik etmediği, ekstremitelerin asimetrik güçsüzlüğü ile karakterize yavaş, progresif bir nöropatidir. Hastalığın seyrinde nadiren vokal kord paralizisi gelişebilir. Bir yıldır el kaslarında güçsüzlük olan 34 yaşında erkek hasta son üç haftadan beri var olan ses kısıklığı yakınması ile kliniğimize başvurdu. Nörolojik muayenede solda belirgin üst ekstremitelerin distallerinde güçsüzlük ve atrofi vardı. Otolaringolojik muayenede solda laringeal sinir felci tespit edildi. Sinir iletim çalışmaları duyusal sinir iletimlerinde anormallik olmaksızın motor sinir liflerinin seçici etkilenimi ile uyumlu çoklu yerlerde ileti bloğu gösterdi. Multifokal motor nöropati tanısı alan hastaya intravenöz immünoglobulin tedavisi başlandı. İleti bloğunun eşlik ettiği multifokal motor nöropati nadiren vokal kord paralizisi ile birlikte olabilir. Hastaların intravenöz immünoglobulin tedavisinden yarar sağlamaları nedeni ile asimetrik motor nöropati ile birlikte laringeal sinir felci olan olgularda bu durum akla gelmelidir. *(Türk Nöroloji Dergisi 2012; 18:114-7)*

Anahtar Kelimeler: Multifokal motor nöropati, vokal kord paralizisi, tanı

Introduction

Multifocal motor neuropathy with conduction block (MMN-CB) is a rare immune-related peripheral neuropathy characterized by asymmetric onset and slowly progressive weakness (1). Onset is between twenty and sixty years of age and men are more frequently affected than women (2). Conduction block (CB) of the motor nerves outside the usual sites of nerve compression is regarded as the neurophysiological hallmark of the disease (3). Cranial nerve paralysis is not a frequent clinical presentation of the disease. MMN-CB associated with vocal cord paralysis (VCP) is rarely reported. We presented a 34-year-old man who was diagnosed with MMN-CB and suffered from VCP during the course of the disease.

Address for Correspondence/Yazışma Adresi: Hatice Balaban MD, Cumhuriyet University Faculty of Medicine, Department of Neurology, Sivas, Turkey
Phone.: +90 346 258 07 38 E-mail: haticebalaban@yahoo.com
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Case Report

A 34-year-old man was referred to our clinic suffering from hoarseness for three weeks. For a year before this condition was reported, he suffered from a slow, progressive weakening in his hands that he predominately felt in the fingers of his left hand. He reported no sensory symptoms. The patient had no history of exposure to neurotoxic chemicals and his medical and family history was unremarkable.

At admission, physical examination showed no abnormal findings in either the chest or the abdomen. Neurological examination revealed asymmetrically diminished muscle strength in the distal part of the upper extremities, according to the Medical Research Council (MRC) scale. Left abductor digiti minimi (ADM), abductor pollicis brevis (APB), and hand flexors were 4/5 muscles weak. There was mild atrophy in the ulnar nerve innervated muscles on the left hand. Right ADM, hand and arm flexors were weak against resistance. The proximal part of the upper extremities and the lower extremities were intact with normal muscle power and tone. Muscle strength of the neck flexors and extensors were normal. Deep tendon reflexes were absent with flexor plantar responses on both sides. The left soft palate was depressed and left vocal cord paralysis was seen on indirect endoscopic larynx examination (Figure 1). Other cranial nerve examination was normal. Sensation was normal for all modalities. No involvement was seen in sphincter, respiratory or autonomic functions on examination.

The electrophysiological nerve conduction studies performed on admission to the clinic showed motor conduction block in both median, left ulnar, and right peroneal nerves with prolonged or absent F waves (Figure 2). CB, adopted as loss of compound muscle action potential (CMAP) amplitude, exceeded 50% percent between 2 contiguous sites of stimulation, the out site and the usual sites of nerve compression.

Left median and left ulnar motor nerves conduction velocities were mildly reduced at the axillary-Erb and Erb-elbow levels. Right ulnar, bilateral median nerve, and bilateral peroneal nerve F waves, as well as the posterior tibial nerves H reflexes, could not be obtained. The left ulnar F wave was prolonged. Other motor nerves and all sensory nerves conduction studies were within normal range (Table 1). Concentric needle EMG study of the left abductor pollicis brevis and abductor digiti minimi muscles revealed high amplitude and long duration in the motor unit potentials. A reduced interference pattern during voluntary contraction was observed. There was no spontaneous muscle fiber activity. Needle EMG study of the other muscles of upper and lower extremities was normal. These electrophysiological findings showed evidence of MMN-CB.

All diagnostic studies gave normal results, including tests for GM1 antibodies, protein electrophoresis, immune electrophoresis, antinuclear antibody, complements 3 and 4, rheumatoid factor, C-reactive protein, vitamin B12, and folate and thyroid hormones. Serological tests were negative for HIV, syphilis and hepatitis B and C viruses. Cerebrospinal fluid protein level analysis was normal. Chest X-ray, thorax and neck computerized tomography and magnetic resonance images of the brain were also normal. Diagnosis of MMN-CB was made with the results of tests and intravenous immunoglobulin (IVIG) treatment was applied. The motor deficit slightly improved within 4 weeks after a five-day course of IVIG (0.4 gr/kg/day). On follow-up neurological examination 8 weeks after admission, left digiti minimi, interosseal muscles, abductor pollicis brevis and hand flexors were 4-5¯/5 muscle weak. The other muscles were intact with normal power and tone. Bilateral vocal cords were seen as a normal on indirect endoscopic larynx examination (Figure 3).

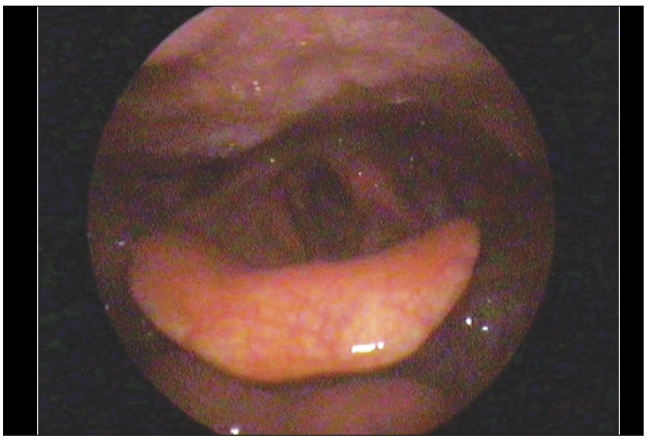

Figure 1. Left vocal cord paralysis.

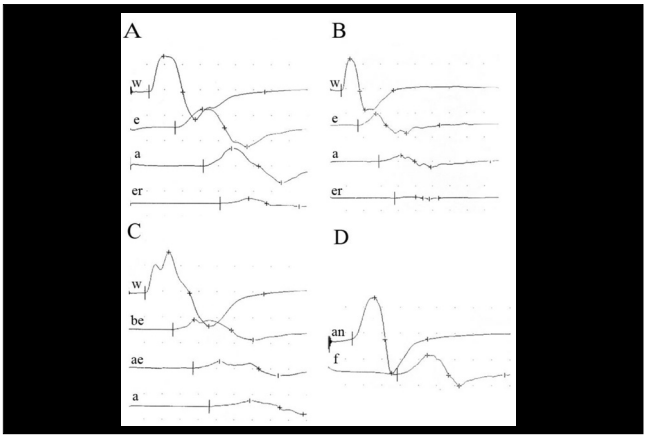

Figure 2. Conduction block for the (A) right median nerve between Erb-axilla, (B) left median nerve between wrist-elbow and Erb-axilla, (C) left ulnar nerve wrist-below elbow, (D) right peroneal nerve ankle-fibula head point sites of the stimulation of patient on admission. w: wrist, e: elbow, a: axilla er: Erb, be: below elbow, ae: above elbow, an: ankle, f: fibula head. Sweep 3ms/division, gain 5mV/division (A, C), 5ms/division, gain 5mV/division (B), 3ms/division, gain 2mV/division (D).

Table 1. Motor nerve conduction study on admission

Peripheral nerve	Distal lat (ms)	Distal CMAP amplitude (mV)	p/d CMAP ratio (%)	Velocity (m/s)	F W	H R
Median, R					NR	
Wrist-APB	3.3	12.6	-41	51.6		
Elbow-wrist		7.4	-9	46.4		
Axilla-elbow		6.8	-76	57.4		
Erb-axilla		1.6				
Median, R						
Wrist-APB	3.6	10.5				
Elbow-wrist		4.0	-62	49	NR	
Axilla-elbow		2.5	-38	34.3		
Erb-axilla		0.4	-83	33.7		
Ulnar, R						
Wrist-ADM	2.4	13.8			NR, +A wave	
Below elbow-wrist		11.9	-14	48.4		
Above-below elbow		10.4				
Erb-above elbow		6.8	-35	45.6		
Axillar-Erb		5.5	-18	51.7		
Ulnar, L						
Wrist-ADM	3.0	14.5			32.2	
Below elbow-wrist		3.8	-74	42.6		
Above-below elbow		2.7				
Erb-above elbow		2.7	-28	31.5		
Axillar-Erb		2.6	-6	57.8		
Tibial R						
Med mal-EHB	2.8	16.9	†	46		NR
FP-med mal		3.9				
Tibial L						
Med mal-EHB	3.0	20.8	†	42		NR
FP-med mal		5.9				
Peroneus R						
Ankle-EDB	4.1	5.9				
Cap fib-ankle		2.9	-60	42	NR	
FP-cap fib		2.9	†	51		
Peroneus L						
Ankle-EDB	3.9	10.3				
Cap fib-ankle		7.1	-31	46.1	48.6	
FP-cap fib		7.0	†	42.3		

R: right, L: left, p/d CMAP ratio: CMAP amplitude ratio on proximal vs. distal stimulation, FW: F wave, HR: H reflex latency, NR=No Response. Med mal: medial malleol, EHB: Extansor hallucis brevis, EDB: Extansor digitorum brevis, Cap fib: Caput fibulae, FP: Fossa poplitea, †:Not evaluated-common entrapment site

Discussion

MMN-CB is a rare immune-mediated disorder of the peripheral nerves. Weakness is usually located in the distal part of the extremities and the upper extremity is affected more early and severe than the lower extremity. Median, ulnar, and radial nerves are affected frequently. MMN is a diagnosis that is based on recognition of a characteristic pattern of clinical symptoms, clinical signs and electrodiagnostic findings (4). CB of motor axons is the fundamental electrodiagnostic finding that distinguishes MMN-CB from amyotrophic lateral sclerosis (ALS) and the other disorders with a comparable clinical phenotype (5). Definite conduction block was diagnosed when CMAP amplitude reduction on proximal versus distal

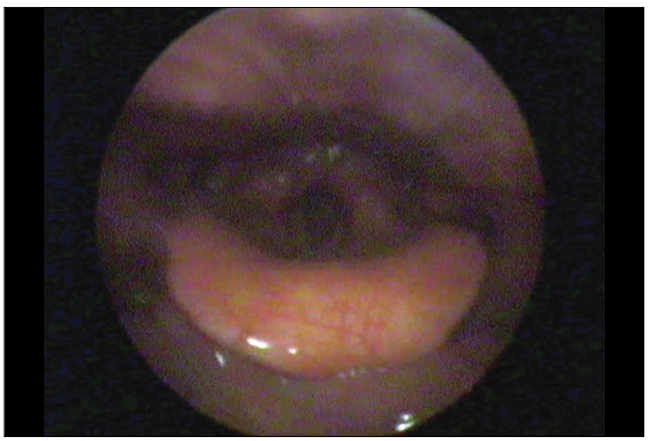

Figure 3. Bilateral normal vocal cords on control examination.

stimulation outside the usual sites of nerve compression of at least 50% in the upper extremities and 60% in the lower were found. One of the electrophysiological criteria is normal sensory conduction in the segments where motor CB exists (4). We did not perform this test in our case. Since the first description of the MMN-CB, many clinical features have been reported. During the course of the disease, cranial nerve involvement has rarely been reported in the literature (6-10).

Kaji et al. (6) described one case with hypoglossal palsy and MMN-CB; Axelsson and Liedholm (7) reported a similar case. Pringle et al. (8) also described a patient with ophthalmoplegia and bibrachial paresis, in which they suggest that their patient had MMN-CB presented with ophthalmoplegia. VCP in MMN-CB has only been reported in the literature twice in the past. The first case was reported by Olchovsky et al. (9) in a patient with Borrelia infection. The second case with VCP was reported by De La Blanchardiere et al. (10). However the author's patient also had HIV-related cytomegalovirus (CMV) infection. To the best of our knowledge, our case is the first in the literature who has vocal cord paralysis with MMN-CB in idiopathic group.

In conclusion, MMN-CB is a rare immune-mediated neuropathy and responds to intravenous immunoglobulin therapy (11). Cranial nerve involvement may lead to diagnostic difficulties. MMN-CB must be kept in mind in cases of cranial neuropathy with asymmetric peripheral neuropathy. Consequently, treatment will be provided upon accurate diagnosis.

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