Case Report / *Olgu Sunumu*

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Two Cases of True Neurogenic Thoracic Outlet Syndrome

Gerçek Nörojenik Torasik Çıkış Sendromlu İki Olgu

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Summary

True neurogenic thoracic outlet syndrome is a very rare clinical entity. Its incidence is about one per million. In this paper, we present the clinical and electrophysiological findings of true neurogenic thoracic outlet syndrome in two patients diagnosed in our electrophysiology laboratory and discuss in the light of the literature.
Key Words: Thoracic outlet, neurogenic, brachial plexus

Özet

Gerçek nörojenik torasik çıkış sendromu çok nadir görülür. İnsidansı yaklaşık olarak milyonda birdir. Bu makalede elektrofizyoloji laboratuvarımızda gerçek nörojenik torasik çıkış sendromu tanısı konan iki olgunun klinik ve elektrofizyolojik özellikleri sunulmuştur ve literatür gözden geçirilmiştir.
Anahtar Kelimeler: Torasik çıkış, nörojenik, brakiyal pleksus

Introduction

Thoracic outlet syndrome (TOS) refers to compression of the neurovascular bundles at the point between the neck and axilla (1). It can be divided into two major categories: neurogenic TOS and vascular TOS. Neurogenic TOS is caused by compression of the lower brachial plexus and is divided into two categories as true and disputed. Disputed or nonspecific neurogenic TOS refers to the clinic entity that symptoms are compatible with TOS, however, no objective neurologic or electrodiagnostic findings are available (2). True neurogenic TOS is considerably rare, the incidence is reported as one in a million (3).

In this paper, we present the clinical and electrophysiological findings of true neurogenic TOS in two patients diagnosed in our clinic.

Case 1

A 55-year-old housewife presented to our outpatient clinic with complaints of pain and paresthesia in her right hand and forearm exacerbating with physical activity and lasting for longer than ten years. She also complained of weakness of this arm for about 2 years. She stated that she noticed some wasting of the muscles of her hand. Her physical examination revealed atrophy of the thenar, hypothenar and interosseal hand muscles at the affected side (Figures 1a-c). Hypoesthesia of the 5th finger and medial aspect of forearm and weakness of intrinsic hand muscles were observed. Muscle strength of abduction of the first and fifth digits were 4/5. There was tenderness on her Erb's point. There were not any vascular signs. Bilateral cervical costae were seen on radiographic examination (Figure 1d).

Bilateral median motor nerve conduction studies on the wrist-elbow segment recording from the abductor pollicis brevis (APB) muscle, and bilateral ulnar motor nerve conduction studies on the wrist-elbow, across elbow, elbow-axilla and the axilla-erb segments recording from the abductor digiti minimi (ADM) muscle were done using superficial electrodes. Bilateral median and ulnar sensory nerve conduction studies on the second and fifth digit-wrist segments recording from the wrists and medial antebrachial cutaneous (MAC) nerve conduction studies recording from the antebrachial region and stimulating from the area medial to the biceps tendon were recorded. In needle electromyography (EMG) APB,

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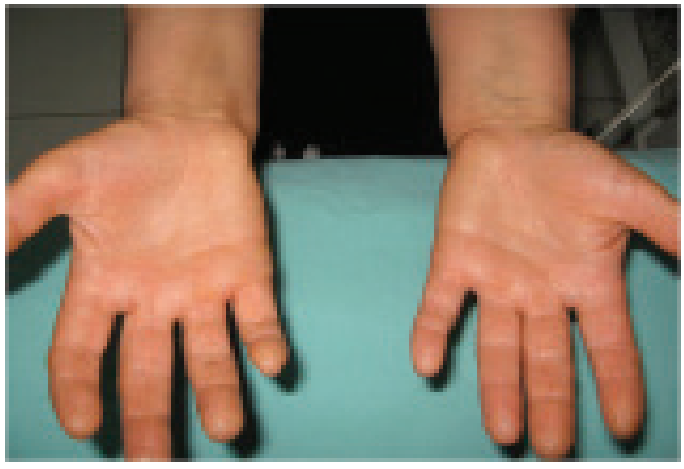

Figure 1a. Thenar and hypothenar atrophies on the right hand of the Case 1.

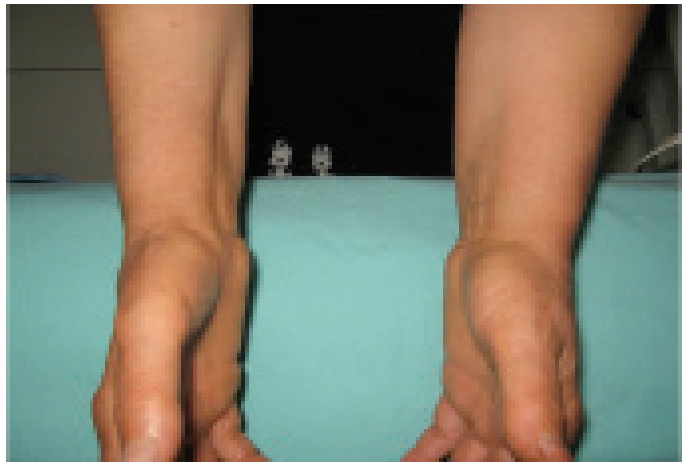

Figure 1b. Thenar atrophy on the right hand of the Case 1.

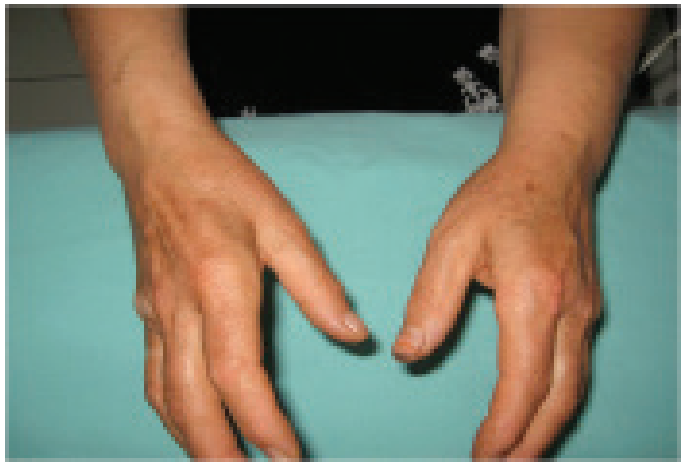

Figure 1c. Interosseal atrophies on the right hand of the Case 1.

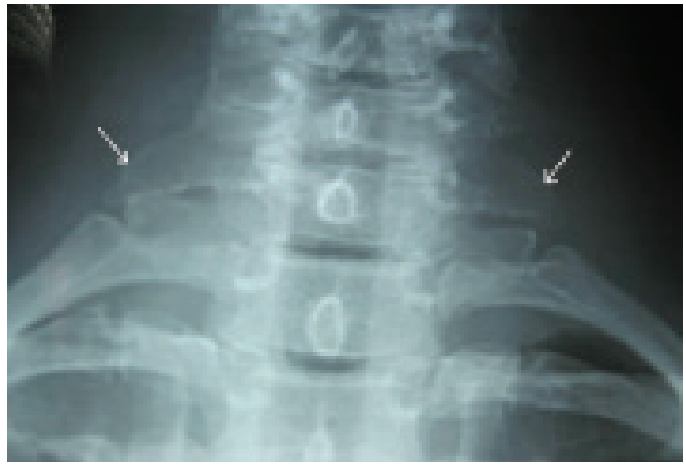

Figure 1d. Bilateral prominent C7 transvers processes on radiography of the cervical vertebral column of the Case 1.

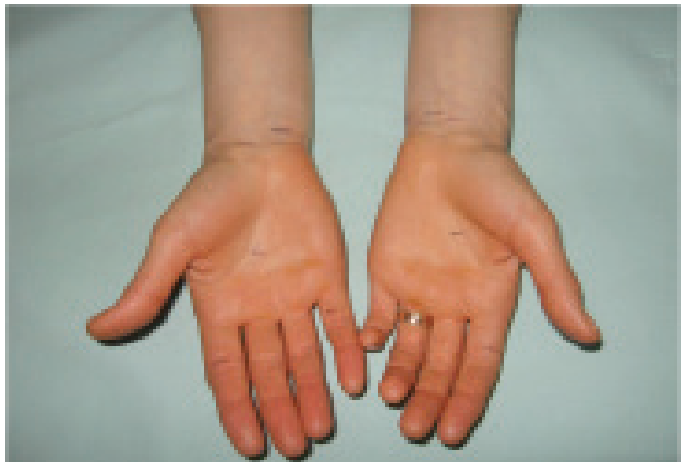

Figure 2a. Thenar and hypothenar atrophies and claw hand on the left hand of the Case 2.

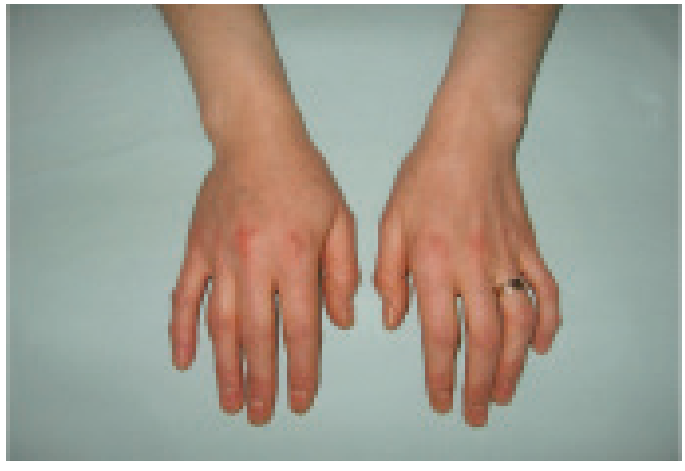

Figure 2b. Interosseal atrophies and claw hand on the left hand of the Case 2.

Table 1. Summary of nerve conduction studies*.

CASE 1					
	Motor		Sensory		
	Median (recording from APB)	Ulnar (recording from ADM)	Median Digit 2-wrist	Ulnar Digit 5-wrist	MAC
	R/L	R/L	R/L	R/L	R/L
Latency (ms)	**4.78**/3.5	3.42/2.76	2.66/2.74	2.26/2.12	**NR**/1.86
Amplitude (mV. µV)	**1.90**/14.02	6.13/12.99	17.4/15.3	**5.4**/9.8	**NR**/21.60
CV (ms)	**43**/54.3	57.8/63.2	48.9/42	44.2/44.8	**NR**/69.9
F wave latency (ms)	29.1/25.8	**32.6**/24.75			
CASE 2					
	Motor		Sensory		
	Median (recording from APB)	Ulnar (recording fromADM)	Median (Digit 2-wrist)	Ulnar (Digit 5-wrist)	MAC
	R/L	R/L	R/L	R/L	R/L
Latency (ms)	2.72/3.04	2.46/3.27	2.7/2.58	2.32/**NR**	1.66/**NR**
Amplitude (mV. µV)	15.97/14.77	17.54/8.77	37.3/52.5	19.8/**NR**	17.4/**NR**
CV (ms)	58.3/52.0	61.6/40.6	50.7/51.6	48.3/**NR**	47.6/**NR**
F wave latency (ms)	29.3/**60.5**	28.5/**35.55**			

NR: No Response. APB: Abductor Pollicis Brevis. ADM: Abductor Digiti Minimi
*Values for amplitude are expressed as mV for motor studies and µV for sensory studies Bold values are abnormal

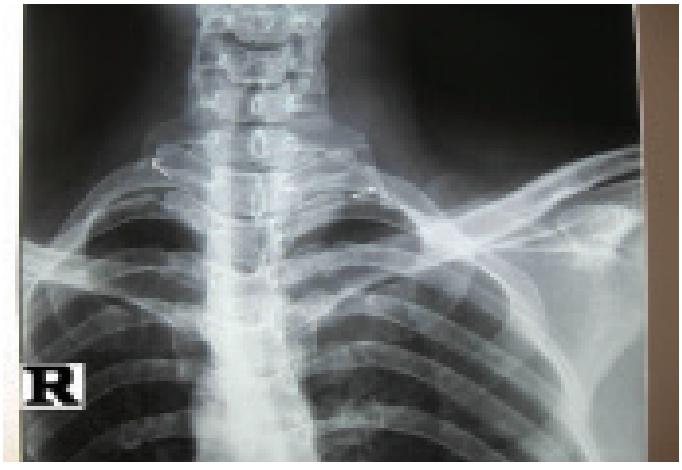

Figure 2c. Bilateral elongated C7 transverse processes and rudimentary costas of the Case 2 (the costa on the left side is longer than the one at the right).

ADM, extensor indicis proprius (EIP) (all by C8, T1 roots and by different peripheral nerve innervated muscles) and some proximal muscles (biceps, triceps) were investigated.

On the affected side, median compound muscle action potential (CMAP) amplitudes were low, median sensory conduction velocity and sensory nerve action potential (SNAP) amplitudes were normal. Ulnar F-wave latency was prolonged and SNAP amplitude was low. MAC SNAP could not be obtained (Table 1). Ulnar CMAP amplitudes and conduction velocities across the wrist-elbow, across the elbow and elbow-axilla segments were normal whereas velocity was found to be decreased across the Erb point-axilla segment, indicating a conduction block through thoracic outlet. However, the amplitude of the ulnar CMAP obtained by wrist stimulation was above the limit value of 5 mV which was rather smaller than the opposite side (6.13/12.99 mV, Table 1).

Needle EMG examination revealed a small amount of denervation potentials in APB and ADM also motor unit rarefaction, increase of polyphasic motor unit potantials (MUP) and chronic neurogenic changes in APB, ADM and EIP muscles, whereas needle EMG findings of the biceps and triceps were normal (Table 2).

Case 2

The second patient was a 41-year-old housewife who presented to our outpatient clinic with the same complaints for about 10 years in her left hand and arm; for two years, the arm has become easily tired and weak. There was atrophy of the thenar, hypothenar and interosseal hand muscles in the left upper extremity (Figures 2a,b). Her 5th finger and medial aspect of the forearm were hypoesthetic like the first case and muscle strength in abduction of the first and fifth digits and opposition of the first digit were 4/5. Her Erb's point was tender as well. There were not any vascular signs. Bilateral cervical costae were seen on radiographic examination (Figure 2c).

In the second patient, median CMAP amplitudes were normal, but median nerve F-wave latency was prolonged compared to the opposite side and to the normal limits. Median sensory conduction velocity and SNAP amplitude were normal. Ulnar F-wave latency was prolonged and ulnar SNAP and MAC SNAP could not be obtained. Ulnar CMAP

Table 2. Summary of neddle EMG findings.

Examined muscles	Fibrillation potentials	Positive sharp waves	Polyphase	MUP duration-amplitude	Recruitment
		CASE 1			
R APB	+	+	↑↑↑	↑	Discret
R ADM	++	++	↑	↑	Discret
R EIP	-	-	↑↑	↑	Reduce
R Biceps	-	-	N	N	Full
R Triceps	-	-	N	N	Full
		CASE 2			
L APB	-	-	↑↑	↑↑	Discret
L ADM	-	-	↑↑	↑↑	Discret
L EIP	-	-	↑	↑	Reduce
L Biceps	-	-	N	N	Full
L Triceps	-	-	N	N	Full

MUP: Motor Unit Potentials, R: Right, APB: Abductor Pollicis Brevis, ADM: Abductor Digiti Minimi, EIP: Extensor İndicis Proprius L: Left, N: Normal

amplitudes and conduction velocities were normal but there was notable difference in ulnar CMAP amplitude between the right and left sides, similar to that in the first case (17.54/8.77 mV, Table1) Motor unit rarefaction, increase of polyphasic MUP's and chronic neurogenic changes in APB, ADM and EIP muscles were recorded. EMG investigation of the proximal muscles was normal (Table 2).

Both patients were advised to consider surgery. The first patient preferred to visit a surgeon in her hometown and did not apply for a control visit, thus, we lost contact with her. The second one refused surgery.

Discussion

True neurogenic TOS is a very rare condition and is seen more commonly in female population (3). There are 11 neurogenic TOS cases diagnosed in our electrophysiology laboratory in which approximately 1200 electroneuromyographic examinations have been performed per year in 13 years of working period. This information was obtained from the archival records but we could not reach the electrophysiological reports of former cases. Forestier et al. (4) reported 4-5 cases/year out of 3500 EMG examinations. Another electroneuromyography clinic reported 2/1000 cases during 17 years of working period (5).

Radiculopathies, motor neuron diseases, entrapment neuropathies, other peripheral neuropathies, and other types of brachial plexopathies should be kept in mind in the differential diagnosis. These possible causes can be discriminated electrophysiologically. Sensory nerve conduction studies are normal in C8-T1 radiculopathies and motor neuron disorders which differ in this way from TOS. Besides, in radiculopathies, needle EMG of the paraspinal muscles are abnormal. Sensory fibers of the median nerve and motor fibers of the deltoid and biceps muscles are affected in upper trunk lesions of the brachial plexus and motor fibers to the triceps muscles are affected in middle trunk lesions of the brachial plexus unlike TOS. All ulnar sensory fibers, all ulnar motor fibers and C8/T1 median motor fibers are carried by the lower trunk, thus, a distinct electrophysiological pattern exists in most patients with neurogenic TOS:

-low/absent ulnar SNAP
-low median CMAP
-borderline/low ulnar CMAP
-normal median SNAP
-low/absent MAC SNAP

The medial antebrachial cutaneous nerve is a sensory nerve that branches out directly from the medial cord of the brachial plexus and innervates the skin on medial side of the forearm. Since an absent or low-amplitude MAC SNAP is a universal finding in all confirmed cases of neurogenic TOS, including mild cases, nerve conduction studies of this nerve are of great importance for patients with suspected lower trunk brachial plexus lesions (2). In another study, it is recommended that MAC sensory study should be performed when other standard electrophysiological tests fail to confirm a lower trunk brachial plexopathy (6).

The first patient's electrophysiological findings corresponded to the electrodiagnostic criteria. In the second one, we found median CMAP amplitudes as normal. Low CMAP amplitude of APB was found in all cases in Levin (7) and Le Forestier's (4) series. Nevertheless, low CMAP amplitude recorded from APB muscle has been reported in 13% to 80% of patient in series of other authors (8-10). We could not obtain MAC SNAP and we showed chronic neurogenic changes in C8/T1 and median/ulnar/radial nerve innervated muscles. Considering these findings, we diagnosed the patient as having true neurogenic TOS.

Some authors claimed that deceleration of ulnar motor nerve conduction across the thoracic outlet is characteristic in patients with TOS, however, subsequent studies showed normal velocities across the Erb's point-axilla and did not correspond with this hypothesis (1). Thus, slowing of ulnar motor nerve conduction between the Erb's point and axilla is not essential. In our cases, ulnar nerve conduction velocity was slow in the first patient whereas it was normal in the second one.

Neural compression could not be confirmed surgically in our cases. If we diagnose new cases with true neurogenic

TOS, we can verify the compressive tissue surgically and hopefully, patients benefit from surgical treatment.

In conclusion, we reported these cases because true neurogenic TOS is a very rare condition and it should be kept in mind in the differential diagnosis of patients with arm or hand pain and atrophy.

Conflict of Interest

Authors reported no conflicts of interest.

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