CLINICAL AND ELECTROPHYSIOLOGICAL FINDINGS IN A CASE WITH INACTIVE LEPROMATOUS LEPROSY*
İnaktif lepramatöz lepralı bir olguda klinik ve elektrofizyolojik bulgular

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Abstract: *Leprosy is a chronic infectious disease caused by Mycobacterium leprae, and mostly involves skin and superficial peripheral nerves. It has two major clinical forms: Lepromatous leprosy and tuberculoid leprosy. The predominant form develops as a result of the nature of the immune response to the microorganism. This microorganism mostly prefers the cooler parts of the body, such as mucous membranes, skin and superficial peripheral nerves for its multiplication. In this report, we aimed to present a 65 year-old female patient with lepromatous leprosy diagnosed and treated 17 years ago. The patient has atrophic cutaneous lesions, bilateral hand and foot deformities and auto-amputations at the distal phalanges of the hands and feet, and bilateral distal severe sensory-motor axonal neuropathy.*

Key Words: *Axon; Leprosy, electrophysiology; Polyneuropathies*

Özet: *Lepra, deri ve periferik sinirleri tutmaya eğilimli Mycobacterium leprae enfeksiyona bağlı olarak gelişen kronik bir hastalıktır. Basillerin mukoz membranlar, deri ve yüzeyel sinirlere affinitesi vardır, çünkü bu sahalar multiplikasyonu için ideal ısıda daha soğuk vücut sahalarıdır. İki majör klinik tip kabul edilmektedir: Lepramatöz ve tüberküloid lepra. Tip mikroorganizmanın immunolojik cevabının natürüne bağlı olarak baskınlık gösterir. Onyedi yıl önce tanı konup tedavi edilmiş, atrofik kütanöz lezyonları, bilateral el ve ayak deformite ve otoampütasyonları olan ve distal ağır sensori-motor aksonal nöropati tespit edilen inaktif lepramatöz lepralı 65 yaşında bir kadın olgu takdim edildi.*

Anahtar Kelimeler: *Akson; elektrofizyoloji; Lepra; Polinöropatiler*

Leprosy is a chronic infectious disease caused by Mycobacterium leprae, with a tendency to involve the skin and the peripheral nerves. It has a tendency to involve mucous membranes, skin and superficial peripheral nerves due to their lower temperature as compared with the body, providing an ideal temperature for the bacillus to multiply. Mycobacterium leprae is one of the common causes of treatable neuropathy in the world. In many cases, the neurological findings are swelling of the nerves, severe sensory deficits, motor paralysis and trophic changes (1-3).

The electrophysiologic findings in leprosy are reported as "lost" or "decreased" amplitudes of sensory nerve action potentials (SNAPs) and compound muscle action potentials (CMAPs), slow nerve conduction velocities, prolonged distal latencies, and focal reducing of conduction rates in locations of nerve swellings (3-11). Late responses, such as F response and H reflex, have been shown to be abnormal (8). It is suggested that findings of chronic denervation are generally observed during electromyographic examinations (4, 11).

Case Presentation

A 65 year-old housewife admitted to the neurology outpatient clinics of Atatürk University, Faculty of

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Medicine in April 2001 complaining of muscle paralyses and hand deformities. She had been diagnosed as having leprosy in Atatürk University Faculty of Medicine 17 years ago. For a 2-year period, she received multiple antilepromatous medicines for treatment and was intermittently followed up for her disease. She has not received any medication for leprosy for 15 years.

In the neurological examination, muscle strength was determined as 2/5 in the distal and 4/5 in the proximal muscles of both upper and lower extremities. In her extremities, there were "gloves and socks" type sensory deficits of both touch and pain. Position and vibration senses were also decreased in the distal parts of the extremities. Facial sensory examination findings were normal. The nerves could not be palpated. Among her deep tendon reflexes, achillles and styloradial reflexes were bilaterally lost, whereas the biceps, triceps and patella reflexes were normal. The minor muscles of the hands and feet were atrophic. There was a contracture and limitation of movement in the right elbow joint. Bilaterally, in all fingers and toes, there were auto-amputations and claw-hand deformities.

In her dermatological examination, multiple hypo and hyperpigmented atrophic plaques of about 1-3 cm diameter, predominantly in the sacral region and the extensor sides of extremities, and loss of eyebrows and body hair supported the diagnosis of Lepromatous leprosy (LL). The case was considered to be at the inactive stage of the disease. The routine blood and urine assessments, fasting blood glucose, urea, creatinine, liver function tests, erythrocyte sedimentation rate, anti-streptolysin O, C-reactive protein, rheumatoid factor, chest X-ray and electrocardiogram of the patient were normal.

Electrophysiologic Studies: *All studies were performed with a Medelec Teca Premerie Plus vE05 electromyograph (Surrey, UK). During the electroneurographic studies, bilateral, symmetric, severe, distally located sensory-motor axonal polyneuropathy (PNP) was detected. The deep and proximally located nerves were found to function normally. The late responses that were studied (F-response, H-reflex) were mildly delayed (Tables I and II).*

In electromyographic examination, the proximal group muscles were normal, whereas the distal group muscles were electrically silent and inexcitable. In the left tibialis anterior muscle, there were chronic denervation findings characterized by long-duration, polyphasic motor unit action potentials and decreased recruitment during maximal contraction. No myopathic potentials were observed (Table III).

Table I. *Motor Nerve Conduction Studies Results*

			Amplitude (mV)		*Latency (msec)*		*Conduction velocity (m/sec)*	
Nerve stimulated	*Stimulation site*	*Recording site*	*RT*	*LT*	*RT*	*LT*	*RT*	*LT*
Facial	*Tragus*	*Frontalis*	-	*1.25*	-	*4.54*	-	-
Spinal accessory	*Erb's point*	*Trapezius*	-	*2.58*	-	*4.16*	-	-
Axillary	*Erb's point*	*Deltoid*	-	*7.11*	-	*5.28*	-	-
Musculocutaneous	*Erb's point*	*Biceps*	*8.26*	*4.52*	*5.08*	*5.58*	-	-
Radial	*Erb's point*	*Triceps*	*4.35*	-	*6.33*	-	-	-
	Spiral groove	*EDC*	*5.01*	-	*4.72*	-	-	-
	Elbow	*EDC*	*4.81*	-	*3.10*	-	*80.2*	-
	Forearm	*EIP*	*0*	-	*0*	-	*0*	-
Median	*Wrist*	*APB*	*0*	*0*	*0*	*0*	*0*	*0*
	Elbow	*APB*	*0*	*0*	*0*	*0*	*0*	*0*
Ulnar	*Wrist*	*ADM*	-	*0*	-	*0*	-	*0*
	Above elbow	*ADM*	-	*0*	-	*0*	-	*0*
Femoral	*Groin*	*Rectus Femoris*	-	*14.2*	-	*5.40*	-	-
Peroneal	*Below fibula*	*TA*	-	*4*	-	*4.74*	-	-
	Popliteal fossa	*TA*	-	*5.84*	-	*8.04*	-	*32.7*
	Ankle	*EDB*	-	*0*	-	*0*	-	*0*
	Below fibula	*EDB- F wave*	-	-	*53.3*	-	-	-
Tibial	*Popliteal fossa*	*GC*	-	*5*	-	*5.8*	-	-
	Popliteal fossa	*GC-H reflex*	-	-	-	*34.5*	-	-
	Popliteal fossa	*AHB*	*0*	*0*	*0*	*0*	*0*	*0*
	Ankle	*AHB*	*0*	*0*	*0*	*0*	*0*	*0*

mV= milliovolt, 0= no response, - = not determined, RT= right, LT= left; EDC= extensor digitorum communis, EIP= extensor indicis proprius, APB= abductor pollicis brevis, ADM= abductor digiti minimi, TA= tibialis anterior, EDB= extensor digitorum brevis, GC= gastrocnemius, AHB= abductor hallucis brevis.

Table II. *Sensory Nerve Conduction Studies Results*

			Amplitude (μV)		***Latency (msec)***		***Conduction velocity (m/sec)***	
Nevre stimulated	***Stimulation site***	***Recording site***	***RT***	***LT***	***RT***	***LT***	***RT***	***LT***
Median	*Wrist*	*Index finger*	*0*	*0*	*0*	*0*	*0*	*0*
Ulnar	*Wrist*	*Little finger*	*0*	*0*	*0*	*0*	*0*	*0*
Radial	*Forearm*	*Snuffbox*	-	*0*	-	*0*	-	*0*
Suralis	*Calf*	*Posterior ankle*	*0*	*0*	*0*	*0*	*0*	*0*

μV=microvolt, 0=no response, RT=right, LT=left, - = not determined

Table III. *Electromyography*

					Voluntary motor unit action potentials			
		Spontaneous activity					***Configuration***	
Muscle	***Insertional activity***	***Fibrillations***	***Fasciculations***	***Activation***	***Recruitment***	***Duration***	***Amplitude***	***Polyphasia***
Left Deltoid	*NL*	*0*	*0*	*NL*	*NL*	*NL*	*NL*	*NL*
Left APB	*NL*	*0*	*0*	*0*	*0*	*0*	*0*	*0*
Left RF	*NL*	*0*	*0*	*NL*	*NL*	*NL*	*NL*	*NL*
Left TA	*NL*	*0*	*0*	*NL*	*↓*	*N/↑*	*NL/↑*	*NL/↑↑*
Left EDB	*NL*	*0*	*0*	*0*	*0*	*0*	*0*	*0*

NL= normal, ↓=slightly reduced, ↑= slightly increased, ↑↑= moderately Increased, 0= no response.
APB= abductor pollicis brevis, TA= tibialis anterior, EDB= extensor digitorum brevis, RF= rectus femoris.

DISCUSSION

The cutaneous lesions of LL are composed mainly of pale lepromatous macules, and lepromatous infiltrations diffusely and symmetrically are distributed on the body. The lepromatous infiltrations may be divided into diffuse, nodular and plaque types. A slow and progression loss of hair begins in the lateral 1/3 of the brow, and later continues on to the eyebrows and body hair (1). During the dermatological examination of the case, multiple hypo and hyperpigmented atrophic plaques predominantly on the sacral region and the extensor sides of the extremities, and loss of body hair and lateral 1/3 of the brow were detected. These findings were supportive of the plaque type lepromatous infiltrations.

Similar to the dermal lesions, the sensory deficits of LL is usually bilaterally symmetric and of "gloves and socks" type. In the axons of the nerves involved, multiple acid-fast microorganisms can be found. If PNP is the beginning manifestation of leprosy, then it may be confused with other cases of PNP such as secondary to diabetes (2, 3).

The neurological findings in leprosy are swollen nerves, serious sensory deficits, motor paralysis, and trophic changes. Due to fusiform swellings, the nerves involved are stiff and palpable. Muscle paralyses usually involve the minor muscles of the hands and feet, and result in progressive atrophy. In addition to claw hand deformities, contractures develop in the fingers. Due to the resorption of phalanges, there is shortening of fingers and toes. In the differential diagnosis of other causes of PNP, sparing of deep tendon reflexes in leprosy is an important criterion. In late periods of the disease; however, reflexes are lost due to severe damage to the nerves (2, 3).

During the neurological examination of our case, bilateral symmetric gloves and socks type loss of sensation, atrophies of the minor muscles of the hands and feet due to paralysis, auto-amputations, and muscle weakness predominantly in the distal segments of the extremities, and bilateral claw hand deformities were detected. The superficial nerves were not palpable. Bilateral styloradial and Achilles reflexes were lost, although other reflexes were normal. These findings indicate a symmetric and distal PNP. As the disease was in the inactive period, no skin or nerve biopsy was performed to reveal the acid-fast microorganisms. Other causes of PNP were ruled out by certain tests including the blood glucose measurement.

In the electrophysiological examination of leprosy, a decrease or loss of the amplitudes of SNAPs and

CMAPs, prolongation of the distal latencies, slowing of sensory and motor conduction rates, and focal slowing of conduction in the locations of nerve swellings have been previously reported (2-11). It has been shown that late responses such as F-wave and H-reflex are abnormal (8). During electrophysiological examinations, it has been proposed that findings of chronic denervation are observed in general (3-11).

In the electrophysiological examination of the present case, sensory and motor responses could not be recorded in the distal segments of both upper and lower extremities symmetrically and bilaterally. However, the recordings from the proximal regions of the nerves and from the deeply localized nerves were normal. The CMAPs and the distal latencies of left facial nerve, as recorded from the frontalis muscle, and of right accessory nerve as, recorded from the trapezius muscle, were normal. These data were supportive of a bilateral symmetric distal PNP predominantly with axonal damage. The complete loss of SNAPs and CMAPs in the distal segments was attributed to the advanced state of the disease. These findings indicate that the nerves in the proximal regions of the extremities and the deeply localized nerves are relatively spared, and this condition confirms the affinity of leprosy to superficial nerves.

In our case, the late responses (F-response, H-reflex) were minimally prolonged. Gupta et al. (8), in their studies discovered a decreased rate of conduction in motor nerves in 36% of the cases, and in sensory nerves in 12% of the cases. They also found that late responses such as H-reflex and F-wave are abnormal in 64% of the cases. They proposed that these late responses are more helpful in the early demonstration of nerve lesions than in conventional nerve conduction studies.

In the electromyographic examination of our case, among proximal group muscles of the upper and lower extremities, the left biceps and rectus femoris muscle activities were normal whereas the left hand thenar group and the left foot extensor digitorum brevis muscles were electrophysiologically silent and inexcitable. In the left leg, there were chronic denervation findings in the tibialis anterior muscle, characterized by long-duration, polyphasic motor unit action potentials and decreased recruitment. There were no myopathic potentials. These findings were considered to result from chronic denervation secondary to nerve damage. Finally, in a study of 40 cases aiming to evaluate the muscular involvement, the electromyographic examination of tibialis anterior muscles revealed findings of chronic denervation in 77.5% of the cases, and no myopathic potential was observed (4).

In conclusion, the clinical findings and the electrophysiological studies in the case presented here indicated the presence of a bilateral, symmetric, severe distal PNP. These findings are parallel to those of the previous reports in the literature. It is important to note that, despite the presence of a severe distal PNP, the proximal segments of the nerves and the deeply localized nerves were relatively preserved.

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