Neuropathy Associated with Leprosy

Jau-Shin Lou, MD, PhD

No one involved in the planning of this CME activity had any relevant financial relationships to disclose. Authors / faculty had nothing to disclose.

CME is available 4/2/2008 - 4/2/2011

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American Association of Neuromuscular and Electrodiagnostic Medicine

2621 Superior Dr NW Rochester, MN 55901

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CME Information
Product: CS21 - Neuropathy Associated with Leprosy

Course Description
Intended Audience
This course is intended for Neurologists, Physiatrists, and others who practice neuromuscular, musculoskeletal, and electrodiagnostic medicine with the intent to improve the quality of medical care to patients with muscle and nerve disorders.

Learning Objectives
Upon conclusion of this program, participants should be able to:
1. formulate a differential diagnosis for distal hand atrophy and sensory loss.
2. design and perform an EMG/NCV study to assess distal hand atrophy.
3. identify the electrodiagnostic pattern associated with mononeuritis multiplex versus distal neuropathy in the upper extremity associated with leprosy.

Release Date: 4/2/2008
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Duration/Completion Time: 1 hour

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Neuropathy Associated with Leprosy

April 2008

CME Available from April 2008 through April 2011

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Presenting Symptom: Left hand atrophy

Case prepared by: Jau-Shin Lou, MD, PhD

Affiliations: Director, EMG laboratory
Director, ALS Center of Oregon Oregon Health and Science University

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Appropriate Audience: Residents and physicians.

Learning Objectives: After completing this educational activity, participants will be able to:
1) Formulate a differential diagnosis for distal hand atrophy and sensory loss
2) Design and perform an EMG/NCV study to assess distal hand atrophy
3) Identify the electrodiagnostic pattern associated with mononeuritis multiplex versus distal neuropathy in the upper extremity associated with leprosy

Level of Difficulty: Intermediate/Advanced.
Neuropathy Associated with Leprosy

History

A 20-year-old woman presented with hand weakness. The history was obtained through a translator. She stated that she developed left hand atrophy numbness and weakness about 5 years ago, right foot numbness, and weakness 3 years ago and similar changes in the left foot at approximately 1 year ago. She denied any body weight loss. She denies any weight loss or other rashes. She denies any known infectious exposure.

Past Med History: Negative
Med: None
FH: Non-contributory

Commentary I

Because different nerves are affected at different time over 5 years and she had both motor and sensory deficits, mononeuritis multiplex is on the top of the differential diagnosis. Brachial and lumbosacral plexopathies or polyradiculopathies involving different extremity over time also need to be considered. Because of sensory involvement, a myopathy is less likely.

Physical Examination

The patient weighed 125 pounds, had a BP of 128/84, and a heart rate of 68. The general physical examination was unremarkable.

The neurological examination showed cranial nerves 2 through 12 were intact.

The motor examination showed:
1) Complete atrophy and 0/5 weakness of the intrinsic hand muscles on the left including abductor pollicis brevis, abductor digiti minimi and first dorsal interosseus.
2) Normal strength in all of the muscles in the forearm including wrist extensors and flexors, flexor pollicis longus, flexor digitorum superficialis, and flexor digitorum profundus.
3) Moderate atrophy of the intrinsic foot muscles was noted.

The sensory examination showed complete sensory loss to all modalities distal to the wrist on the left hand and reduced sensation to all modality distal to the ankle on both sides.
Reflex studies showed 2+ bilateral biceps, triceps, and knee jerk. Ankle jerk was 0.

Figure 1. These two pictures demonstrate atrophy of the intrinsic muscles of the left hand.

Commentary II

Physical examination showed that motor and sensory deficits are limited to the hands distal to the wrists in the upper extremities and the feet distal to the ankles. These finding made mononeuritis multiplex, plexopathy or polyradiculopathy less likely. Weakness and sensory deficits usually are present proximal to the wrists or ankles in mononeuritis multiplex, plexopathy or polyradiculopathy. Nerve entrapments at the wrists and ankles need to be considered.
Laboratory and Electrophysiologic Data

The Chem 20, complete blood count with differential, rapid plasma reagin, anti-nuclear antibody, RA, erythrocyte sedimentation rate, serum protein electrophoresis, B12, HbA1c, and hepatitis C tests were all normal.

<table>
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<th>NERVE</th>
<th>SIDE</th>
<th>STIM SITE</th>
<th>RECORD</th>
<th>cm</th>
<th>AMPL uv</th>
<th>LAT</th>
<th>CV</th>
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<td>Wrist</td>
<td>5th digit</td>
<td>14</td>
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<td>51 (5th -W)</td>
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<tr>
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<td>6</td>
<td>7.1</td>
<td>4.1 (Prolonged)</td>
<td>51 (BE-W)</td>
</tr>
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</table>

NEEDLE ELECTROMYOGRAPHY

INSERTional activity: N, sust, unsust
FIBribillation and POSitive: 0, 1+, 2+, 3+, 4+
AMPlitude: N, inc or dec 1+, 2+, 3+, 4+
DURation: N, inc or dec 1+, 2+, 3+, 4+
Interference Pattern (IP): 0, -1, -2, -3, -4

<table>
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<td>n</td>
<td>n</td>
<td>n</td>
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</tr>
<tr>
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</table>
Summary of abnormal findings:

1. Absent compound muscle action potentials (CMAPs) recording from left APB, FDI, EDB and AH.
2. Absent sensory nerve action potentials (SNAPs) recording from the thumb, 2nd, and 5th fingers and lateral malleolus.
3. Complete denervation of the muscles distal to the left wrist and ankles but not in proximal muscles.

Commentary III

EMG/NCV studies confirmed the clinical findings that only muscles distal to the wrist and ankles are involved. Those muscles proximal to the wrist or ankle were normal. Sensory nerve fibers are also affected in the hands and feet.

Further History

Further history was taken at the follow-up visit 1 month later. During the same time she developed weakness, she had skin pigmentary changes in her left forearm, face, and bilateral legs. Physical examination revealed widespread hypopigmented linear patches on the cheeks, eyelids, arms, legs, and trunk. Her left ulnar nerve and left greater auricular nerve were tender and palpable. A diagnostic test was performed.
Figure 2. The picture shows the hypopigmented areas of the left forearm.

Biopsy from the Hypopigmented Area
Figure 3. Skin biopsy shows granulomatous changes that are typically seen in tuberculoid leprosy. The acid-fast stain for the bacilli was negative (not shown), which is common in patients with tuberculoid epilepsy.

**Diagnostic Impression**

Tuberculoid leprosy affecting peripheral nerves and skin.

**Commentary IV**

Leprosy is caused by an acid-fast bacillus, *Mycobacterium leprae*. The bacilli have a predilection for areas of low temperature: Mucous membrane, skin, cornea, superficial nerves. According to the host's immune response against mycobacterial antigen, leprosy is divided into two major categories: tuberculoid leprosy and lepromatous leprosy. Tuberculoid leprosy is characterized by active immune reaction against *Mycobacterium leprae*. The disease is restricted to a few peripheral nerves or skin lesions as seen in our case. Lepromatous leprosy is characterized by lack of immune response against *Mycobacterium leprae*, and results in extensive proliferation of bacilli in skin and nerves and is associated with severe tissue destruction.

Leprosy is common in South and Central America, China, India, and Africa and uncommon in Europe or North America. In the United States, the number of new cases has increased due to immigration from endemic areas.

The diagnosis of leprosy is not difficult if the possibility of leprosy is kept in mind. The diagnosis is made with diagnostic skin and neuritis lesions. The differential diagnoses include mononeuritis multiplex due to vasculitis, von Recklinghausen disease, hypertrophic interstitial neuritis, and syringomyelia.

The World Health Organization recommends multidrug therapy regimens consisting of a combination of rifampin, dapsone, and clofazimine. Patients with tuberculoid leprosy are generally treated with two drugs (rifampin and dapsone) for 6 months. Patients with lepromatous leprosy are treated with three drugs (rifampin, dapsone, and clofazimine) for 2 years.
Bibliography