Livedo Reticularis Idiopathica Associated with Mononeuropathy Multiplex Syndrome and Bilateral Ulnar-median Nerve Anastomosis

Tae Jin Choi, M.D., Chul Jong Park, M.D., Jong Yuk Yi, M.D., Tae Yoon Kim, M.D., Chung Won Kim, M.D.

Department of Dermatology, Catholic University Medical College, Seoul, Korea

A 31-year-old woman had a livedo reticularis of reticulated, bluish discoloration on both arms and legs for 18 months with a burning pain in the right hand and numbness in both lower legs.

The findings of the electromyography were consistent with mononeuropathy multiplex and bilateral ulnar-median nerve anastomosis.

A biopsy specimen from the right sural nerve showed perivascular lymphocytic infiltration and occasional myelin digestion chambers, which were consistent with vasculitic neuropathy.

We present a patient with livedo reticularis idiopathica associated with mononeuropathy multiplex syndrome who also has bilateral ulnar-median nerve anastomosis.


Key Words: Livedo reticularis, Mononeuropathy multiplex syndrome, Ulnar-median nerve anastomosis

Livedo reticularis is a specific dermatologic manifestation characterized by blood stasis in the superficial venous drainage and mottled bluish discoloration of the skin.

Mononeuropathy multiplex is a peripheral neuropathy characterized by degeneration of two or more nerve trunks. The onset of peripheral neuropathy is characteristically heralded by a severe burning dysesthetic pain in the limbs. Other symptoms include muscle weakness, muscle atrophy, paraesthesiae, and sensory deficits. Mononeuropathy multiplex has been reported to be associated with the systemic vasculitides, especially polyarteritis nodosa and rheumatoid vasculitis.

Ulnar-median nerve anastomosis was first reported by Marinacci and in contrast to the frequent occurrence of median-ulnar nerve anastomosis, is rare.

We present a patient with livedo reticularis idiopathica associated with mononeuropathy multiplex syndrome and bilateral ulnar-median nerve anastomosis.

REPORT OF A CASE

A 31-year-old woman with an intermittently pruritic bluish discoloration on her hands and legs for 18 months presented with an intractable burning pain and tingling sensation in the right hand and numbness in both lower extremities which suddenly developed 6 months ago (Fig.1 A&B).

Her family history was non-contributory.

The following results of laboratory tests were negative or within normal limits: complete blood count, erythrocyte sedimentation rate, blood chemistry, urinalysis, stool examination and he-
Table 1. Motor nerve conduction study

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Amplitude (mV)</th>
<th>Conduction velocity (m/sec)</th>
<th>Distal latency (msec)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>R</td>
<td>L</td>
<td>R</td>
</tr>
<tr>
<td>Median</td>
<td>1.2</td>
<td>14.0</td>
<td>52</td>
</tr>
<tr>
<td>Ulnar</td>
<td>9.2</td>
<td>7.5</td>
<td>60</td>
</tr>
<tr>
<td>Peroneal</td>
<td>5.2</td>
<td>1.9</td>
<td>45</td>
</tr>
<tr>
<td>Tibial</td>
<td>10.8</td>
<td></td>
<td>41</td>
</tr>
</tbody>
</table>

Table 2. Sensory nerve conduction study

<table>
<thead>
<tr>
<th>Nerve</th>
<th>Amplitude(mV)</th>
<th>Distal latency(msec)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>R</td>
<td>L</td>
</tr>
<tr>
<td>Median</td>
<td>15.2</td>
<td>48.8</td>
</tr>
<tr>
<td>Ulnar</td>
<td>66.0</td>
<td>2.8</td>
</tr>
<tr>
<td>Peroneal</td>
<td>3.2</td>
<td>4.3</td>
</tr>
<tr>
<td>Sural</td>
<td>5.6</td>
<td>6.4</td>
</tr>
</tbody>
</table>

Fig. 1. A and B. Livedo reticularis in a woman who had a mononeuropathy multiplex syndrome.

eumononeuropathy multiplex involving both median, sural and peroneal nerves (Fig.2), and bilateral ulnar-median nerve anastomosis (Fig.3). Nerve conduction studies showed as follows (Table 1& 2): 1) There were small amplitudes in the right median and left peroneal nerves. 2) The left superficial peroneal nerves were not involved. 3) Compound muscle action potentials were seen on the abductor pollicis brevis recording on the stimulation of ulnar nerve bilaterally. A needle electromyography showed profuse abnormal spontaneous activities at rest and single voluntary motor unit action potentials on volition in the right ab-

Hepatitis B surface antigen. Radiographs of the chest and abdomen, liver sonograph and electrocardiogram were within normal limits. Immunologic studies including complement (C3, C4), immunoglobulin (G, A, M, E), cryoglobulin and cold agglutinin, and thyroid function tests were within normal limits. Serologic test results were also negative for antibodies to double-stranded deoxyribonucleic acid, Sm antigen, ribonucleoprotein, SSA and SSB antigens, and cardiolipin antigen, but the rheumatoid factor had increased to 60.5 IU/ml (normal 0-20), which does not fulfill the diagnostic criteria of rheumatoid arthritis. Physi-
Fig. 2. Abnormal spontaneous activity on needle EMG. Positive sharp wave(a) and fibrillation potential(b) that mean denervation.

Fig. 4. Histologic findings on the left lower leg show perivascular mononuclear cell infiltration in the papillary dermis (H & E stain, ×400).

Fig. 3. Compound motor action potentials by ulnar and median nerve stimulation. Recordings from right abductor pollicis brevis muscle to stimulation of the ulnar(A) and median(B) nerve at the wrist(a), below elbow(b), at the elbow(c) and below axilla(d).

Abductor pollicis brevis and pronator teres.

A biopsy specimen from the left lower leg showed perivascular mononuclear cell infiltration and mild edema in the papillary dermis (Fig. 4). Direct immunofluorescence was negative for fibrinogen, complement (C3, C1q), and immunoglobulin (G, A, M).

A biopsy specimen from the right sural nerve revealed perivascular lymphocytic infiltration and occasional myelin digestion chambers, which was consistent with vasculitic neuropathy (Fig. 5).

She was treated with acetylsalicylic acid (Aspirin®) 500mg daily and pentoxifylline (Trental®) 400mg two times a day for livedo reticularis, and had a systemic administration of corticosteroid and physical training for the mononeuropathy multiplex.
Livedo reticularis idiopathica associated with…

Fig. 5. Right sural nerve showing perivascular lymphocytic infiltration and occasional myelin digestion chambers (H & E stain, ×400).

DISCUSSION

Livedo reticularis (livedo racemosa) is a mottled or reticulated, pinkish or reddish blue discoloration of the skin, mostly on the extremities, especially the lower legs around the ankles. The pathogenesis of livedo reticularis is still unknown and most cases are usually unassociated with any underlying diseases. However, in some instances, it has been reported to be associated with a number of conditions, including collagen vascular diseases, adverse effects of drugs, vasculitis, infection, metabolic disorders, neoplasms, hematologic diseases, neurologic disorders, pancreatitis, and emboli.

The syndrome of mononeuropathy multiplex is defined as the lesion of two or more peripheral nerve trunks. Mononeuropathy multiplex lesions usually develop asynchronously and produce sensorimotor deficit distributed asymmetrically. Characteristically, the onset of peripheral neuropathy is heralded by a severe burning dysesthetic pain in the limbs. Other symptoms include muscle weakness, muscle atrophy, paraesthesiae, and sensory deficits. Mononeuropathy multiplex can be associated with several disorders such as diabetes, vasculitis, amyloidosis, direct tumor involvement, and paraneoplastic syndromes. As with vasculitic mononeuropathies, the nerves most often affected are the peroneal, ulnar, and median. Involvement of homologous nerves in corresponding limbs (e.g., both peroneal nerves) is common.

Marinacci was the first to describe ulnar-median nerve anastomosis in which the patient, following median nerve trauma, developed denervation of the forearm flexor muscles supplied by the median nerve, but the hand muscles remained unaffected. He suggested that it showed an almost normal sensation in the traumatic hand, because the afferent fibers also traveled with the anastomosis. A median-ulnar nerve anastomosis in the forearm is observed in 6% to 31% of the normal population but ulnar-median nerve anastomosis in the forearm is rare.

In most patients with polyarteritis nodosa, livedo reticularis and neuropathy occur together. Baumgartel et al reported a patient with cold-induced generalized livedo reticularis, peripheral polyneuropathy, repeated acral ulcerations, purpura and essential cryofibrinogenemia. Our patient had livedo reticularis, mononeuropathy multiplex and bilateral ulnar-median nerve anastomosis. She had no evidence of polyarteritis nodosa or associated diseases. She was treated with systemic corticosteroid, acetylsalicylic acid and pentoxifylline. Low doses of acetylsalicylic acid decrease platelet clumping and reduce the cyclooxygenase activity of platelet by 89%, but high doses have the effect of increasing the thrombotic tendency by preventing prostacyclin formation. Pentoxifylline increases blood flow through the microcirculation by increasing erythrocyte flexibility and reducing blood viscosity. After treatment she enjoyed a marked relief of pain, but unfortunately the pain has recurred and the livedo reticularis is still present.

Although a high frequency of mononeuropathy multiplex without connective tissue disease has been previously reported, mononeuropathy multiplex usually occurs early in the course of systemic vasculitis or rheumatoid disease. In our case, there was no evidence of systemic vasculitis or rheumatic disease. However a sural nerve biopsy from our patient revealed perivascular lymphocytic infiltration and occasional myelin digestion chambers, which was consistent with vasculitic neuropathy. The findings of the electromyography were consistent with mononeuropathy multiplex and bilateral ulnar-median nerve anastomosis. We describe a case of livedo reticularis, mononeuropathy multiplex and bilateral ulnar-median nerve anastomosis.
REFERENCES