Clinical, Electrophysiological and Histopathological Diagnosis of Chronic Polyradiculoneuropathy

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ABSTRACT

Peripheral neuropathies are among the most common neurological diseases. They are either inherited or acquired and are often associated with various disorders. This study was designed to 1- Evaluate the role of conventional and non-conventional neurophysiological studies in diagnosis of chronic polyneuropathy and 2- Evaluate the role of histopathologic study for diagnostic purposes. This study was carried out in the Neurology Department, Assiut University Hospital. The study was performed on 25 consecutive patients (20 males and 5 females) who were admitted to our department over the period of one year (May 1st 2002 - April 30th, 2003). They were presented with manifestations of chronic polyneuropathy with duration of illness more than 6 months. Also, the study included age and sex matched 20 normal control individuals (14 males and 6 females). All patients and control were subjected to: A- Complete history taking and clinical examination. B- Neurophysiological studies including (Distal latency, Nerve conduction, F response, H reflex, Conduction block study, and Quantitative electromyography). C- Sural nerve biopsy for patient group only. Patients were classified according to clinical and neurophysiological data in to 3 groups; Chronic inflammatory demyelinating polyneuropathy (CIDP), Hereditary polyneuropathy, and Polyneuropathy of undetermined aetiology. Comparison between three studied groups revealed; a statistical difference in the CMAP amplitude of ulnar nerve being more reduced in CIDP and those of undetermined aetiology compared to hereditary group. Conduction block were found mainly in CIDP group. Sural nerve biopsy revealed segmental demyelination, axonal degeneration, active remyelination, axonal sprouting, and necrotizing vasculitis in different percentage among the three studied groups. (Egypt J. Neurol. Psychiat. Neurosurg., 2005, 42(2): 493-503).

INTRODUCTION

The assessment and investigation of a possible neuropathy is one of the most common clinical problems facing the general neurologist. Studies of the prevalence of neuropathy in the community are rare but suggest a figure of between 2-8%, making peripheral neuropathy at least as common as stroke. Despite this high prevalence of neuropathy, it is only a small proportion of patients with neuropathies who are referred for detailed evaluation, principally those individuals with disabling disease, or with none of the obvious risk factors such as diabetes or alcoholism¹.

The diagnosis of peripheral neuropathies can be frustrating, time consuming and costly. Careful clinical and electrodiagnostic assessment, with attention to the pattern of involvement and the type of nerve fibers mostly affected, narrows the different diagnosis and helps to focus the laboratory evaluation. A meticulous approach to
the evaluation and differential diagnosis of a patient with peripheral neuropathy is essential, based on important elements of the clinical history and physical examination, the use of electromyography and nerve conduction studies, autonomic testing, cerebrospinal fluid analysis and nerve biopsy findings.

**Aim of the work is:**

1. To evaluate the role of conventional and non-conventional neuropsychological studies in diagnosis of chronic polyneuropathy (more than 6 months).
2. To evaluate the role of histopathologic study for diagnostic purposes.

**SUBJECTS AND METHODS**

The study includes all patients admitted to Neurology department, Assiut university hospital over the period of one year (From May 1st 2002 to April 30th 2003), who presented with manifestations of chronic polyneuropathy with duration of illness more than 6 months.

Twenty-five patients of chronic polyneuropathy (20 males and 5 females) were recruited along the period of the study whose age ranged from 16-59 years with a mean age 36.4±10.4 years. A group of 20 normal persons (14 males and 6 females) with a mean age 38.6±22.5 years and age range 19-57 years were chosen as a control group. Patients with polyneuropathy due to metabolic causes (Diabetes, renal diseases, liver diseases, …etc.) were excluded.

All patients were subjected to:

A) Complete history taking, clinical examination, both general and neurological; according to a standard neurological sheet, currently used in the Neurology department, Assiut University Hospital.

B) Neurophysiological studies including:

1- Nerve conduction studies of the following nerves: median and ulnar nerves (Motor and sensory), common peroneal nerve (motor), and sural nerve (Sensory).
2- F response study of the following nerves: median, ulnar, and posterior tibial nerves.
3- H-reflex study of the posterior tibial nerve.
4- Conduction block study of the median and ulnar nerves.
5- Quantitative electromyography of the following muscles: abductor pollicis brevis, abductor digitii minimi, biceps, extensor digitorum brevis, and rectus femoris muscles.

C) Nerve biopsy study:

The nerve selected to be biopsed was the sural nerve. The sural nerve meets six criteria cited by Dyck and Lougren for selection of a peripheral nerve for biopsy.

The control group were subjected to complete clinical examination and the same neurophysiological studies.

Patients were classified according to clinical and neurophysiological data into:

**Group 1:** Twelve Patients (48%) diagnosed as Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP) who fulfilled criteria of Ad Hoc subcommittee for demyelinating neuropathy.

**Group 2:** Nine Patients (36%) diagnosed as Hereditary Polyneuropathy.

**Group 3:** Four Patients (16%) diagnosed as Polyneuropathy of undermined aetiology.

**Consent:** Written consents were taken from the patients after explaining to them the purpose of the study, steps of the work, benefits and risks. Agreement of the Ethical Committee of Faculty of Medicine Assiut University was taken.
RESULTS

Quantitative EMG results:
Statistical comparison was carried between the patients and control group using student’s “t” test.

1- **Amplitude**: There was a statistically significant increase in the mean amplitude of Motor unit potentials of abductor pollicious brevis, Abductor digiti minimi and Extensor digitorum brevis muscles when compared with the control values. However, these finding were not detected from Biceps and Rectors femoris muscles. Eighty percent of cases had abnormal amplitude above the upper limit of normal range (mean for the control group ± 2SD).

2- **Duration**: A statistically highly significant increase in the mean duration of MUPs of the all studied muscles was found when compared with the control group. Eighty four percent of cases had abnormal duration (above the upper limit of normal range).

3- **Size index (area/amplitude + log amplitude)**: There was a statistically highly significant increase in the mean size index of the studied muscles when compared with control group. Eighty four percent of cases had abnormal size index (above the upper limit of normal range).

4- **Phase count**: A statistically highly significant increase in the mean phase count of the studied muscles was found when compared to the control group. All cases were above upper limit of normal range.

5- **Turn count**: There was a statistically highly significant increase in the mean turn count of the studied muscles when compared to control group. All cases were above upper limit of normal range.

6- **Polyphasic MUPs%**: A statistically highly significant increase in the mean values of polyphasic MUPs% of the studied muscles was found when compared to the control group. All cases were above upper limit of normal range.

There was a statistically highly significant positive correlation between the amplitude of MUPs and duration of illness (Fig. 1). No other statistically significant correlations were found between other parameters and duration of illness. There was a statistically significant negative correlation between size index and motor power (Fig. 2). Otherwise, no statistical significant correlations were found between other parameters and motor power.

Neurophysiological data of the studied three groups:
Comparison between the three above mentioned groups was done using ANOVA test.

Table (1) shows a statistically significant difference in distal motor latencies of median and ulnar nerves, being prolonged in CIDP group compared to other groups.

Also, a statistically significant reduction in the CMAP amplitude of ulnar nerve being more reduced in CIDP and those of undetermined aetiology compared to the hereditary group.

Table (2) shows a statistically significant difference in distal sensory latencies of median and ulnar nerves, being more prolonged in CIDP group compared to the other two groups.

Table (3) shows a statistically significant difference in F response latencies of median and ulnar nerves, being more prolonged in CIDP group compared to the other two groups.

Comparison between the three groups for quantitative EMG parameters revealed no consistent statistically significant difference between the three groups apart from the following findings:

A statistically significant difference in mean phase count of APB muscle was found between the three groups, being more in CIDP group compared to the other two groups.

A statistically significant difference in mean amplitude of MUPs of EDB muscle was found, being higher in hereditary polyneuropathy group compared to the other two groups. Also a statistically significant difference in polyphasic MUPs % elicited from EDB muscle was found.
between different groups, being higher in CIDP group compared to the other two groups.

Table (4) shows that, sural nerve biopsy from patients of CIDP revealed segmental demyelination in all patients (100%) with axonal degeneration in 5 patients (41.6%), 4 patients showed active remyelination (33.3%), and 6 patients showed axonal sprouting (50%).

Sural nerve biopsy from patients of hereditary polyneuropathy demonstrated segmental demyelination in all patients (100%), with remyelination in 5 patients (55%) with no axonal degeneration could be detected.

While biopsy of patients of polyneuropathy of undetermined etiology demonstrated axonal degeneration in 3 patients (75%), segmental demyelination in one patient (25%), and necrotizing vasculitis in 3 patients (75%).

Fig. (1): Correlation between duration of illness and amplitude of MUPs.

Fig. (2): Correlation between motor power and MUPs size index.
### Table 1. Mean values±SD of motor conduction studies of the three studied groups.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Median nerve</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal latency</td>
<td>5.7±1.8</td>
<td>4.2±0.8</td>
<td>4.4±0.6</td>
<td>0.05</td>
</tr>
<tr>
<td>MCV</td>
<td>33.7±8.3</td>
<td>44.6±11.9</td>
<td>40.1±6.4</td>
<td>0.05</td>
</tr>
<tr>
<td>Amplitude</td>
<td>2.8±2.7</td>
<td>4.3±2.1</td>
<td>2.0±0.9</td>
<td>0.18</td>
</tr>
<tr>
<td>Duration</td>
<td>18.0±5.4</td>
<td>15.3±3.7</td>
<td>12.5±2.9</td>
<td>0.09</td>
</tr>
<tr>
<td><strong>Ulnar nerve</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal latency</td>
<td>4.5±1.3</td>
<td>3.3±0.7</td>
<td>3.6±0.6</td>
<td>0.47</td>
</tr>
<tr>
<td>MCV</td>
<td>34.3±11.5</td>
<td>43.7±102</td>
<td>42.1±3.7</td>
<td>0.11</td>
</tr>
<tr>
<td>Amplitude</td>
<td>2.4±1.4</td>
<td>4.6±2.1</td>
<td>2.4±0.4</td>
<td>0.01</td>
</tr>
<tr>
<td>Duration</td>
<td>18.4±4.8</td>
<td>14.5±5.7</td>
<td>13.6±3.7</td>
<td>0.08</td>
</tr>
<tr>
<td><strong>Common peroneal nerve</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal latency</td>
<td>7.3±2.6</td>
<td>6.1±2.1</td>
<td>6.3±2.2</td>
<td>0.58</td>
</tr>
<tr>
<td>MCV</td>
<td>26.9±7.3</td>
<td>32.1±16.1</td>
<td>34.3±5.8</td>
<td>0.43</td>
</tr>
<tr>
<td>Amplitude</td>
<td>0.6±0.5</td>
<td>0.6±0.8</td>
<td>0.8±0.4</td>
<td>0.78</td>
</tr>
<tr>
<td>Duration</td>
<td>22.2±3.9</td>
<td>20.7±4.9</td>
<td>18.5±4.0</td>
<td>0.09</td>
</tr>
</tbody>
</table>

Significant: P< 0.05  
* N.B.:  Group 1: Chronic inflammatory demyelinating polyneuropathy (CIDP).  
Group 2: Hereditary polyneuropathy.  
Group 3: Polyneurpathy of undetermined etiology.

### Table 2. Mean values±SD of sensory conduction studies of the three studied groups.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Median nerve</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal latency</td>
<td>5.8±1.2</td>
<td>4.3±0.8</td>
<td>4.5±0.5</td>
<td>0.04</td>
</tr>
<tr>
<td>SCV</td>
<td>30.6±7.7</td>
<td>36.2±7.1</td>
<td>37.4±4.6</td>
<td>0.14</td>
</tr>
<tr>
<td><strong>Ulnar nerve</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal latency</td>
<td>5.5±1.8</td>
<td>4.2±1.0</td>
<td>3.7±0.5</td>
<td>0.05</td>
</tr>
<tr>
<td>SCV</td>
<td>31.2±8.1</td>
<td>35.7±10.0</td>
<td>38.9±6.2</td>
<td>0.27</td>
</tr>
<tr>
<td><strong>Sural nerve</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Distal latency 10</td>
<td>4.4±1.4</td>
<td>5.0±1.2</td>
<td>4.0±1.8</td>
<td>0.41</td>
</tr>
<tr>
<td>Distal latency 14</td>
<td>6.4±1.7</td>
<td>6.6±0.7</td>
<td>5.4±1.7</td>
<td>0.51</td>
</tr>
<tr>
<td>SCV</td>
<td>22.9±6.2</td>
<td>21.6±2.9</td>
<td>24.5±5.9</td>
<td>0.70</td>
</tr>
</tbody>
</table>

Significant: P<0.05

### Table 3. Mean values±SD of late response (F wave and H reflex) of the three studied groups.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Group 1</th>
<th>Group 2</th>
<th>Group 3</th>
<th>Significance</th>
</tr>
</thead>
<tbody>
<tr>
<td>F response</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median nerve</td>
<td>36.3±6.8</td>
<td>30.9±6.4</td>
<td>33.3±1.3</td>
<td>0.05</td>
</tr>
<tr>
<td>Ulnar nerve</td>
<td>38.8±10.1</td>
<td>30.3±5.5</td>
<td>32.3±1.3</td>
<td>0.04</td>
</tr>
<tr>
<td>Posterior tibial nerve</td>
<td>41.3±7.2</td>
<td>37.5±7.1</td>
<td>37.0±4.3</td>
<td>0.09</td>
</tr>
<tr>
<td>H reflex</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Posterior tibial nerve</td>
<td>38.7±7.04</td>
<td>36.3±9.9</td>
<td>34.6±1.1</td>
<td>0.48</td>
</tr>
</tbody>
</table>

Significant: P<0.05

### Table 4. Pathological changes among the different studied groups.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>All patients</th>
<th>Group (1) n=12</th>
<th>Group (2) n=9</th>
<th>Group (3) n=4</th>
</tr>
</thead>
<tbody>
<tr>
<td>No.</td>
<td>%</td>
<td>No.</td>
<td>%</td>
<td>No.</td>
</tr>
<tr>
<td>Segmental demyelination</td>
<td>22</td>
<td>88%</td>
<td>12</td>
<td>100%</td>
</tr>
<tr>
<td>Active remyelination</td>
<td>9</td>
<td>36%</td>
<td>4</td>
<td>33.3%</td>
</tr>
<tr>
<td>Onion bulb formation</td>
<td>9</td>
<td>36%</td>
<td>5</td>
<td>41.6%</td>
</tr>
<tr>
<td>Schwann cell proliferation</td>
<td>11</td>
<td>44%</td>
<td>7</td>
<td>58.4%</td>
</tr>
<tr>
<td>Axonal degeneration</td>
<td>8</td>
<td>32%</td>
<td>5</td>
<td>41.6%</td>
</tr>
<tr>
<td>Axonal sprouting</td>
<td>6</td>
<td>24%</td>
<td>6</td>
<td>50%</td>
</tr>
<tr>
<td>Necrotizing vasculitis</td>
<td>3</td>
<td>12%</td>
<td>0</td>
<td>0%</td>
</tr>
</tbody>
</table>

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Picture (1): Semithin transverse section of sural nerve biopsy (from a patient with CIDP) showing demyelination and presence of remyelination indicated by relatively thin myelinated axon in proportion to axon diameter (arrow).

Picture (2): Semithin transverse section of sural nerve biopsy (from a case of hereditary polyneuropathy) showing segmental demyelination, schwann cell proliferation and onion bulb formation (arrow).

Picture (3): Semithin transverse section of sural nerve biopsy (from a patient with polyneuropathy of undetermined aetiology) showing vasculitis of vasa nervosa appearing as fibrous scarring and thickening of vessel wall and endothelial proliferation with narrowing of vascular lumen (arrow).

Picture (4): Semithin transverse section of sural nerve biopsy (from a case of CIDP) showing segmental demyelination, remyelination indicated by presence of relatively thin myelinated nerve fibers in proportion to their axon diameter, and axonal sprouting (arrow) appearing as four thin myelinated axons surrounded by one schwann cell (arrow).
Electron micrograph of ultrathin section of sural nerve biopsy (from a patient of hereditary polyneuropathy) shows decreased numbers of myelinated nerve fibers with evidence of remyelination appearing as axons with relatively thin myelin sheath in proportion to their diameters. X 2700.

Electron micrograph of ultrathin section of sural nerve biopsy (from a patient with polyneuropathy of undetermined aetiology) showing marked proliferation of endothelial lining of vasa nervosa with thickening of its wall (Vasculitis). X 2700

Electron micrograph of ultrathin section of sural nerve biopsy (from a patient with CIDP) showing decreased number of myelinated fibres with schwann cell proliferation. X 4000
DISCUSSION

Clinicians use electrodiagnostic information to confirm clinical findings, localize specific subclinical abnormalities, and to identify the underlying pathophysiology. Although the primary role of clinical neurophysiology is diagnostic, test results may be sufficiently sensitive to identify sub-clinical findings and to monitor changes related to disease progression or treatment response\(^5\). In this study, patients were considered to have peripheral neuropathy if confirmed by clinical examination as well as specific abnormal neurophysiological studies characteristic for polyneuropathy.

Electromyography examination is an essential diagnostic tool in diagnosis of peripheral neuropathy and can be considered an extension of clinical examination. The widespread use of computers in EMG machines led to the development of new testing procedures in the clinical EMG laboratory\(^6\).

In this study, we use Multi MUPs analysis method for quantitative EMG study in proximal and distal muscles of upper and lower limbs. There was statistically significant difference in the mean amplitude of MUAPs of the patients group compared to the control group. This was in agreement with Stalberg and Flack\(^7\), who reported that the amplitude is a sensitive parameter in discriminating neurogenic from myogenic changes and stated that amplitude is typically increased in neuropathies, after collateral reinnervation and muscle fiber hypertrophy.

The duration of MUAPs in our studied muscles showed a highly significant increase in patients compared to control group. These results agreed with Pfeiffer and Kunze\(^8\), who also reported that the duration is a standard indicator for neuropathic changes. Also, positive correlation was found between the amplitude of MUAP and duration of illness in patients group. These results were in concordance with those of Terebuh and Johnson\(^9\), who stated that with process of reinnervation, healthy motor axon give rise to extensive collaterals for denervated muscle fibers (axonal sprouting), with a resultant increase in MUAP amplitude.

As regard the size index (SI), there was a highly statistically significant increase in mean SI of patient group compared to control group. Also, there was a statistically significant negative correlation between SI and motor power of the studied muscles. These results agreed with Akaboshi\(^10\), who stated that SI is an excellent parameter to distinguish neuropathic from normal MUAPs, and it is not affected by electrode position.

The percentages of suggested subclassification of our patients are partially agreed with Barohn\(^11\), who found, in his study on 402 consecutive patients referred to the University of Texas neuromuscular outpatient clinics, that hereditary neuropathy represent one third of the studied patients, acquired demyelinating polyneuropathy was found in 18%, and 23% of patients diagnosed as chronic sensory polyneuropathy with no identifiable cause, and the remaining patients had chronic polyneuropathy due to metabolic cause (renal, Hepatic, DM ...et) or toxin exposure.

The slight difference between this study and our present study may be due to small number of our studied patients and exclusion of polyneuropathy of acute onset, and those due to metabolic causes.

As regard clinical presentation of the studied groups in this study, a statistically significant more proximal motor weakness was observed among CIDP group compared to the other groups. A statistically significant radicular sensory impairment was observed among CIDP groups, and this can be explained by pathological involvement of proximal parts, of nerves and roots among these patients. This was further supported by the findings that measurement of late responses among the different groups in this study revealed prolonged F response latencies in CIDP group compared to the other groups which again confirms the involvement of proximal nerves and roots by the neuropathic process in CIDP patients.
These findings are consistent with Barohn and Saperstein\textsuperscript{12}, who stated that the finding of weakness in both proximal and distal muscle group in a symmetric fashion is the hallmark for acquired immune demyelinating polyneuropathies, both the acute form Guillain Barre syndrome and the chronic form CIDP.

Neurophysiological assessment of the three groups shows statistically prolonged distal motor latencies and distal sensory latencies of median and ulnar nerves in CIDP group compared to the other groups. These results emphasize the value of distal latency as a sensitive parameter indicating demyelination process. These finding can be interpreted by the more severe and more rapid course of the neuropathic process in CIDP compared to hereditary polyneuropathy, in which the course of illness is more slower and show lesser degree of affection.

Although in most cases, it is difficult to provide a clinical diagnosis based upon specific microscopic findings in the nerve biopsy, there are some conditions in which nerve biopsy can provide specific diagnostic information such as; vasculitis, amyloidosis, HNPP, leprosy, sarcoidosis and tumor infiltration\textsuperscript{13}.

CIDP with purely demyelinating lesions was found in 7 patients (58.3%), and CIDP with mixed demyelinating and axonal lesions were found in 5 patients (41.7%).

These finding are partially consistent with Villa\textsuperscript{14} who found, on his study of 30 patients with CIDP, that all the studied sample showed demyelination, with 9 samples showed associative axonal degeneration. Also, Molenaar\textsuperscript{15} found that 61% of patients with CIDP had sural nerve biopsy with predominantly demyelination.

The relation of whether segmental demyelination and axonal degeneration are independent of each other, or whether there is interdependence between them, was the question of many previous studies. Said\textsuperscript{16}, through their work on patients with Guillain Barre syndrome, came to the conclusion that axonal degeneration developed independently of demyelination. Ashour\textsuperscript{17} studied the histopathological changes in 36 cases of Guillain Barre syndrome. They demonstrated partial demyelination, remyelination with mild axonal degeneration in the studied biopsies. One patients in their studies showed marked axonal degeneration with no accompanying segmental demyelination.

Axonal loss has more long-term prognostic impact than active demyelination, inflammatory infiltrates, or onion-bulb formations. Axonal degeneration occurs in varying degrees in lesions that are attributed primarily to demyelination. This phenomenon is evident in severe lesions of spinal roots and proximal nerve trunks in patients with Guillain-Barre syndrome\textsuperscript{18}.

The axonal lesions occurring in the course of demyelinating neuropathy have been attributed to a bystander effect in inflammatory foci, to immune attack directed toward epitopes on the axon, or to increased endoneurial pressure\textsuperscript{19}. Therefore, in demyelinating disorders of the peripheral and central nervous systems, the long-term prognosis depends more on the amplitude of axonal loss, as recently emphasized in MS, than on demyelination\textsuperscript{20}.

In group of patients with hereditary polyneuropathy, the following changes were observed in their sural nerve biopsies; segmental demyelination, remyelination, onion bulb formation and Schwann cell proliferation. These result correlate with the finding described by Dyck and Lambert\textsuperscript{21}, who stated that in cases of peroneal muscular atrophy (HSMN type one), the sural nerve biopsy showed reduced myelinated fiber density, hypertrophic onion bulb formation, and some axonal atrophy. In this group of common demyelinating Charcot-Marie-Tooth neuropathies (type 1A, 1B, 1C,) with autosomal dominant inheritance, no distinction between subtypes can be made based on morphology, and nerve biopsy does no more to establish prognosis than the combination of the clinical examination, electrophysiology and family history\textsuperscript{22}.

In Dejerine-Sottas neuropathy, the nerve pathology demonstrates a spectrum of changes from congenital amylolination to hypomyelination and others show decreased numbers of nerve.
fibers, demyelination, remyelination, and onion bulb formation. Attempts have been made to classify these cases solely by morphology, but not all agree to this approach. Also in the type of Charcot-Marie-Tooth neuropathies (CMT2, CMT4 and CMTX), nerve biopsy has no specific picture enough to permit a definite diagnosis.

In the third group of patients with undetermined polyneuropathy, the following changes in biopsies was observed: axonal degeneration in 3 patients, segmental demyelination in one patient, necrotizing vasculitis in endoneural vessels in 3 patients. Detection of vasculitis in sural nerve biopsy has special importance since this is a treatable condition. The advent of genetic tests for many neuropathies has reduced the need for biopsy, but it remains the primary method of establishing a diagnosis of vasculitic neuropathy when history is not available from elsewhere.

These findings agreed with Molenaar, who stated that, although sural nerve biopsy may give much information about degree of demyelination, axonal injury and inflammatory cells infiltration, it is not confirmatory to diagnose CIDP.

In addition, the pathologic changes of demyelination or remyelination are not unique to CIDP and can be demonstrated in other neuropathic disorders. It is important to emphasize that there is no diagnostic nerve biopsy findings for CIDP but only suggestive pathologic features.

However, it is sometimes difficult to assess the activity of a chronic polyneuropathy. Nerve biopsy can be used to detect active nerve lesion and inflammatory cell infiltrate which reflect the stage of disease progression. Also, searching for axonal injury, manifestation of regeneration as; remyelination and axonal sprouting, may play an important role in detection of prognosis of these patients. In contrast, detection of blood vessels abnormalities such as necrotizing vasculitis in the studied nerve biopsy is a unique finding which is conclusive for the diagnosis of vasculitis and can’t be diagnosed by any other mean of studies.

Finally, on comparing the neurophysiological studies to the pathological finding in detecting the type of neuropathy, it was found that neurophysiological studies had a sensitivity of 87.5% in detecting of demyelinating form of polyneuropathy, 75% for axonal form of polyneuropathy, and only 40% for mixed form. These results magnify the role of neurophysiological studies in diagnosis of chronic polyneuropathies and detecting the underlying pathological mechanism.

REFERENCES


