Clinical features and tibial nerve somatosensory evoked potential findings in patients with neuromyelitis optica spectrum disorder

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I. BACKGROUND
Neuromyelitis optica spectrum disorder (NMOSD), autoimmune immunopathology with secondary produced demyelination, is distinct from multiple sclerosis (MS), an inflammatory demyelinating disease. The tibial nerve somatosensory evoked potentials (SSEPs) are useful tool for detecting the spinal cord lesion and evaluating its nature of the patients with inflammatory neurological diseases such as multiple sclerosis. However, the tibial SSEPs profiles of NMOSD have been investigated by only two reports so far [Watanabe 2009; Ito 2014].

II. OBJECTIVE
To investigate clinical features and the tibial SSEPs findings in patients with NMOSD.

III. METHODS
• We retrospectively reviewed 26 patients who fulfilled the diagnostic criteria with NMOSD according to the 2015 international panel for NMO [Wingerchuk DM 2015] at our hospital (Hiroshima City Hiroshima Citizens Hospital) between April and December 2016.
• Of the 17 NMOSD patients with spinal cord lesion, 11 patients had the tibial SSEPs studies.
• The central sensory conduction time (CSCT) was calculated as latency between N21 and P38. (Fig. 1)
• Z-score was given using our established normal values considering height, age and gender. Absolute value of the Z-score exceeding 2.5 was considered abnormal.

Fig. 1 The tibial SSEPs and CSCT

CSCT:16.6 (Z-score 0.46)
CSCT:20.4 (Z-score 3.97)
Cz'-Fpz
L1S-Icc
Icc-GTi
PFi-K

IV. RESULTS
• The onset age was 44.7 ± 10.4 years (range 17 – 66).
• Twenty four (92%) were female and two were male.
• Follow-up duration was 10.5 years (range 0.3 – 33).
• Twenty seven (77%) were positive for anti-AQP4 antibody, two (8%) were positive for anti-MOG antibody, and four (15%) were negative for both anti-AQP4 and anti-MOG antibodies.
• The frequency of transverse myelitis presentation in all course was 85%. Seventeen (77%) patients with myelitis presentation had Longitudinally extensive spinal cord lesion (LESCL). (Table 1)
• In NMOSD patients with spinal cord lesion, Z-scores of CSCT were 1.82 ± 1.58, and eight (73%) out of 11 patients showed normal CSCT. (Fig. 2, Fig. 3)
• Three patients showed prolonged CSCT (Z-scores were 3.07, 3.97 and 4.34). (Fig. 2, Fig. 3)
• Disease duration, lengths of the spinal cord lesions and frequency of relapse in patients with abnormal tibial SSEPs take on higher values than those with normal SSEPs. (Table 2)

Fig. 2 Z-scores of CSCT of NMOSD patients

Table 2 Comparison of clinical features between NMOSD patients with normal and abnormal CSCT

<table>
<thead>
<tr>
<th>Z-score of CSCT</th>
<th>Prolonged (n=3)</th>
<th>Normal (n=8)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Female –male ratio (Female)</td>
<td>100% (3/3)</td>
<td>88% (7/8)</td>
<td>0.23</td>
</tr>
<tr>
<td>Positive for anti-AQP4 antibody</td>
<td>100% (3/3)</td>
<td>63% (5/8)</td>
<td>0.77</td>
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<tr>
<td>Age at disease onset (years ±SD)</td>
<td>42.3 ± 24.5</td>
<td>47.02 ± 11.6</td>
<td>0.77</td>
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<tr>
<td>Duration from onset to time of study (years ±SD)</td>
<td>13.6 ± 12.2</td>
<td>8.1 ± 19.2</td>
<td>0.42</td>
</tr>
<tr>
<td>Vertebral lengths of involved spinal segments (±SD)</td>
<td>5.6 ± 2.3</td>
<td>3.3 ± 1.9</td>
<td>0.23</td>
</tr>
<tr>
<td>Frequency of relapse</td>
<td>6.3 ± 7.7</td>
<td>3.0 ± 3.6</td>
<td>0.43</td>
</tr>
<tr>
<td>Annualized relapse rate</td>
<td>0.57 ± 0.50</td>
<td>0.51 ± 0.35</td>
<td>0.72</td>
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</tbody>
</table>

Fig. 3 Representative SSEPs findings of NMOSD patients with normal CSCT and prolonged CSCT

V. DISCUSSIONS
Pathological studies demonstrated that the inflammation, necrosis, and cavitation in NMOSD affect not just the central grey matter but white matter in spinal cord [Wingerchuk DM 2007].
Our study demonstrated that disease duration, lengths of the spinal cord lesions and frequency of relapse in patients with abnormal tibial SSEPs take on higher value than those with normal SSEPs.

VI. CONCLUSIONS
Our findings indicate that the nature of the lesion in the spinal cord is axonopathy rather demyelinating. However, the prolonged CSCT (central sensory conduction time) was observed in some NMOSD cases due to severe spinal cord involvement.

REFERENCES