Chronic inflammatory demyelinating polyradiculoneuropathy associated with intracranial hypertension

The association of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) and intracranial hypertension (IHT) is rare. We report a patient with CIDP who also has IHT.

This was a 28-year-old woman who was referred to a local hospital in November 2013 with one year history of relapsing weakness and numbness of her extremities. On admission, she was unable to walk and hold chopsticks. The cerebrospinal fluid (CSF) protein was raised at 1.27 g/L with no cells. The pressure was raised at 30 cmH2O. The cranial MRI was normal, and electromyography (EMG) showed absence of F-waves of right median nerve and ulnar nerve. She was initially misdiagnosed to have Guillain-Barre syndrome (GBS) and was treated with a course of intravenous immunoglobulin. She was discharged from the hospital with significant improvement.

In November, 2014, she was admitted to our hospital with a three months history of progressive unsteady gait and distal numbness in all extremities. She also complained of severe headache and slightly blurred vision. On examination, muscle strength in the upper extremities was grade 3/5, and in the lower extremities, grade 4/5. There was glove and stocking distribution reduced sensation to touch and pain as well as vibration and proprioception loss in all extremities. The deep tendon reflexes were diminished in all limbs with absent knee jerks. Fundus examination showed bilateral disc edema. EMG showed slowed nerve conduction, prolonged motor distal latencies and absent F-wave latencies, which were consistent with demyelinating polyradiculoneuropathy. Magnetic resonance venography (MRV) was normal.

Table 1 is a summary of the CSF examination performed on 4 occasions. As shown, at the time of her admission to our hospital, the CSF protein was 0.73 g/L with no cells, the intracranial pressure was 32 cmH2O and oligoclonal bands were negative. She was diagnosed to have CIDP and was started on prednisolone 60 mg/day. In the following days, her motor strength, paresthesia, headache, blurred vision and papilloedema all improved. The intracranial pressure dropped to 24 cmH2O after 10 days, with the CSF protein being lower at 0.49 g/L. There was no further improvement clinically after another week when she had a flu-like illness. Repeat lumber puncture showed an opening pressure of 29.5 cmH2O, and protein 0.54 g/l. With added antibiotics and prednisolone being continued, the CSF opening pressure improved to 21 cm H2O and protein to 0.41 g/L. There was no headache and blurred vision, with further improvement in motor strength and paresthesia after a month of treatment.

The association of CIDP and IHT is rare; we found only 5 cases previously reported in the literature. All the reported patients showed elevation of intracranial pressure and CSF protein, which improved with steroid and other treatments. The clinical presentation of IHT could start simultaneously or months to years after the initial symptoms of CIDP. In our patient, there was also fluctuation of the IHT accompanying the relapse of limb weakness and CSF protein.

The precise mechanism of CIDP associated IHT remains uncertain. Some authors hypothesized that high CSF protein contributes to IHT by increasing CSF outflow resistance or by increasing intracranial venous pressure at sites of CSF absorption. The levels of intracranial pressure in our patient varied according to the fluctuations of CSF protein, consistent with CSF protein contributing towards IHT. This
Table 1: Details of CSF analyses

<table>
<thead>
<tr>
<th>Date</th>
<th>Pressure (cmH2O)</th>
<th>White cells (10^6)</th>
<th>Protein (g/L)</th>
<th>Glucose (mmol/L)</th>
<th>Other examinations of CSF</th>
</tr>
</thead>
<tbody>
<tr>
<td>November 29, 2013</td>
<td>30.0</td>
<td>0</td>
<td>1.27</td>
<td>3.91</td>
<td>-</td>
</tr>
<tr>
<td>November 25, 2014</td>
<td>32.0</td>
<td>0</td>
<td>0.73</td>
<td>4.19</td>
<td>Oligoclonal bands were negative</td>
</tr>
<tr>
<td>December 5, 2014</td>
<td>24.0</td>
<td>1</td>
<td>0.49</td>
<td>4.16</td>
<td>-</td>
</tr>
<tr>
<td>December 12, 2014</td>
<td>29.5</td>
<td>0</td>
<td>0.54</td>
<td>4.06</td>
<td>-</td>
</tr>
<tr>
<td>December 18, 2014</td>
<td>21.0</td>
<td>1</td>
<td>0.41</td>
<td>4.08</td>
<td>-</td>
</tr>
</tbody>
</table>

- indicates not performed

Hypothesis is also supported by other diseases, such as subarachnoid hemorrhage, chronic meningitis and poliomyelitis, which also demonstrate an association between CSF protein and IHT. However, there have also been cases of Guillain-Barré syndrome with IHT and normal CSF protein levels reported, suggesting that excessive CSF protein may not be the only explanation for IHT. Conversely, cases of progressive visual failure and polyradiculopathy with areflexic quadriparesis secondary to raised intracranial pressure have also been reported, suggesting that the polyradiculoneuropathy could be a consequence of IHT. The proposed mechanism was mechanical compression of nerve roots due to increased CSF pressure distending the subarachnoid space.

In conclusion, we report a case of CIDP associated IHT. The precise mechanism of the association remains controversial.

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REFERENCES