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CORRESPONDENCE

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 cmH_2O. 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 cmH_2O 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 cmH_2O 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 cmH_2O, and protein 0.54 g/l. With added antibiotics and prednisolone being continued, the CSF opening pressure improved to 21 cm H_2O 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 diagnosis of CIDP in our patient was based on prolonged clinical course, motor-sensory neuropathy, absence of other causes, EMG showing evidence of demyelinating polyradiculoneuropathy, raised protein and absence of pleocytosis in CSF, and response to steroids. With a close temporal relationship between the IHT and CIDP, both improving with steroid treatment, and the absence of other causes such as cerebral venous thrombosis or idiopathic intracranial hypertension[1], we thought the IHT in our patient is associated with her CIDP.

The association of CIDP and IHT is rare; we found only 5 cases previously reported in the literature.[2-6] 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 the 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.[2] 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

Date	Pressure (cmH_2O)	White cells (10^6)	Protein (g/L)	Glucose (mmol/L)	Other examinations of CSF
November 29 , 2013	30.0	0	1.27	3.91	-
November 25,2014	32.0	0	0.73	4.19	Oligoclonal bands were negative
December 5, 2014	24.0	1	0.49	4.16	-
December 12 , 2014	29.5	0	0.54	4.06	-
December 18 , 2014	21.0	1	0.41	4.08	-

– 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.[7] 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.[1] 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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