Liver transplantation in hepatic myelopathy

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Abstract
Hepatic myelopathy (HMy) is a rare neurological complication of cirrhosis that involves spastic paraplegia caused by lateral cord demyelination. It occurs spontaneously or is encountered more often as a result of surgical shunting of portosystemic shunts. It can cause progressive extremity deficit and has been shown to affect lower extremities more often than upper extremities. Herein, we report a young adult woman presenting with spasticity and paraparesis in extremities after intrahepatic portosystemic shunting (TIPS) application and underwent deceased liver transplantation (LT). A 39-year-old woman underwent deceased LT because of cryptogenic liver cirrhosis. She underwent a TIPS procedure 5 years ago. After that, hepatic encephalopathy and spasticity appeared. She was on the waiting list for 3 years. Neurological findings after LT significantly decreased, but did not return to normal. After the emergence of neurological findings, the earlier LT can provide improvement in neurological findings.

Keywords: Cirrhosis; hepatic myelopathy; liver transplantation.

Introduction
Hepatic myelopathy (HMy) is a very rare complication of cirrhosis. It is caused by portosystemic shunting, which can precipitate to bypass the liver.[5] It can occur spontaneously or be encountered more owing to surgical portosystemic shunts. It causes progressive extremity deficit, and it has been shown that lower extremities are more often affected than upper extremities.[1] Herein, we report a young adult woman presenting with spasticity and paraparesis in extremities after performing transjugular intrahepatic portosystemic shunting (TIPS), and deceased donor liver transplantation (DDLT).

Case Report
A 39-year-old woman was diagnosed with cryptogenic liver cirrhosis in 2006, had recurrent esophageal bleeding, and was performed endoscopic band ligation multiple times. Therefore, TIPS was applied to the patient in 2011. After this procedure, attacks of hepatic encephalopathy started and then spasticity and paraparesis began to occur in both lower extremities, eventually causing gait defects gradually. Her autoimmune and viral parameters were all negative. There was no abnormality in her blood test results except for thrombocytopenia and hyperbilirubinemia. She was referred to the neurology department and was diagnosed with HMy eventually. She had no living donor candidate, therefore she was admitted to the list for liver transplantation (LT) with a model for end-stage liver disease (MELD) score of 16. She was on the waiting list for 3 years, and DDLT was performed in 2016. There was no problem during the postoperative follow-up period. After 3 months from transplantation, her spasticity was reduced partially, and electromyography was found normal. Her spasticity significantly decreased and there was no need to walk with crutches anymore. She has been followed up without any problems in the outpatient clinic for 6 years.

Discussion
HMy is an unusual neurological complication of chronic liver disease, causing progressive symmetrical loss of myelin in the lateral pyramidal tracts with spastic plegia in extremities with a minimal sensory deficit. Early diagnosis and early treatment, such as LT, are crucial to improving the quality of life.[6] The cervical spine demyelination is the predominant finding, but as the disease progresses, it is likely to be permanent. A few case reports state that HMy may occur after the TIPS procedure, similar to the patient in our case report.[4] The pathophysiology of HMy was not understood properly, which is caused by the portosystemic shunts in liver dysfunction. Portosystemic shunts cause some essential nutritional deficiencies to the nervous system, and some nitrogenous products such as ammonia and mercaptans precipitate to bypass the liver.[4] Many patients with HMy possess normal or minimal sensory findings. No correlation was found between the grade of hepatic dysfunction and the stage of identifying HMy. Generally, the traditional imaging methods may not help in the diagnosis. Hepatic myelopathy can be diagnosed with history, neurological examination, and neurological imaging to exclude other myelopathies. The prognosis is poor because neurological damages are progressive and irreversible.

Treatments such as reducing blood ammonia levels, other conservative medical treatments, or surgical treatments such as colonic diversion, ligation, and shunt repair have been tried.[6,8] However, there is no effective treatment that can cure, especially after clinical findings are established. It has been accepted that the best effective treatment is LT. However, Counsell and Wurlow reported a case with HMy, but did not find any improvement even after 18 months after LT.[9] They suggested that this result may be due to the high Child-Pugh score and various degrees of neurological complications. In our case, there was a significant im-
improvement in neurological findings in the early period after LT. Caldwell et al.\(^{[10]}\) suggested that LT should be performed in the early stage to consider MELD exception points to the patients with HMy.

Consequently, LT can be reasonable in patients with HMy due to decompensated chronic liver disease with Child-Pugh B and C. Nevertheless, for patients who have a normal liver function or Child-Pugh A, the effect of LT and other treatments is controversial.

### Conclusion

In young patients with chronic liver disease, chronic encephalopathy, and/or slow-onset upper motor damage signs and symptoms, progressive spastic paraplegia in lower extremities in the absence of primary neurological disease should be remembered as a rare complication of HMy. In these patients, LT can provide improvement in neurological findings.

**Informed Consent:** Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

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### References

1. Scobie BA, Summerskill WH. Permanent paraplegia with cirrhosis. Arch Intern Med 1964;113:805-810. [CrossRef]