CASE REPORT

Hepatic myelopathy: an unusual neurological complication of chronic liver disease presenting as quadriparesis

Prakash Kori, Ritesh Sahu, Anupam Jaiswal, Rakesh Shukla

Department of Neurology, King George's Medical University, Lucknow, Uttar Pradesh, India

Correspondence to
Professor Rakesh Shukla,
rakeshshukla_rakesh@rediffmail.com

SUMMARY

Hepatic myelopathy is a rare neurological complication of chronic liver disease, which is usually seen in adults, presenting as pure motor spastic paraparesis. It is almost always associated with portosystemic shunts and hepatic decompensation. We report a rare case of a young adult man presenting as rapidly progressive spastic quadriparesis because of hepatic myelopathy and associated spontaneous splenorenal shunt and early hepatic decompensation.

BACKGROUND

Hepatic myelopathy is a rare neurological complication of chronic liver disease first described by Leigh and Card¹ in 1949 followed by a detailed description of hepatic or portosystemic myelopathy by Brown² and Zieve *et al.*³ This neurological condition is not easily identified and often overlooked in clinical practice. Hepatic myelopathy is rarely found without hepatic encephalopathy. In most of the reported cases it was associated with pure motor spastic paraparesis without bowel and bladder involvement; however, spastic quadriparesis has rarely been reported. Here we report a case of a young adult who presented with pure motor quadriparesis without bowel and bladder complaints and normal sensory examination without any impairment of cerebellar and extrapyramidal system.

CASE PRESENTATION

A 28-year-old man presented to our department of neurology with a history of difficulty in walking because of stiffness and weakness in both lower limbs. The weakness gradually progressed over 3 months to involve bilateral upper limbs. There was no history suggestive of sensory impairment and symptoms of bowel and bladder involvement were absent. No history of any chronic illness was noted. There was also no history of any ocular abnormalities, seizures, diabetes mellitus, hypertension, tuberculosis, trauma, fever, exposure to drugs, toxins, radiation or sexual promiscuity. On examination his vitals were stable; there was mild pallor without any icterus, clubbing or lymphadenopathy. Asterixis was not seen. No external markers of chronic liver disease were evident. Per abdomen examination showed mild splenomegaly. Central nervous system examination revealed the patient to be conscious, alert with mild impairment of attention. Cranial nerves were intact. Motor power of all limb muscles was grade 2/5 (MRC grading).

Muscle tone was increased in all limbs (Grade 1 spasticity according to modified Ashworth Scale) with brisk deep tendon reflexes and plantar response was extensor bilaterally. All modalities of sensation in upper and lower extremities were normal. There was no evidence of cerebellar and extrapyramidal dysfunction. There were no signs of meningeal irritation.

INVESTIGATIONS

Investigation revealed haemoglobin 10 g/dl (normal range (N) 14–16 g/dl), normal total white blood cell count and platelet count. Liver function tests showed normal total serum bilirubin (N 0.3–1.3 mg/dl), aspartate (N 12–38 U/l) and alanine transaminase (N 7–41 U/l) level. Alkaline phosphatase level was raised (240 U/l, N 30–180 U/l). ELISA for serum hepatitis B surface antigen (HBsAg) was positive. Other viral markers like hepatitis C virus (HCV RNA) and HIV were negative. Serum albumin was low 3.0 g/dl (N 4.1–5.3 g/dl). Prothrombin time was prolonged with International Normalised Ratio (INR) of 1.6 (N 1). Blood urea, serum creatine and electrolytes were within normal limits. Serum ammonia level was high (80 µg/dl, N 15–60 µg/dl). Blood mercaptan level was above normal range (N 5–10 nmol/ml). Serum vitamin B₁₂ (N 279–996 pg/ml) and folic acid levels (N 5.4–18.0 ng/ml) were within normal limits. Ultrasound abdomen showed coarse echo texture of liver with mild splenomegaly and extensive splenorenal collaterals suggestive of chronic liver disease. Portal vein diameter was 18 mm (N 10–15 mm). EEG showed generalised slowing of background suggestive of encephalopathy. Upper gastrointestinal (GI) endoscopy showed grade 2 oesophageal varices. Magnetic resonance imaging (MRI) of cervical, thoracic, lumbosacral spine with contrast did not show any abnormal findings. Nerve conduction studies were within normal limits.

TREATMENT

The patient was treated conservatively with β-blockers along with antihepatic encephalopathy measures with which patient responded. Upper GI endoscopic variceal banding was performed. The patient was offered hepatic transplantation but he refused owing to financial constraints.

OUTCOME AND FOLLOW-UP

On follow-up after 6 months the patient showed improving trend with respect to increase in motor

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power up to grade 3/5 with no evidence of further hepatic decompensation.

DISCUSSION

The first documentation of hepatic myelopathy from the Indian subcontinent was provided by Pant *et al.*⁴ There is a paucity of literature of hepatic myelopathy causing quadriparesis, that is, mainly affecting cervical cord. Upper extremities are usually spared or minimally affected if at all.⁵ Our patient had cervical myelopathy causing quadriparesis wherein the upper limbs are equally involved as the lower limbs, which have rarely been reported. Hepatic myelopathy is a frequently overlooked complication in patients with chronic liver disease, which is likely to be more frequent than the few case reports, following the initial description by Leigh and Card in 1949.

The pathogenesis of hepatic myelopathy is poorly understood. It is generally accepted that portocaval shunts or less commonly splenorenal shunts play an important role even in the absence of liver dysfunction.^{6–7} It has been postulated that alteration of the hepatic metabolism leads to deficiencies of essential nutrients for the central nervous system and nitrogenous products such as ammonia, fatty acids, indoles and mercaptans, bypassing the liver through the portocaval shunt, play a contributory role.⁸ These nitrogenous products cause injury to the axon cylinder, neuronal cell bodies and myelin. Furthermore, a discrepancy exists between the tissue reaction in the corticospinal tract and sparing of other systems in the spinal cord, where axonal degeneration and demyelination, cytoplasmic astrocytosis and round cell infiltration occur, and the reaction in the brain and brain-stem, where there is proliferation of Alzheimer's type II glia. This discrepancy of the sensitivities indicates that there are two pathogenic mechanisms, one responsible for the lesions in the brain and brain-stem and the other for those in the spinal cord. Alternatively, a single factor may be responsible for the development of both lesions, and morphological discrepancy reflects differences in the sensitivity of the tissues in these.⁹ Our patient had extensive splenorenal collaterals signifying spontaneous shunt. Differential diagnosis of hepatic myelopathy includes amyotrophic lateral sclerosis, multiple sclerosis, paraneoplastic effect, radiation myelopathy, human T-Lymphotropic Virus Type 1 associated myelopathy and vascular spinal cord disease. Our patient had chronic liver disease because of hepatitis B infection, and presented as pure motor quadriparesis with sparing of bowel and bladder and sensory system. Normal MRI and biochemical investigations ruled out a compressive lesion as well as other causes of non-compressive myelopathy, thus features of early hepatic encephalopathy and spastic quadriparesis, in our patient, favoured the diagnosis of hepatic myelopathy. We believe that the overall clinical picture and results of laboratory studies described above are sufficient to make a diagnosis of hepatic myelopathy. The unusual thing about our patient is that cervical spinal cord was affected, which is rare compared with thoracic cord involvement in hepatic myelopathy. Thus, a high index of suspicion is

required so that the diagnosis of this uncommon condition is not missed in a case of chronic liver disease. Hepatic myelopathy has been shown to be relentlessly progressive, unresponsive to ammonia lowering measures and liver transplantation (barring rare documentations of recovery).^{10–12} The liver transplant could fully reverse the effects of hepatic myelopathy in patients with early disease; whereas it has little effect in patients with advanced disease.¹⁰ One case report also showed that therapy to decrease ammonia level at the earliest stage was effective for gait disturbance.¹³

Learning points

- ▶ Hepatic myelopathy should be considered as a differential diagnosis in a patient with quadriparesis with chronic liver disease.
- ▶ MRI should be advised to rule out other common causes of myelopathy.
- ▶ Early diagnosis and prompt management of liver disease may improve overall prognosis of neurological complications.

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Competing interests None.

Patient consent Obtained.

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