Case Report

Hepatic myelopathy: A rare complication following extrahepatic portal vein occlusion and lienorenal shunt


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A 19-year-old gentleman presented with slowly progressive spastic paraparesis, 2 years after the therapeutic lienorenal shunt for portal hypertension secondary to cirrhosis and portal vein occlusion. After 2 years of initial evaluation, the motor functions had not worsened further. He did not have any obvious clinical or EEG features of hepatic encephalopathy. Other causes for myelopathy were ruled out. Contribution of portal vein occlusion to portosystemic shunting has not been reported previously in patients with 'hepatic myelopathy.' This uncommon complication needs to be considered in patients with shunt surgery for relieving portal hypertension.

Key words: Hepatic myelopathy, lienorenal shunt, portal vein occlusion

Introduction

Shunting of blood from the portal to systemic circulation in patients with chronic liver disease is associated with wide spectra of neurological manifestations. The syndrome of spastic paraparesis, called hepatic myelopathy (HM)[1] or portal-systemic myelopathy,[2] a well-recognized entity, is often subclinical in majority[3] and potentially reversible following timely liver transplantation.[4,5]

We report slowly progressive spastic myelopathy observed in a young man, secondary to portal-vein occlusion and cirrhosis of unknown etiology.

Case Report

A 19-year-old farmer manifested symptoms of stiffness of legs, tripping over objects and tingling sensation of legs of 2 years duration. There was no involvement of upper limb, sphincter or erectile function, cognitive or behavioral changes, tremors or myoclonus. Four years back, he had recurrent hematemesis and pitting pedal edema. There was no history of jaundice or alcohol consumption. He had no family history of liver or neurological illness. He was earlier diagnosed to have extrahepatic portal-vein obstruction with portal-vein cavernoma causing cirrhosis of liver, splenomegaly and grade II esophageal varices. He underwent splenectomy with left lienorenal shunt, following which hematemesis and limb edema remitted.

He had sparse axillary and pubic hair. There was no skin hyperpigmentation, KF ring, pedal edema, spider-nevi, gynecomastia, testicular atrophy and caput medusae. Liver span was reduced to 8 cm. He scored 30/30 on MMSE (Folstein). He had sparse axillary and pubic hair. There was no skin hyperpigmentation, KF ring, pedal edema, spider-nevi, gynecomastia, testicular atrophy and caput medusae. Liver span was reduced to 8 cm. He scored 30/30 on MMSE (Folstein). There was bilateral Achilles’ tendon contracture and spasticity with pyramidal weakness in both lower limbs. Muscle stretch reflexes in the lower limbs were exaggerated and plantar response was extensor bilaterally. Gait was spastic. There was no other deficit.

There was reduced hemoglobin (11.6 gm/dL) and normocytic, normochromic anemia. Liver function tests revealed normal serum bilirubin (0.6 mg/dL) and alanine aminotransferase (39 U/L), minimally raised aspartate aminotransferase (68 U/L), reduced total protein (5 gm/dL) and albumin (2.6 gm/dL) and abnormal A:G ratio (1.1:1). Plasma ammonia was elevated to 99 mmol/L (N=11-32 mmol/L). Renal function tests and serum electrolytes were normal. Craniospinal magnetic resonance imaging (MRI) was normal [Figure 1a, b, c]. CSF was acellular with normal protein (18 mg/dl). Immunological tests (CSF) for syphilis, tuberculosis and HTLV-1 were negative. CSF immunoelectrophoresis showed oligoclonal bands. Visual, brain stem auditory and somatosensory evoked potentials and motor (median, ulnar, common peroneal) and sensory (median, ulnar, sural) nerve conduction studies were within normal limits. Thyroid functions and serum TSH estimation were normal. Serum HIV (ELISA) was negative. Serum Hexoseaminidase level was normal. Ultrasound (abdomen) showed features of cirrhosis of liver with multiple portal vein collaterals and nonvisualization of spleen. Doppler study showed functioning lienorenal shunt. Contrast-enhanced CT (abdomen) confirmed these observations [Figure 1d].

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Upper gastrointestinal endoscopy disclosed esophageal varices. Stool for occult blood was negative. Liver biopsy was attempted but sample could not be obtained. Investigations for Wilson’s disease and markers for chronic viral hepatitis were negative.

A diagnosis of spastic paraparesis due to HM secondary to portacaval shunting was considered. In view of significant esophageal varices, lienorenal shunt was not reversed. He was treated conservatively with protein restriction, physiotherapy and baclofen. At follow-up after 2 years, he was neurologically stable. The myelopathic features, including MRC grade and activities of daily living had not worsened. Repeat liver function tests were normal except for reduced total protein (4.8 gm/dl) and albumin (2.3 gm/dl) and raised plasma ammonia of 89 mmol/L. EEG showed normal background activity without any triphasic waves or paroxysmal activity [Figure 1d]. His MMSE (Folstein) score remained 30/30; however, detailed neuropsychological assessment revealed slight impairment in visual learning and memory, abstract thinking and working memory, suggesting minimal involvement of bifrontal (right>left) and right temporal lobe functions.

**Discussion**

Leigh and Card were the first to describe the occurrence of pure motor spastic paraparesis following liver failure and many such reports have been documented thereafter. Gospe et al were the first to report a progressive HM due to hepatic failure in a 14-year-adolescent. Interestingly, ours is the second such report of HM, with onset at 17 years. Interval between shunt surgery and onset of neurological illness is varied and ranges from 6 to 36 months. The delay is longer in patients with lienorenal shunts. The characteristic profile consists of insidious progression of lower limb spasticity and weakness. Typically, patients have bounts of hepatic encephalopathy. Our patient did not have any obvious clinical or electrophysiological (EEG) features of hepatic encephalopathy.

Our patient developed symptoms within 2 years of shunt surgery with a profile consistent with HM. Interestingly, there was no worsening of neurological features for the last 2 years. We speculate that early onset of myelopathy may have been due to significant shunting as a result of several factors: cirrhosis and spontaneous shunting, portal-vein occlusion and cavernoma leading to anastomosis and shunting and lastly, therapeutic lienorenal shunting. Portacaval shunting plays a substantial role in the pathogenesis of HM. Spontaneous shunting occurs within the liver as a compensatory mechanism. In cirrhosis with portal hypertension, surgical shunting is performed to reduce the portal pressure, as in portacaval shunts and lienorenal shunt. In the latter, portal vein is anastomosed to the splenic vein. HM was initially noted following surgical placement of a portacaval shunt, but it may also occur after transjugular intrahepatic portosystemic shunt (TIPS). Our patient had cirrhosis at the time of presentation and also had additional spontaneous portacaval shunting. Doppler and CT scan confirmed the functioning of lienorenal shunt. In addition, there was hyperammonemia and absence of some secondary sexual features. Symptomatic myelopathy following lienorenal shunt is uncommon. Cranial MRI, CSF studies, serum B₁₂ and multimodal evoked potentials were normal. There was no clinical or MRI evidence to suggest adrenoleucodystrophy. Hexoseaminidase levels were normal. Infections like tuberculosis, syphilis, HTLV, HIV were ruled out. The significance of positive oligoclonal bands in CSF is unclear. Whether it will provide insight regarding pathophysiological mechanism needs to be explored.

Autopsy studies in patients with HM have shown selective demyelination in the corticospinal tracts, Betz cell loss and some authors have even regarded HM as a restricted form of encephalopathy. Nitrogenous toxins, e.g., ammonia that inappropriately enters the systemic circulation as a result of shunting, are implicated in its pathogenesis. HM responds
poorly to treatment. Liver transplantation may reverse some of the neurological changes.\textsuperscript{[4,5]}

\textbf{Conclusion}

We report an unusual instance of chronic HM following lienorenal shunting at a young age without any obvious features of hepatic encephalopathy. This rare complication needs to be considered in patients offered shunt surgery for relieving portal hypertension. Portal vein occlusion as a contributory cause to portosystemic shunting has not been previously reported in patients with hepatic myelopathy.

\textbf{References}


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