Case Report JOURNAL OF CLINICAL AND EXPERIMENTAL HEPATOLOGY

Recanalized Paraumbilical Vein Leading to Hepatic Myelopathy Causing Spastic Paraparesis in a Patient with Chronic Liver Disease

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A 34-year-old male visited our hospital with complaints of recurrent episodes of altered behavior since past 6 months along with difficulty in walking since past 3 months. He was diagnosed of chronic liver disease in the past. Examination revealed spasticity and brisk deep tendon reflexes in both the lower limbs. His blood investigations and spinal cord imaging was normal. Based on his clinical features, a possibility of portosystemic shunting leading to portosystemic encephalopathy (PSE) and shunt myelopathy was suspected. A computed tomography portography showed a recanalized paraumblical vein draining portal blood into external iliac veins. Patient underwent shunt occlusion (Figure- 2). One month after the procedure, while there was no recurrence of symptoms of PSE, those of myelopathy remained unchanged. Shunt myelopathy is a rare complication of spontaneous or iatrogenic portosystemic shunts. Unlike PSE, the management of shunt myelopathy is uncertain due to limited evidence. Limited evidence suggests reversal of myelopathy after early shunt occlusion, highlighting the irreversible changes that may set in spinal cord due to delayed diagnosis. Our case highlights an important but a rare complication of portosystemic shunting in chronic liver disease which should be kept in mind if these patients develop symptoms attributable to spinal cord disease. (J Clin Exp Hepatol xxxx;xxx:xxx)

Hepatic myelopathy is an infrequent complication of cirrhosis.[1] It is seen in patients of advanced cirrhosis with recurrent hepatic encephalopathy and having an associated portosystemic shunt. The most commonly reported symptom is spastic motor weakness with a predilection to affect lower limbs although quadriparesis has also been reported.[2] Hepatic myelopathy has also been reported in patients after the creation of shunt in cases of portal cavernoma and after transjugular intrahepatic portosystemic shunting (TIPS).[3] The most common spontaneous portosystemic shunt seen and reported in literature is the lienorenal shunt.[4] Here we report a case of hepatic myelopathy who presented with spastic motor weakness due to a recanalized umbilical vein shunt.

CASE REPORT

A 34-year-old male presented with recurrent episodes of altered behavior which included altered sleep wake cycle and incoherent speech for six months. He had been incidentally diagnosed with compensated cirrhosis caused by hepatitis B virus 8 months back and was on oral antiviral therapy. These episodes of altered behavior were discerned to be hepatic encephalopathy based on history, exclusion of other causes, and elevated ammonia levels (125 umol/L at baseline) in the blood. He also complained of difficulty in walking with a feeling of tightness in both the lower limbs which had been gradually increasing for the last 3 months. It was associated with a sensation of tripping while walking. There was no history of upper limb involvement, foot drop, numbness, paraesthesias, bladder involvement, or speech disturbance. There was no prior history of neurological illness, back pain, or trauma. On examination, the tone was increased in the lower limbs with hyperreflexia and power was found to be reduced equally across muscle groups in the lower limb. He had no prior history of jaundice, gastrointestinal bleed prior, and no hepatocellular carcinoma on imaging.

The motor weakness was of upper motor neuron type and localized anatomically to the lower spinal cord. Other systemic neurological examination including sensory parameters, cranial nerve examination and higher mental functions appeared intact. He, however, was found to be in covert hepatic encephalopathy based on the number

Keywords: portosystemic encephalopathy, shunt, cirrhosis, TIPS
Received: 9.4.2022; *Accepted:* 27.9.2022; *Available online:* xxx
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Abbreviations: CLD: Chronic liver disease; MELD: model for end stage liver disease; PSE: Portosystemic encephalopathy; TIPS: Transjugular intrahepatic portosystemic shunt
https://doi.org/10.1016/j.jceh.2022.09.008

Please cite this article as: Mundhra et al., Recanalized Paraumbilical Vein Leading to Hepatic Myelopathy Causing Spastic Paraparesis in a Patient with Chronic Liver Disease, Journal of Clinical and Experimental Hepatology, https://doi.org/10.1016/j.jceh.2022.09.008

connection test (completion of task in 90 s against a reference of 30 s).

Laboratory parameters revealed a low platelet count and a serum albumin of 3.1 g/dL while other parameters of liver function, including bilirubin, liver enzymes, and prothrombin time, were all within normal limits. The MELD score was 8. He had no history of alcohol use, no diabetes, and a normal level of vitamin B12. Gastroscopy showed small low risk esophageal varices. Spinal and brain imaging in the form of magnetic resonance imaging was normal. A triple phase cross sectional imaging in the form of computed tomography scan was done. It showed a dilated recanalized paraumbilical vein draining portal blood into external iliac veins (Figure. 1). No other significant shunt was noted. The diameter of shunt was 16 mm. The liver on imaging was reported as nodular in outline with caudate lobe hypertrophy with a span of 9.6 cm in craniocaudal outline. Based on his clinical features, a possibility of portosystemic shunting leading to portosystemic encephalopathy (PSE) and shunt myelopathy was suspected.

After discussion in a multi-disciplinary team meeting, it was decided to occlude the recanalized paraumbilical vein with the support of interventional radiology to prevent further encephalopathy episodes and possibly prevent

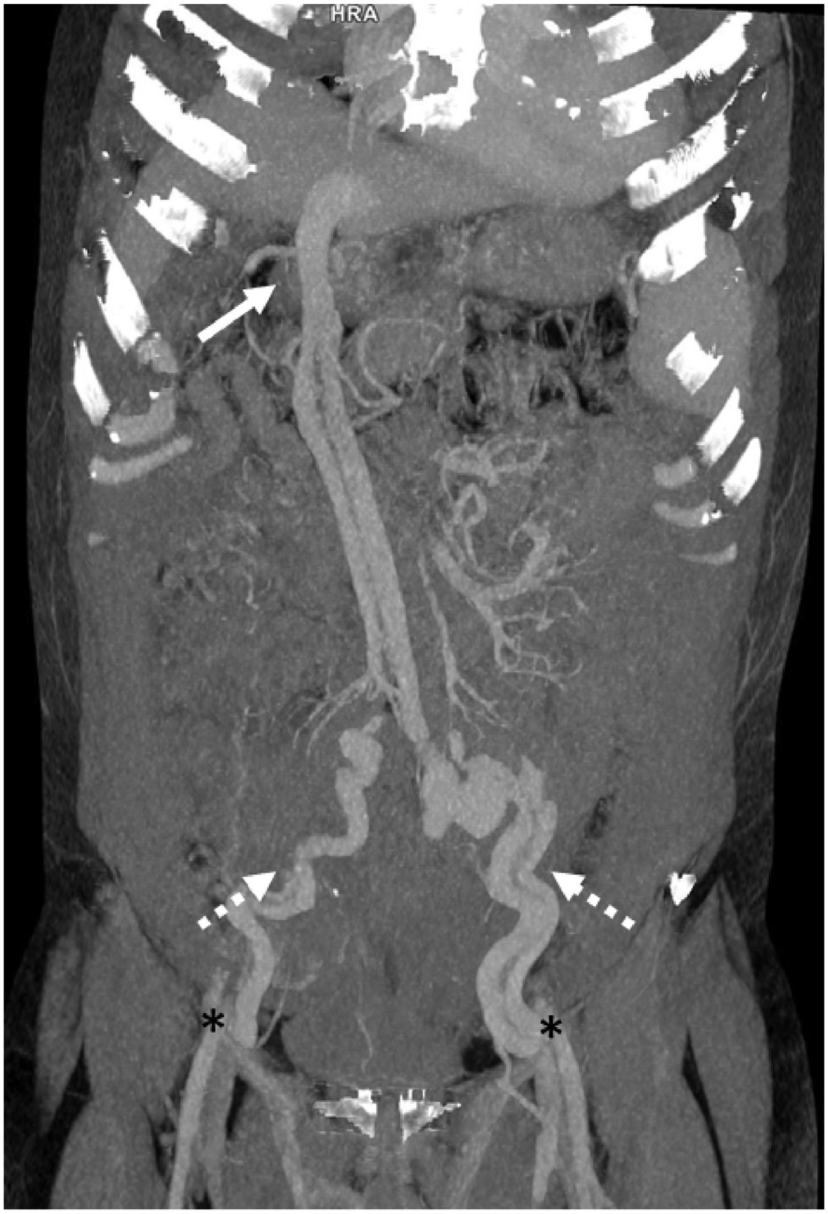

Figure 1 Portosystemic shunting by recanalized paraumbilical vein. Coronal maximum intensity projection of contrast enhanced CT shows enlarged recanalized paraumbilical vein (arrow) communicating with bilateral inferior epigastric veins (dashed arrows) to drain into the external iliac veins (asterisk). CT, computed tomography.

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progression of myelopathy. The patient was counseled regarding possible worsening of portal hypertension and its complications. He underwent shunt occlusion using vascular plug deployment in the recanalized paraumbilical vein under combined ultrasound and digital subtraction angiography guidance (Figure. 2). Ultrasound guidance for gaining access to the shunt, following the access, 10F vascular sheath was placed and rest of the procedure was completed under DSA. Shunt closure was done with a 22 mm Amplatzer™ Vascular Plug followed by gel foam mixed with sclerosant was injected proximal to the plug and the inferior epigastric veins were embolised with glue (Figure. 2). Despite an anticipated risk of worsening of portal hypertension, it was decided to close the main stump of shunt to reduce the risk of clinical failure.

On follow-up, the patient had no further recurrence of hepatic encephalopathy in the next 12 months of follow-up. Although there was no progression of the spasticity or worsening of deficits, no improvement was seen in neurological symptoms. Follow-up imaging at one month showed collapsed collateral with no significant flow in the paraumbilical vein. On repeat gastroscopy, no enlargement of varices was noted at 12 months after procedure. A repeat ammonia done at 1 year of follow-up was 55 umol/L. Baclofen was started for improving spasticity in view of limited improvement of myelopathy related symptoms.

DISCUSSION

PSE is a neuropsychiatric syndrome which develops in patients with liver dysfunction and having a significant portosystemic shunt. The blood from splanchnic circulation is circulated through collaterals rather than hepatic sinusoids. Due to the shunting, ammonia and other toxic metabolites reach and breach the blood brain barrier instead of being detoxified by the liver. In most cases, hepatic myelopathy is associated with hepatic encephalopathy.[1] In India, the first documented case of hepatic myelopathy was reported by Pant *et al.*[5]

The pathogenesis of hepatic myelopathy is poorly understood primarily because of the uncommon nature of the disease. It mostly occurs with other features of liver dysfunction, however, in rare cases, may be the presenting manifestation of cirrhosis with relatively preserved liver function tests.[6]

It has been postulated that shunting of the blood deprives the brain of essential vitamins and amino acids and this deficiency plays a role in myelopathy.[1] Also some waste products like ammonia, indoles and mercaptans damage the axon cylinder, cell bodies and myelin.[7] There are also theories regarding the differential response to injury in the corticospinal tract leading to a relative sparing of the other tracts, however none of those theories have been adequately proven. For the diagnosis of hepatic myelopathy, it is important to have a high index of suspicion in the appropriate clinical context along with the exclusion of structural and primary neurological diseases (Table 1).

Our case is unique because the patient had myelopathy as one of the presenting features (along with encephalopathy) on a background of compensated chronic liver disease. The collateral in our case leading to portosystemic shunting was the recanalized paraumbilical vein which is a relatively unusual shunt to be associated with PSE or myelopathy. Umbilical vein recanalization has been reported to occur in ~20% of patients with cirrhosis.[8] However, in this study, it was not found to increase hepatic encephalopathy as compared to those without umbilical vein recanalization. It is likely that the larger the diameter, as in our case, the higher the chance of hepatic encephalopathy due to higher portosystemic shunting.[9] Chronic HE due to paraumbilical vein shunting has been reported.[10]

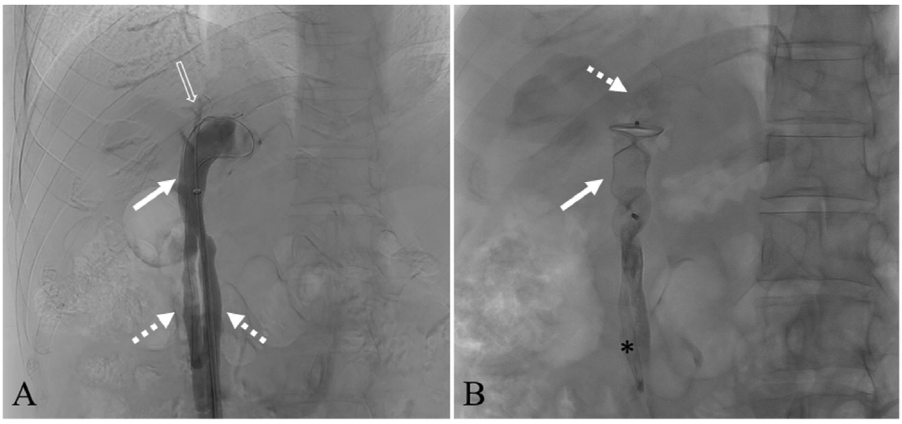

Figure 2 Percutaneous plug assisted closure of recanalized paraumbilical vein. Diagnostic venogram through left inferior epigastric vein (**a**) shows enlarged paraumbilical vein (arrow) draining into bilateral inferior epigastric veins (dashed arrows) with filling of small portal branches (open arrow). Shunt closure (**b**) done with a 22 mm **Amplatzer™ Vascular Plug** (arrow). Gelfoam mixed with sclerosant was injected proximal to the plug (dashed arrow) and the inferior epigastric veins were embolized with glue (asterisk).

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Table-1 Differential Diagnosis of Spinal Cord Involvement in Patients With Cirrhosis.[14]

Differential Diagnosis of Hepatic myelopathy	
Brain Pathology	Demyelinating processes Hydrocephalus Parasagittal space occupying lesion Arnold chiari malformation Other structural abnormalities at craniocervical junction
Spinal cord pathology	Compressive myelopathy Vascular myelopathy (spinal cord infarction, bleeding, vasculitis) Radiation myelopathy Genetic (Hereditary spastic paraplegis, adrenoleukodystrophy) Metabolic/Nutritional (B12 deficiency – Subacute combined degeneration, copper deficiency, lathyrism) Neoplastic (intra or extramedullary tumors, metastasis, paraneoplastic myelitis) Infections (AIDS, HTLV-1 associated myelopathy, erbs spastic paraplegia – syphilis) Motor neuron disease (amyotrophic lateral sclerosis) Hepatolenticular degeneration

Paraumbilical vein recanalization has been reported to be responsible for hepatic encephalopathy in case reports but ours is the first case as per our knowledge to show its association with hepatic myelopathy.

The duration of shunting required for the occurrence of changes in nervous system is difficult to assess in spontaneous shunts. The median duration of onset of myelopathy from the creation of portosystemic shunt has been assessed in artificially created shunts like TIPS or lienorenal shunt for portal hypertension. Portocaval shunts develop myelopathy quicker than partial, non-portocaval shunts (median duration 16 months vs. 60 months).[11]

Treatment of hepatic myelopathy is often not rewarding. Some people have demonstrated the usefulness of shunt closure by measuring ammonia levels reduction.[12] In our case, the encephalopathy showed a dramatic reduction; therefore, we did not measure ammonia level during follow-up. Hepatic myelopathy is more difficult to treat and has been shown previously to be irreversible in later stages. The possible reason for this is poor regenerative capacity of neurons and the lasting damage cause on myelin and cell bodies by nitrogenous waste products. Hepatic myelopathy secondary to TIPS is also a known complication with similar pathogenesis. It has been shown in these cases, early treatment (<6 months from onset to treatment) and intervention has a much better likelihood in predicting response.[13] It is likely that a similar mechanism played a role in our patient but for a possible longer subclinical phase, thus preventing the reversal or improvement in neurological status. Unlike hepatic encephalopathy, which is reversible, liver transplant may not be a relevant option for progressive or non-improving myelopathy.

No liver disease or type of shunt has been shown to increase predisposition to myelopathy. So, being more aware of this condition and astute clinical examination may be the only way to make an early diagnosis which is of paramount importance in such cases. A higher index of suspicion should be kept in patients with recurrent hepatic encephalopathy. This may lead to expedited intervention and prioritization to prevent further neurological damage.

CREDIT AUTHORSHIP CONTRIBUTION STATEMENT

Sandeep Mundhra – Investigation, Data curation, Writing original draft.

Srikant Mohta – Investigation, Data curation, Writing original draft.

Shivanand Gamanagatti – Investigation, Resources, Writing – review and editing.

Sanchit Sharma- Resources, Writing – review and editing.

Naren Hemachandran – Investigation, Supervision.

Anoop Saraya – Resources, Writing – review and editing.

CONFLICTS OF INTEREST

None.

FUNDING

None.

DISCLOSURES

None.

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INFORMED PATIENT CONSENT FOR PUBLICATION

Obtained.

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