

Case Report

Hepatic encephalomyelopathy: a complication following liver cirrhosis caused by Budd-Chiari syndrome and HBV

Jun Cheng*, Shou-Wei Jiang*, Zhong-Song Zhou, Qiu-Lin Sun

Department of Infectious Diseases, The First Affiliated Hospital of Anhui Medical University, Hefei, Anhui Province, China

*authors contributed equally to this work

Abstract

Progressive encephalomyelopathy is a rare neurological complication of chronic liver disease, even manifesting progressive spastic paraparesis. Few reports detailing the clinical and diagnostic aspects of this uncommon cause of neurological deterioration in patients with hepatic insufficiency have been published. Early recognition of this disorder will become more important in the future as patients with liver disease survive longer due to medical advances, including liver transplantation. The case of a patient with hepatic encephalomyelopathy associated with Budd-Chiari syndrome and HBV-related cirrhosis is presented.

Key words: Budd-Chiari syndrome; HBV; Liver cirrhosis; Hepatic encephalomyelopathy

J Infect Dev Ctries 2014; 8(4):551-553. doi:10.3855/jidc.3052

(Received 27 September 2012 – Accepted 19 October 2013)

Copyright © 2014 Cheng *et al.* This is an open-access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Introduction

The development of hepatic encephalomyelopathy (HEM) in patients with a broad range of liver diseases is rare, but there is a significant impact on mobility and quality of life [1]. HEM is usually associated with portal-caval shunts and the patients with HEM survive longer due to medical advances, including liver transplantation [2-4]. We report a patient with Budd-Chiari syndrome and hepatitis B virus (HBV) infection, who developed symptoms and signs of HEM later in life.

Case Report

A 37-year-old woman was admitted to the hospital with a 20 days history of walking difficulties. The bilateral legs weakness progressed and created difficulties in rising from a chair and climbing stairs in the previous two years. There were no symptoms related to cranial nerve dysfunction, tremors, or myoclonus. During that period, she experienced several episodes of mild encephalopathy. She had been diagnosed with Budd-Chiari syndrome and chronic hepatitis B for more than 10 years. Liver biopsy in 2008 demonstrated evidence of cirrhosis, and by 2010 gastroscopy revealed grade I esophageal varices. No evidence of diabetes mellitus, hypertension, and

cardiopulmonary disease was found. Her past history was not significant for alcohol consumption. She denied bowel, bladder, or upper gastrointestinal bleeding. There was no family history of liver or neurological illnesses.

The patient had palmar erythema, spider angioma, and splenomegaly of about 2.0 cm on physical examination. K-F rings and skin hyperpigmentation were absent. She was alert, cooperative and had no speech disorder. Neurological examination revealed normal mental status and the presence of asterixis. Upper extremities strength was weak, especially for the left upper limb, with normal function. Upper extremity tendon reflexes were normal. Hoffman's sign was absent. In the lower extremities the tone was more noticeably increased with brisk deep tendon reflexes and there was clonus at the ankles. Plantar reflexes were extensor bilaterally. There was neither atrophy nor fasciculations. Both knee extensors and plantar flexors were normal in strength. Upper extremities sensation was without any deficits, but proprioception and vibration were mildly decreased in the toes, there was hyperesthesia to pinprick in the feet. The sensation of the trunk and crissum was normal. Gait was markedly spastic. There was no evidence of cerebellar dysfunction.

There was normocytic hypochromic anemia (hemoglobin: 102g/L). Hemogram disclosed thrombocytopenia (platelet: $32 \times 10^9/L$) and prothrombin time was prolonged (19.9 seconds). Stool was tested negative for occult blood. Liver function revealed raised serum total bilirubin ($68.09 \mu\text{mol/L}$), normal ALT, minimally raised AST (50U/L), reduced total protein (59.3g/L) and albumin (31.7g/L) and abnormal A: G ratio (1.15:1). Plasma ammonia was elevated to $131 \mu\text{mol/L}$. Renal function and serum electrolytes were normal. The patient's electrocardiogram was normal. Serum antibodies to HTLV-1, HIV, and HCV were absent. Markers of HBV infection assay were positive. The level of HBV-DNA was not detectable. Blood Vitamin B1, Vitamin B12, folate values, thyroid functions, and serum hexoseaminidase levels were in normal ranges. The upper endoscopy revealed grade II esophageal varices. CSF contained normal levels of glucose, proteins, and cell counts. Immunological tests of CSF for syphilis, tuberculosis were negative. MRI of the spinal cord and brain showed no intrinsic abnormality apart from bilateral symmetric pallidal hyperintensity. 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. Electroencephalogram showed normal background activity without triphasic waves or paroxysmal activity. Abdominal CT revealed a cirrhotic liver, thrombosis of the portal vein, and extensive collateralization and varices.

After 3 weeks of conservative treatment with protein restriction, physiotherapy, and oral lactulose, the patient's mobility improved, with decreased spasticity but without significant changes in the neurological examination follow up though the asterixis was absent at this time. Plasma ammonia level decreased to $57 \mu\text{mol/L}$. The patient returned home following discharge.

Discussion

Hepatic myelopathy (HM) is commonly found in combination with hepatic encephalopathy (HE). At present, the pathophysiology of HEM remains poorly understood. The diagnosis of HEM has to be established on clinical grounds after exclusion of other possible pathologies [5]. It has been hypothesized that ammonia and other neurotoxins may bypass the liver through spontaneous or surgical porto-systemic shunts. Porto-systemic shunts were found with radiological investigation in this patient. Neuropathological studies

demonstrated selective demyelination of the lateral corticospinal tracts with varying degrees of axonal loss. These lesions were seen especially within the cervical levels of the spinal cord but occasionally within the brainstem with no involvement of other tracts [2,3,6-7].

HEM caused by Budd-Chiari syndrome and HBV-related cirrhosis is uncommon. The patient developed symptoms within two years after diagnosis of cirrhosis. Moreover, there was no considerable worsening of neurological features for the last 2 years. We suspect that early onset of encephalopathy and myelopathy may be caused by significant shunting as a result of several factors: cirrhosis and spontaneous shunting, portal-vein occlusion, and cavernoma leading to anastomosis. There are two factors involved in the cirrhosis including vascular disease and liver cell damage. Portacaval shunting plays a substantial role in the pathogenesis of HEM. Spontaneous shunting occurs within the liver as a compensatory mechanism. The patient had cirrhosis at the time of presentation and additional spontaneous portacaval shunting. Computer tomography (CT) scan confirmed the functioning of the portal-vein shunt and a twisted splenic vein. In addition, hyperammonemia was detected. The rare complication needs to be considered in patient as spontaneous portacaval shunting for relieving portal hypertension. However, whether it will provide insight regarding pathophysiological mechanism needs to be explored [1,8,9-10].

Spastic paraparesis caused by HM has been shown to be refractory to medical treatment. In most patients, therapies aiming at reducing nitrogen absorption or lowering plasma ammonia levels have been generally unrewarding, which is similar to the therapeutic schedule of HE. Symptomatic treatment and administration have been demonstrated to temporarily benefit HEM symptoms. Protein restriction, oral neomycin, lactulose, and colonic exclusion have failed to produce significant improvement. At best, these measures prevented or decreased the numbers of encephalopathic episodes. Even so, progressive paraparesis was observed even when encephalopathy was effectively averted [1,8,11-12]. The improvement in spastic paraparesis was reported after liver transplantation in recent studies.

References

1. Campellone JV, Lacomis D, Giuliani MJ, Kroboth FJ (1996) Hepatic myelopathy. Case report with review of the literature. *Clin Neurol Neurosurg* 98: 242-246.

2. Utku U, Asil T, Balci K, Uzunca I, Celik Y (2005) Hepatic myelopathy with spastic paraparesis. *Clin Neurol Neurosurg* 107: 514-516.
3. Demirci M, Tan E, Elibol B, Gedikoğlu G, Saribaş O (1992) Spastic paraparesis associated with portal-systemic venous shunting due to congenital hepatic fibrosis. *Neurology* 42: 983-985.
4. Counsell C, Warlow C (1996) Failure of presumed hepatic myelopathy to improve after liver transplantation. *J Neurol Neurosurg Psychiatry* 60: 590.
5. Qu B, Liu C, Guo L, Yang Y, Li JH, Yu L, Lv Y (2009) The role of liver transplantation in the treatment of hepatic myelopathy: case report with review of the literature. *Transplant Proc* 41: 1987-1989.
6. Troisi R, Debruyne J, de Hemptinne B (1999) Improvement of hepatic myelopathy after liver transplantation. *N Engl J Med* 340: 151.
7. Mendoza G, Marti-Fàbregas J, Kulisevsky J, Escartin A (1994) Hepatic myelopathy: a rare complication of portacaval shunt. *Eur Neurol* 34: 209-212.
8. Robinson CE, Anzarut A (1978) Myelopathy in cirrhosis. *Paraplegia* 16: 3-4.
9. Panicker J, Sinha S, Taly AB, Ravishankar S, Arunodaya GR (2006) Hepatic myelopathy: a rare complication following extrahepatic portal vein occlusion and lienorenal shunt. *Neurol India* 54: 298-300.
10. Wang MQ, Dake MD, Cui ZP, Wang ZQ, Gao YA (2001) Portal-systemic myelopathy after transjugular intrahepatic portosystemic shunt creation: Report of four cases. *J Vasc Interv Radiol* 12: 879-881
11. Rab SM, Hussain SM (1985) Spastic paraparesis with cirrhosis of liver. *Br J Clin Pract* 39: 244-6.
12. Bechar M, Freud M, Kott E, Kott I, Kravvic H, Stern J, Sandbank U, Bornstein B (1970) Hepatic cirrhosis with post-shunt myelopathy. *J Neurol Sci* 11: 101-107.

Corresponding author

Qiu-Lin Sun
Department of Infectious Diseases
the First Affiliated Hospital of Anhui Medical University
230022, 218 Jixi Road, Hefei
Anhui Province, China
E-mail: sql0815@126.com.

Conflict of interests: No conflict of interests is declared.