ISSN: 2754-4931

Journal of Gastroenterology & Hepatology Reports

SCIENTIFIC Research and Community

Case Report

Open Access

A Case of Acquired Hepato-Cerebral Degeneration after Splenorenal Shunt from Ethiopia

Abdu Mohammed[1]*, AbateBane[2], Getahun Mengistu[3] and Fekadu Ayalew[4]

[1]Department of Internal Medicine, Gastroenterology and Hepatology, Adera Medical center, Addis Ababa, Ethiopia

[2]Department of Internal Medicine, Gastroenterology and Hepatology, CHS, Addis Ababa University, Addis Ababa, Ethiopia

[3]Department of Neurology, CHS, Addis Ababa University, Addis Ababa, Ethiopia

[4]Department of Radiology, Adera Medical center, Addis Ababa, Ethiopia

ABSTRACT

Acquired hepato-cerebral degeneration (AHD) is a neurologic syndrome caused by liver dysfunction and long-standing portosystemic shunting. There is scarce and conflicting information regarding the clinical course of AHD. We report the case of a 25-year-old male who underwent splenectomy and splenorenal shunt for recurrent UGIB 20 to Esophageal varices 20 to Non-Cirrhotic portal hypertension 20 to Non-Cirrhotic portal fibrosis and hypersplenism, in Mumbai, India. He presented with slow movement, hypophonia, gait instability, and rigidity of the lower extremities. In this paper, we discuss the patient's case with literature review.

***Corresponding author**

Abdu Mohammed, Department of Internal Medicine, Gastroenterology and Hepatology, Adera Medical center, Addis Ababa, Ethiopia.
E-mail: abdumohammed7@gmail.com

Received: January 06, 2022; **Accepted:** January 12, 2022; **Published:** January 17, 2022

Keywords: Acquired Hepato-Cerebral Degeneration, Non-Cirrhotic Portal Hypertension, Non-Cirrhotic Portal Fibrosis, Ethiopia

Abbreviations
AHD: Acquired Hepato-Cerebral Degeneration
BG: Basal Ganglia.
CAHD: chronic acquired Hepato-cerebral Degeneration
HE: Hepatic Encephalopathy
Mn: Manganese
MRI: Magnetic Resonance Imaging
NCPF: Non-Cirrhotic Portal Fibrosis
NCPH: Non-Cirrhotic Portal Hypertension
PI: Pallidal Hyperintensity
TIPSS: Transjugular Intrahepatic Portosystemic Shunts.

Introduction

Acquired hepato-cerebral degeneration (AHD) is a neurologic syndrome characterized by parkinsonism, ataxia, or other movement disorders and by neuropsychiatric and cognitive manifestations in patients with chronic liver disease, especially those who develop portosystemic shunting spontaneously or induced surgically [1,2]. Patients typically present with dysarthria, ataxia, tremor, involuntary movements, and altered mental status, and often do not respond to conventional medical therapy for hepatic encephalopathy. It is often under-recognized as a cause of cognitive impairment in patients with liver disease. Recently, its pathogenesis has been associated with metal accumulation in the basal ganglia, mainly manganese, increasing the interest of clinicians and researchers towards the condition.T1-weighted magnetic resonance imaging (MRI) typically shows bilateral pallidal hyperintensities [3,4]. We report a case of a patient with clinical and radiological characteristics of AHD, who presented with a movement disorder resulting from manganese accumulation in the basal ganglia.

Case Report

A.T.(MRN- 58469) is a 25- year-old male who has a history Non-Cirrhotic Portal Hypertension (NCPH) 20 to Non-Cirrhotic Portal Fibrosis (NCPF). He suffered from recurrent episodes of UGIB 20 to esophageal varices and hypersplenism. He had repeated sessions of EVL done at Adera Medical center in Addis Ababa, followed by secondary prophylaxis with non-selective beta-blockers. In 2018, 3 years back, he had a splenectomy and splenorenal shunt at Fortis Hospital in Mumbai, India. However, after the procedure, he developed splenic vein thrombosis and was started with anticoagulant treatment.

Recently he presented to our medical center with a complaint of trouble in keeping his balance, slowness of movement, and hypophonia of 1 and half years duration. A few months later, he noticed that he had difficulty positioning himself in bed, keeping his balance, and was unable to walk without support. At presentation, on physical examination, he was noted to have

hypophonia, gait instability, and cogwheel rigidity of the lower extremities.

Investigation showed elevated serum ammonia (183 µmol/L – reference range: 16-60 µmol/L). Magnetic resonance imaging (MRI) of the brain showed symmetrical hyperintensity on T1 weighted images in the globus pallidus and crura of the midbrain bilaterally. Similar symmetrical hyperintensity is also seen on FLAIR images in the Crura of the midbrain bilaterally (Figure1)

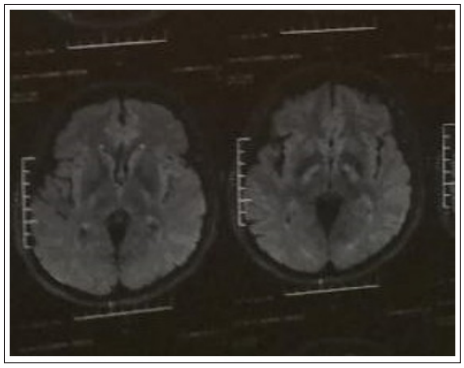

Figure 1: Magnetic resonance imaging (MRI) of the brain showed a symmetrical hyperintensity on T1 weighted images in the globus pallidus and crura of the midbrain bilaterally,at Adera Medical Center, Addis Ababa, 2021

This MRI Scan also revealed heavy metal deposition in bilateral basal ganglia and midbrain, consistent with manganese deposition probably as a result of a portosystemic shunt.

CT scan done, in March 2021, showed normal liver parenchyma with patent splenic, portal vein without any intraluminal filling defect suggesting thrombus or wall thickening. The left branch of the portal vein was larger compared to the right (Figure 2).

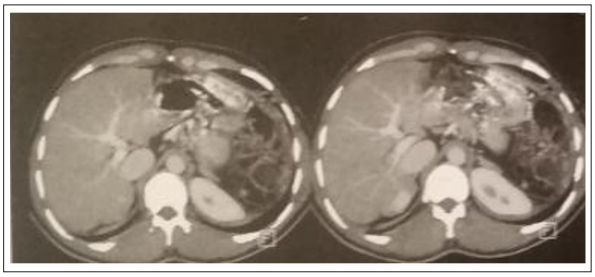

Figure 2: CT scan showing normal liver parenchyma And a large left branch of the portal vein as compaired to the right, at Adera Medical Center, Addis Ababa, 2021

MR Elastography showed the mean shear stiffness value of the liver to be 4.4 kpa (Figure 3).

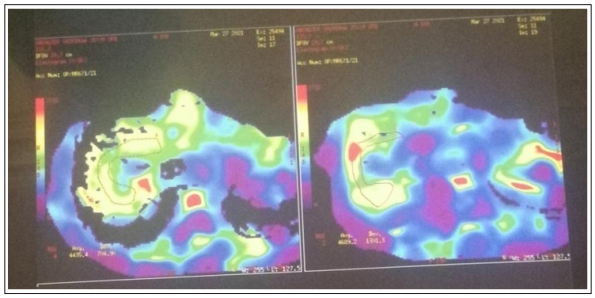

Figure 3: MR Elastography of the patient with 4.4kpa mean shear stiffness value of the liver at Adera Medical Center, Addis Ababa, 2021

Recent liver biochemical test and CBC was normal. Abdominal ultrasonography with Color Doppler demonstrated normal liver echo pattern, size (14.3cm), and smooth contour. The portal vein had normal caliber (0.8cm). Hepatic veins and the intrahepatic portion of IVC had normal diameters &flow. The spleen was not seen.

After an evaluation by a neurologist, a diagnosis of acquired hepato-cerebral degeneration (AHD) was made, and he was put on Levodopa and carbidopa. The patient was also started on ammonia-decreasing therapy (lactulose, L-Ornithine L-Aspartate granules) with diet manipulation (recommended diet rich in branched-chain amino acids). He also continued treatment with warfarin. However, there was no clinical improvement noted in this patient.

Discussion

Acquired hepato-cerebral degeneration is an exceptional type of hepatic encephalopathy which was first described by van Woerkom in 1914, but remained unrecognized until 1965 when Victor et al. p published their observations [5,6]. AHD has been described in patients with severe liver disease of many causes, and notably in patients with surgically or spontaneously induced portosystemic shunts. It is a rare syndrome. It's frequency and prevalence remain largely uncertain. In some reports, AHD occurs in 0.8-2 % of patients with cirrhosis [7,8]. There are also reports of AHD after splenorenal shunt developing eight years later [8].

The clinical picture of AHD includes neuropsychiatric movement disorders including ataxia, tremor, chorea, dysarthria, parkinsonism, dystonia, and myoclonus [6]. In this case, the clinical features of acquired hepato-cerebral degeneration started after 1 and half years of splenorenal shunt procedure done for recurrent esophageal variceal bleeding.

The pathogenesis of AHD remains unclear but metal intoxication seems to play a role in the disease. Brain manganese (mn) overload may contribute to AHD outcome [3,4,11-13]. Some studies suggest that manganese plays a major role in the development of the disease [4,12-13]. The hepatobiliary system clears manganese from both blood and cerebrospinal fluid and in some patients manganese concentrations are higher than would be expected. Therefore, the toxic substances that are not removed by the hepatobiliary system due to portosystemic shunts and liver dysfunction enter into the systemic circulation [14]. As a result, manganese deposition in the brain (especially in the basal ganglia, brainstem, cerebral cortex, and the surrounding white matter) is thought to induce neuronal loss and it is the major determinant of basal ganglia dysfunction, leading to the predominantly extrapyramidal central nervous system symptoms of cirrhosis [11].

The diagnostic modalities used to confirm or refute the diagnosis of AHD involve a combination of clinical examination and neuroimaging [4]. In chronic AHD specifically, MRI often shows basal ganglia (BG) changes with hyperintense signal changes on T1-weighted imaging mainly in the pallidum [9,16-17]. It is believed that the high signal intensity on T1 is due to the rise in manganese concentration within the CNS, with preferential deposition in the globus pallidus [11]. Blood tests can be less helpful in the diagnosis of Chronic AHD(CAHD). Blood manganese levels can vary in patients with CAHD thus, are not diagnostic. Levels were significantly higher in patients with a previous portocaval anastomosis or transjugular intrahepatic portosystemic shunt (TIPS) suggesting portosystemic shunting of manganese may lead to increased blood levels and ultimately to deposition in the brain with potentially deleterious effects [4].

In our patient, AHD resulted from the splenorenal shunt bypassing the detoxication effect of the liver.

A study done in 2004 showed that there was a significant correlation between blood manganese levels with pallidal hyperintensity (PI) seen on MRI, however, no correlations between blood manganese level and liver synthetic function or neurological functions were seen. In addition, there was no correlation between PI and neurological function. The importance of shunting, however, was highlighted by the fact that most patients with chronic acquired hepato-cerebral degeneration (CAHD) either have advanced liver disease with established portal hypertension and/or surgical shunts, TIPS, or spontaneous portosystemic shunts [18]. Even if diagnosed, the treatment options for CAHD are limited. Trials of dopamine agonists because of the Parkinson disease-like symptoms may be initiated, however, most patients do not respond to this, as was the case with our patient [4,19]. There have been reports of improvement in symptoms with Rifaximin, a non-absorbable antibiotic used for the treatment of hepatic encephalopathy, however, these studies are limited to small case series(20).And in our case no feature of hepatic encephalopathy.

With the link between portosystemic shunting and CAHD, investigation for shunts is warranted with reported benefits in the obliteration of shunts [21]. Liver transplantation has been reported to be effective for some patients with CAHD in small case series [22-26]. However, many patients present with advanced neurological symptoms, which often leads to exclusion from transplantation candidacy.

The formation of portosystemic shunts may also aggravate the course of the disease by promoting increased shunting of manganese to the systemic circulation and the brain and thus shunts should be sought after and considered for closure if clinically appropriate. In our patient's case, treatment using both ammonia-lowering therapies (lactulose, L-Ornithine L-Aspartate granules) and diet manipulation (a diet rich in branched-chain amino acids) were tried with no improvement.

Conclusion

In summary, CAHD is a rare complication occurring in cirrhotic patients with a portosystemic shunt, which develops either spontaneously or after surgery. According to studies, brain manganese overload plays a major role in the development of AHD. Which leads to debilitating neurological symptoms not readily amenable to pharmacological treatments. Increased awareness of CAHD will hopefully lead to increased recognition and ideally to the development of better therapeutic options for such a rare and challenging clinical disease.

References

1. Jog MS, Lang AE (1995) Chronic acquired hepatocerebral degeneration: case reports and new insights. Mov Disorder 10: 714-722.
2. Ferrara J, Jankovic J (2009) Acquired hepatocerebral degeneration. J Neurol 256: 320-332.
3. Butterworth RF, Spahr L, Fontaine S, Layrargues GP (1995) Manganese toxicity, dopaminergic dysfunction and hepatic encephalopathy. Metab Brain Dis 10: 259-267.
4. N Rajoriya, M Brahmania, Jordan J Feld (2019) "Implications of Manganese in Chronic Acquired Hepatocerebral Degeneration" Ann hepatology 1: 274-227.
5. Van Woerkem W (1914) La cirrhose hepatique avec alterations dans les centresnerveuxevoluant chez des sujetsd'agemoyen. Nouvelle Iconographie de la Salpetriere, Clinique des Maladies du SystemeNerveux 7: 41-45.
6. Victor M, Adams RD, Cole M (1965) The acquired (non-Wilsonian) type of chronic hepatocerebral. Medicine 44: 345-396.
7. Fernández-Rodriguez R, Contreras A, De Villoria JG, Grandas F (2010) Acquired hepatocerebral degeneration: clinical characteristics and MRI findings. Eur J Neurol 17: 1463-1470.
8. A FBlease LR, C Waugh, GW Mccaughan et al. (1989) "Development of chronic hepatocerebral degeneration eight years after a distal splenorenal (Warren) shunt : Gut 30: 1419-1423
9. Ferrara J, Jankovic J (2009) Acquired hepatocerebral degeneration. J Neurol 256: 320-332.
10. Layrargues GP (2001) Movement dysfunction and hepatic encephalopathy. Metab Brain Dis 16: 27-35.
11. Burkhard PR, Delavelle J, Du Pasquier R, Spahr L (2003) Chronic parkinsonism associated with cirrhosis: a distinct subset of acquired hepatocerebral degeneration. Arch Neurol 60: 521-528.
12. Huang CC, Chu NS, Lu CS, Chen RS, Schulzer M, et al. (2007) The natural history of neurological manganism over 18 years. Parkinsonism RelatDisord 13: 143-145.
13. Rose C, Butterworth RF, Zayed J et al. (1999) Manganese deposition in basal ganglia structures results from both portal-systemic shunting and liver dysfunction. Gastroenterology 117: 640-604.
14. Rovira A, Alonso J, Cordoba J (2008) MR Imaging findings in hepatic encephalopathy. Am J Neuroradiol 29: 1612-162.
15. Spahr L, Butterworth RF, Fontaine S et al. (1996) Increased Blood Manganese in Cirrhotic Patients: Relationship to Pallidal Magnetic Resonance Signal Hyperintensity and Neurological Symptoms. Hepatology 24: 1116-1120
16. Lee J, Lacomis D, Comu S, Jacobsohn J, Kanal E (1998) Acquired hepatocerebral degeneration: MR and pathologic findings. Am J Neuroradiol 19: 485-785.
17. Shulman LM, Minagar A, Weiner WJ (2003) Reversal of parkinsonism following liver transplantation. Neurology 60: 519.
18. Crossgrove J, Zheng W (2004) Manganese toxicity upon overexposure. NMR Biomed 17: 544-553.
19. Tuschl K, Mills PB, Parsons H, Malone M, Fowler D et al. (2008) Hepatic cirrhosis, dystonia, polycythaemia and hypermanganesaemia-a new metabolic disorder. J Inherit Metab Dis 2008; 31: 151-63.
20. Kok B, Foxton MR, Clough C, Shawcross DL (2013) Rifaximin is an efficacious treatment for the parkinsonian phenotype of hepatic encephalopathy. Hepatology 58: 1516-1517.
21. Hisahara S, Matsushita T, Kitamura M, Mezawa S, Nonaka M et al. (2014) Long-term clinical and radiological improvement of chronic acquired hepatocerebral degeneration after obliteration of portosystemic shunt: Report of a case. J Neurol Sci 346: 303-306.
22. Powell EE, Pender MP, Chalk JB, Parkin PJ, Strong R et al. (1990) Improvement in chronic hepatocerebral degeneration following liver transplantation. Gastroenterology 98: 1079-1082.
23. Servin-Abad L, Tzakis A, Schiff ER, Regev A (2006) Acquired hepatocerebral degeneration in a patient with HCV cirrhosis: complete resolution with subsequent recurrence after liver transplantation. Liver Transpl 12: 1161-1165.
24. Parkes JD, Murray-Lyon IM, Williams RO (1970) Neuropsychiatric and electroencephalographic changes after

transplantation of the liver. QJ Med 39: 515-527.

25. Stracciari A, Guarino M, Pazzaglia P, Marchesini G, Pisi P (2001) Acquired hepatocerebral degeneration: full recovery after liver transplantation. J Neurol NeurosurgPsychiarty 70: 136-137.

26. Wijdicks EF, Wiesner RH (2003) Acquired (non-Wilsonian) hepatocerebral degeneration: complex management decisions. Liver Transpl 9: 993-994.