BJMS
Borneo Journal of Medical Sciences

CASE REPORT

Non-Cirrhotic Portal Hypertension: A Case of Bleeding Gastro-Oesophageal Varices with Non-Cirrhotic Liver Reported in East Malaysia

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Received: 2 July 2018

Accepted: 30 October 2018

Keywords: non-cirrhotic portal hypertension, normal liver, variceal bleeding

ABSTRACT

Non-cirrhotic portal hypertension (NCPH) is clinically defined as the presence of portal hypertension in the background of non-cirrhotic liver. It is diagnosed by the findings in ultrasound of the hepatobiliary system and also oesophagogastroduodenoscopy (OGDS) that consistent with that of a portal hypertension, but otherwise has a relatively normal liver function and echotexture. The treatment mainly focuses on primary and secondary prophylaxis of variceal bleeding both pharmacologically like non-selective beta-blockers and octreotide, and non-pharmacologically like endoscopic band ligation of varices and sclerotherapy. In advance cases, sometimes surgery such as Porto-systemic shunt or splenectomy may be required especially in patients with uncontrolled variceal bleeding or with symptomatic hypersplenism. Here we report a case of a young man who presented with upper gastro-intestinal bleeding, which was initially thought from a bleeding ulcer but was found to be secondary to oesophageal and gastro-oesophageal varices. Apart from having mild ascites, he has no other features of portal hypertension. His liver biochemistry and echotexture were also normal. Unfortunately, the patient was lost to follow up while he was still in the early stage of investigating the condition. The purpose of this case report is to share an uncommon occurrence of NCPH in East Malaysia, where liver cirrhosis predominates the aetiology of portal hypertension. Also, to the best of our knowledge, there is a very limited reporting of a similar case in this region.

INTRODUCTION

Portal hypertension often developed in the background of liver cirrhosis. It is the end result of multiple conditions associated with increased resistance in portal circulation and ultimately leads to life-threatening complications such as variceal bleeding, splenomegaly and ascites[1]. However, portal hypertension can occur in a non-cirrhotic liver in a condition that can be broadly grouped as Non-Cirrhotic Portal Hypertension (NCPH). NCPH frequently associated with a wide range of vascular changes in the portal system. Unlike those due to liver cirrhosis, it is associated with normal or mildly elevated hepatic venous pressure gradient[2]. The leading conditions of NCPH are classified anatomically as pre-hepatic, hepatic, and post-hepatic[3] according to the site of resistance in portal circulation. For the diagnosis of NCPH, it is essential to rule out other causes of chronic liver diseases such as viral hepatitis B and C, chronic alcoholism, non-alcoholic fatty liver disease, autoimmune hepatitis, hereditary causes like hereditary hemochromatosis and Wilson's disease, and also drug induced causes as well. The principal of management is aimed to prevent and treat complications which expected to occur from portal hypertension. Therefore, prompt diagnosis of the disorder is very important for initiating the correct management.

CASE PRESENTATION

A 25-year-old man with no known medical illness presented with progressive abdominal pain and distention, and passing blackish stool for the past 2 weeks prior to admission, which was accompanied by symptoms of anaemia. He also had reduced appetite and loss of weight for the past one year. Otherwise, he has neither history of haematemesis nor any other bleeding elsewhere, and he denied having any fever, altered bowel habit, night sweats or jaundice. There is no significant family history. On examination, patient was well and comfortable. He was cachectic and pale, and there was mild ascites present. Otherwise, there were no jaundice, no lymphadenopathy, no pedal oedema, and no stigmata of chronic liver disease. The abdomen was soft and not tender, and there was no palpable mass or organomegaly felt. As for his investigations, his haemoglobin was 3.7g/dl, microcytic and hypochromic picture and his reticulocyte count was 4.2%. The platelet count was normal and the coagulation time was not prolonged. His renal and liver function test (Total Bilirubin: 5.9umol/L, Alanine Aminotransferase (ALT): 12u/L, Alkaline Phosphatase: 94u/L) was normal, and his albumin: globulin ratio was reduced (albumin: 20 g/L, globulin: 32g/L). There was no cardiomegaly but the right costophrenic angle was blunted on radiographic imaging of the chest. Echocardiography was done and it was normal. Subsequently he was subjected to Oesophagogastroduodenoscopy (OGDS) and it showed Grade I Oesophageal Varices (Figure 1) and Gastro-oesophageal Varices grade 2 with Stigmata of Recent Haemorrhage (Figure 2). Endoscopic sclerotherapy was performed to secure the bleeding.

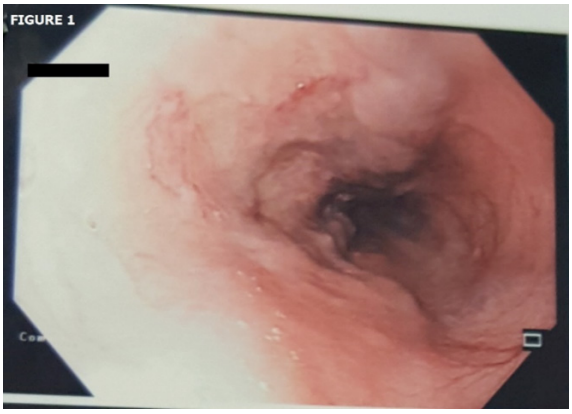

Figure 1 Oesophagogastroduodenoscopy shows Grade 1 Oesophageal Varices

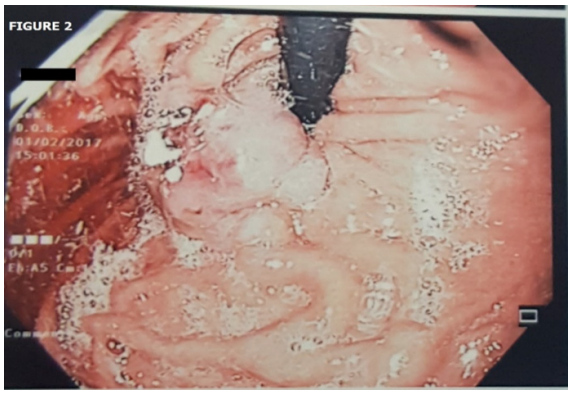

Figure 2 Oesophagogastroduodenoscopy shows Gastro-Oesophageal Varices with stigmata of recent bleed.

As it was initially thought the UGIB was due to bleeding ulcer, the patient was started on intravenous (IV) Pantoprazole 8 mg hourly infusion and planned for 72 hours. However, after the OGDS showed variceal bleeding, he was then given IV Terlipressin 2 mg bolus, then 1 mg 4 hourly for 24 hours. He was also given IV Ceftriaxone 1 gm once daily, and was started on oral Propranolol 20 mg twice daily thereafter. He was also transfused 4 pints of packed red cells. The repeated Hb was 8 g/dL. Preliminary investigations for liver cirrhosis such as HBsAg, Anti-HCV, HIV antibody, stool microscopy for Schistosoma egg and others were all negative. Ultrasound of the hepatobiliary system showed enlarged liver, dilated hepatic sinusoids and minimal ascites, but otherwise the liver parenchymal structure was normal (Figure 3), and the portal vein was patent on Doppler (Figure 4). Patient was consulted for liver biopsy. However, he defaulted and was lost to follow up.

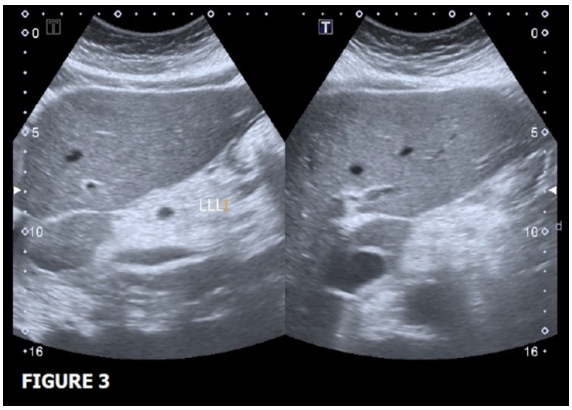

Figure 3 Ultrasound scan of the hepato-biliary system shows mild hepatomegaly.

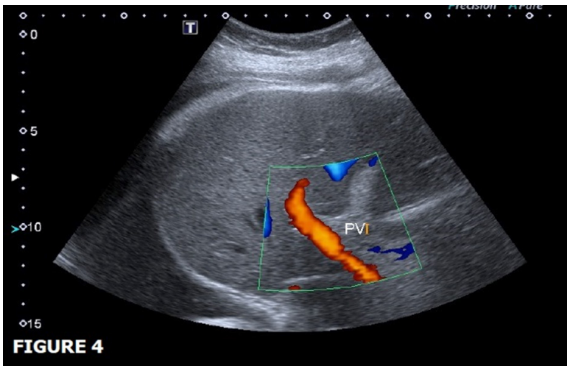

Figure 4 Doppler ultrasound scan of the portal vein shows normal patency of the portal vein.

DISCUSSION

Portal hypertension is a clinical condition defined by pathological increase in the pressure of the portal venous system with the gradient between portal vein and inferior vena cava more than 5 mmHg[1]. This usually occurs as a result of increased resistance in the portal circulation and ultimately lead to the formation of extensive network of portosystemic collaterals in multiple sites that divert portal blood to the systemic circulation and bypassing the liver[2]. In most cases, the resistance occurs within the liver (as in the case of liver cirrhosis). But there are some cases where it can also occur from pre-hepatic site (like portal vein thrombosis) or post-hepatic site (like Budd-Chiari syndrome). As portal hypertension progresses, splanchnic blood flow will increase as a result of local

release of humoral vasodilatory mediators, such as vascular endothelial growth factor, prostacyclin, nitric oxide and others that will cause splanchnic vasodilation and angiogenesis. These mediators will also affect the systemic circulation and lead to hypotension and vascular underfilling, which triggers the release of endogenous systemic vasoactive substance and plasma volume expander by renal sodium retention, and lead to increased cardiac output[2]. The haemodynamic changes in both splanchnic and systemic circulation will ultimately lead to the main complications of portal hypertension, like bleeding varices causing upper gastrointestinal bleeding (UGIB), ascites, hepatic encephalopathy, hepatorenal and hepatopulmonary syndrome[1,2]. All of these complications are essentially a sequalae of the abnormal collateral formation and hyperdynamic splanchnic and systemic circulations.

Liver cirrhosis is the most common cause of portal hypertension worldwide[2]. But portal hypertension can also develop in the background of normal functioning liver, in a group of condition collectively referred to as noncirrhotic portal hypertension (NCPH). In NCPH, the hepatic venous pressure gradient is often normal or only mildly elevated and usually lower than the portal venous pressure. The causes of NCPH are primarily vascular in origin and generally can be classified according to anatomical location of blood flow resistance, such as pre-hepatic, hepatic, and post-hepatic. The hepatic causes are further subdivided into pre-sinusoidal, sinusoidal and post-sinusoidal[3]. Although NCPH only account less than 10 per cent of cases in the Western world, it is the leading causes of portal hypertension in other parts of the world. NCPH is clinically defined by the presence of features of portal hypertension, for example like splenomegaly, varices and ascites with preserved liver functions and patent hepatic and portal veins. It is usually diagnosed by ultrasound of the hepatobiliary system and esophagogastroduodenoscopy (OGDS) findings consistent with portal hypertension, but normal liver function and histology.

NCPH is said to affect predominantly the low socio-economic population of the developing world. But it is also reported in higher strata population from all parts of the world[4]. Epidemiologically, it constitutes approximately 10 – 30% of patients with variceal bleed worldwide[5]. And it is thought that NCPH is largely under-diagnosed particularly from the parts of the world where it is rare. Overall NCPH is estimated to contribute 14% of portal hypertension in adults[6].

The management of NCPH are geared at preventing and treating the complication that may arise as a sequalae of portal hypertension, for example like variceal bleeding. Therefore, early recognition and prompt diagnosis of these conditions is very important so that the appropriate therapeutic measure which is to reverse the natural course of the disease or prophylactically to prevent disease progression can be initiated. As data are still very limited regarding the best approach to management of this particular condition, patients are usually treated in the manner similar to those of due to cirrhosis.

The prognosis is fortunately good whereby only minority of cases will develop recurrent attacks of uncontrolled bleeding, portal vein thrombosis, and also hepatopulmonary syndrome. Long-term survival of well-treated oesophagogastric varices and properly-timed shunt surgery is nearly 100 and 80%, respectively[7]. Liver functions will usually remain well preserved until very late, but 20 – 33% patients will develop parenchymal atrophy with subsequent decompensation. The commonest causes of death are bacterial infection (31%), followed by progressive liver failure (25%), uncontrolled variceal bleeding (17%), and intestinal infarction (8%)[8, 9].

In this article, we report a patient who presented with UGIB which at first was thought due to peptic ulcer disease. However, his OGDS found gastro-oesophageal varices. Apart from having mild ascites, he has no other signs of chronic liver disease. Upon further test noted the liver biochemistry and echotexture were normal. His portal vein was also patent. The next step of investigation for this patient is to do liver biopsy to confirm no cirrhotic changes over the liver and also to look for histological changes consistent with NCPH such as prominent sclerosed wall of portal vein and its branches, obliteration of small portal venules, also to find site of venous obstruction, if any. He was supposedly to be monitored regularly with Doppler ultrasound at six monthly intervals to check the patency of the portal vein, and also secondary prophylaxis for variceal bleeding with non-selective beta-blockers, but unfortunately, he defaulted follow up. Therefore, this case report suffers one limitation where the patient was lost to follow up before the diagnosis is known. However, the objective of this case report is to illustrate the uncommon occasion of UGIB secondary to portal hypertension in a relatively normal liver function and parenchymal structure.

CONCLUSION

In conclusion, this case report served to remind the reader that portal hypertension does not occur exclusively as a complication of liver cirrhosis, but it can also happen due to multiple other causes with the background of normal liver function and architecture.

ACKNOWLEDGEMENTS

The authors would like to thank the patient for his cooperation in relation to the writing of this case report. The author would also like to thank the Director General of Health Malaysia for his permission to publish this article.

CONFLICT OF INTEREST

The authors declare that they have no competing interests in publishing this case.

CONSENTS

Written informed consent was obtained from the patient to publish the case. A copy of written consent is available for review by the Chief Editor.

REFERENCES

1. Sanyal AJ, Bosch J, Blei A et al. (2008). Portal hypertension and its complications. Gastroenterology 134: 1715 – 1728.
2. García-Pagán JC, Gracia-Sancho J, Bosch J. (2012). Functional aspects on the pathophysiology of portal hypertension in cirrhosis. J Hepatol 57: 458.
3. Sarin SK, Khanna R. (2014). Non-cirrhotic portal hypertension. Clinics in liver disease 18: 451 –476.
4. Siramolpiwat S, Seijo S, Miquel R et al. (2014). Idiopathic portal hypertension: natural history and long-term outcome. Hepatology 59: 2276 – 2285.
5. Schouten JNL, Nevens F, Hansen B et al. (2012). Idiopathic noncirrhotic portal hypertension is associated with poor survival: results of a long-term cohort study. Aliment Pharmacol Ther 35: 1424 – 1433.
6. Goel A, Madhu K, Zachariah U et al. (2013). A study of aetiology of portal hypertension in adults (including the elderly) at a tertiary centre in southern India. Indian J Med Res 13: 922 – 927.
7. Sarin SK, Kumar A, Chawla YK et al. (2007). Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment. Hepatol Int 1: 398 – 413.
8. Okuda, K. (2002). Non-cirrhotic portal hypertension versus idiopathic portal hypertension. Journal of Gastroenterology and Hepatology 17.
9. Murai Y, Ohfuji S, Fukushima W et al. (2012). Prognostic factors in patients with idiopathic portal hypertension: two Japanese nationwide epidemiological surveys in 1999 and 2005. Hepatol Res 42: 1211 – 1120.