A Case of Idiopathic Non-Cirrhotic Portal Fibrosis

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Abstract

"Idiopathic non-cirrhotic portal hypertension" is a rare diagnosis characterised by increased intrahepatic portal pressures in the absence of underlying liver disease and caused by an unexplained aetiology. We present a one-of-a-kind "case of a 16-year-old male" with recurrent variceal bleeding who had an "elevated hepatic venous pressure gradient" and "portal hypertension sequelae" without underlying liver illness. There is limited treatment available because management is mostly geared at preventing illness consequences. This example emphasises the importance of future investigation into this disease entity and its pathophysiology.

Keywords: "Non-cirrhotic portal hypertension, Hepatic Venous Wedge Pressure, Variceal Bleeding".

Introduction

"Idiopathic non-cirrhotic portal hypertension (INCPH)" is an exclusion diagnosis characterised by increased "portal venous pressure" in the absence of "cirrhosis, hepatoportal flow blockage, splanchnic venous thrombosis, or other causes of liver disease". [1] "INCPH is responsible for 3%–5% of portal hypertension (PH) cases and 14%–27% of non-cirrhotic PH cases. It primarily affects males with a median age of 40 years in Western cultures; a higher occurrence at a younger age is reported in Eastern countries, which is thought to be attributable to socioeconomic disadvantages and bad living conditions".

INCPH manifests as PH consequences such as “variceal haemorrhage, ascites, portal vein thrombosis, and hepatic encephalopathy”. Without appropriate diagnostic testing, the underlying aetiology is unknown. To rule out underlying liver disease, a thorough workup is recommended, including “laboratory testing, hepatic imaging scans, and a liver biopsy”.

Case Report

A 16-year-old guy arrived with episodes of gastrointestinal haemorrhage. A long-standing bulk in the “left upper quadrant (splenomegaly)”, anaemia, and the “consequences of hypersplenism”. To come to a diagnosis of NCPF we should rule out all other causes of “Portal Hypertension and Splenomegaly”. For which certain investigations to be done. They are CE – CT ABDOMEN to rule out Cirrhosis of Liver, Viral serology for infectious Cause, Sr.Ferritin ,Sr. Cerruloplasmin for Inheritable Liver Disease, TB Qunatiferon for TB Spleen , ANA , LDH , ESR and CRP for Auto immune Diseases, IgG levels for Hodgkins Lymphoma, USG Hepatic Doppler for Portal vein size and Pathologies and finally Liver Biopsy which is Confirmatory for NCPF. A big and abnormally large spleen is the most common cause of portal hypertension in NCPF.

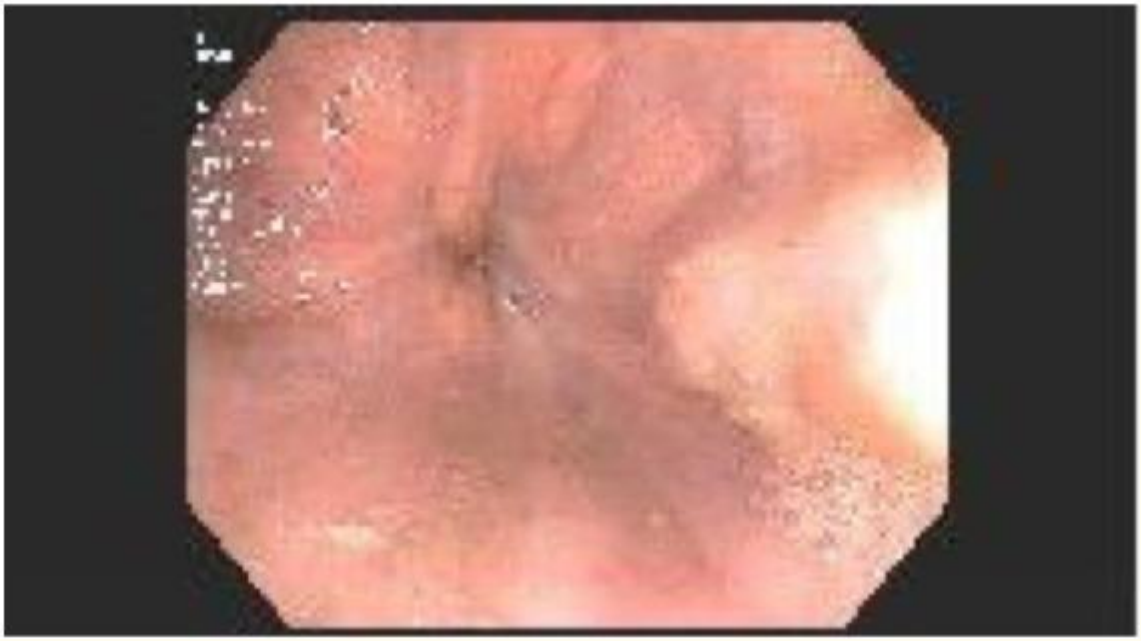

Figure 1. "Enteroscopy reveals grade 1 esophageal varices".

"Hepatic serologies" for the aforementioned underlying illnesses, such as "primary biliary cirrhosis, autoimmune and chronic hepatitis, Wilson's disease, and hemochromatosis", were all negative. The "inferior vena cava, portal vein, and right and middle hepatic veins" were all found to be patent in the "liver and portal vein vascular duplex".

The patient received a transjugular liver biopsy after being diagnosed with probable cirrhosis with no definite aetiology. "The hepatic venous wedge pressure was 21 mm Hg, the free hepatic venous pressure was 7 mm Hg, and the hepatic venous pressure gradient was 15 mm Hg, all of which indicated PH. A liver biopsy revealed normal liver tissue, with no signs of congestive hepatitis or cirrhosis" (Figure 2). Patient developed recurrent episodes of Variceal Bleeding. Patient referred to higher centre for Splenectomy where splenectomy was done and Patient improved Symptomatically.

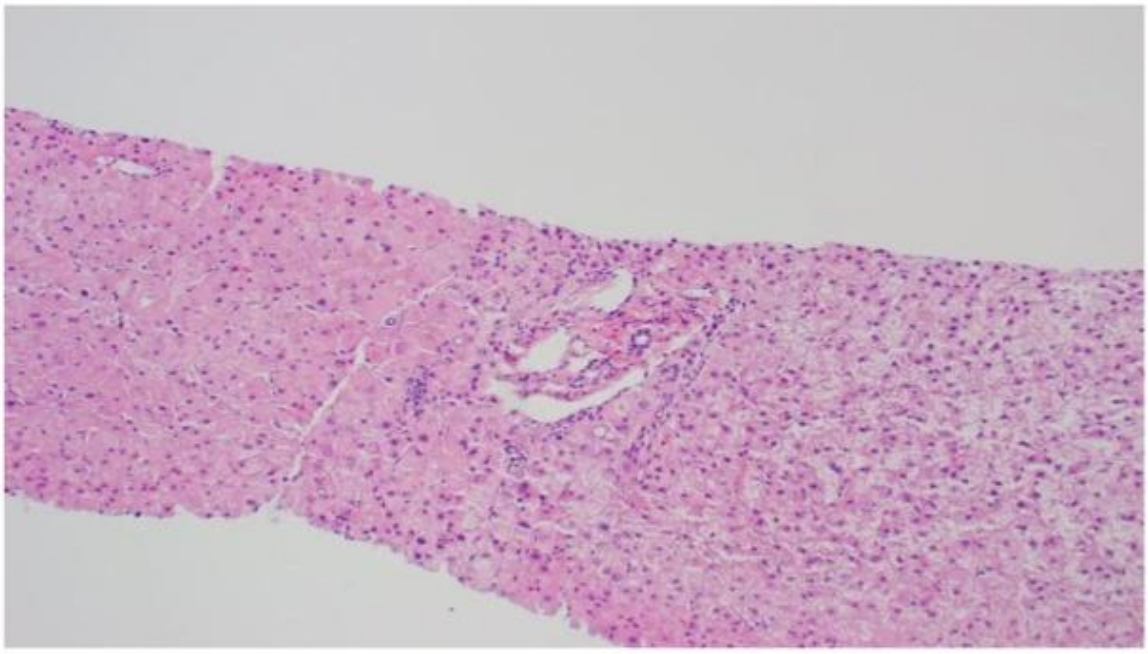

Figure 2. "Liver biopsy with normal liver tissue, no evidence of congestive hepatopathy or cirrhosis".

Discussion

The aetiology of INCPH is unknown; similar to our patient, the most common presenting sign of INCPH is stomach or esophageal variceal haemorrhage with maintained liver function. The prognosis of variceal haemorrhage is improved due to normal liver function; acute encephalopathy is a rare consequence. [2] "Splenomegaly and ascites are present in approximately 95% and 50% of patients, respectively, and are associated with a poor prognosis. Portal vein thrombosis is relatively prevalent, accounting for 13% to 46% of cases".[1-3]

INCPH has no well accepted diagnostic criteria, is underdiagnosed, and is frequently misinterpreted as cirrhosis. [1,2] “Liver function tests are normally normal, hepatic and portal veins are unobstructed, and the hepatic venous pressure gradient is elevated”. [3] As a result of hypersplenism, laboratory testing may reveal anaemia and thrombocytopenia.6 Serology and liver biopsy must be used to rule out “viral hepatitis, alcoholic and nonalcoholic steatohepatitis, autoimmune hepatitis, hemochromatosis, Wilson's disease, and primary biliary cirrhosis”. [1,2] The goal of management is to prevent illness consequences. The liver transplant is postponed until the disease worsens.

Conclusion

INCPH is a challenging condition to diagnose and treat. This condition is poorly understood; only a few studies have looked at the development, diagnostics, and therapy of INCPH. This instance raises awareness of a rare disease entity and highlights the need for additional research to avert disastrous consequences.

References

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