International Journal of Advances in Medicine
Chanda R et al. Int J Adv Med. 2023 Sep;10(9):670-672
http://www.ijmedicine.com
pISSN 2349-3925 | eISSN 2349-3933
DOI: https://dx.doi.org/10.18203/2349-3933.ijam20232554

Case Report

Face says it all: a curious case of non-cirrhotic portal fibrosis

Abhishek Chanda*, Mayurbahan Mukherjee, Mainak Mandal, Mohammad Saddam Hossain, Nirmalya Roy

Department of Medicine, KPCMCH, Kolkata, West Bengal, India

Received: 25 May 2023
Accepted: 14 August 2023

***Correspondence:**
Dr. Abhishek Chanda,
E-mail: abhishekchandabkcpc@gmail.com

ABSTRACT

Non cirrhotic portal fibrosis is a disease of obscure etiology and it causes portal hypertension, massive splenomegaly and well tolerated episodes of variceal bleeding young adults due to obliterative portovenopathy in young adults from low socioeconomic backgrounds, having normal or close to normal hepatic functions. It is also called as idiopathic portal fibrosis, though the etiology of NCPF is unclear but thought to be from chronic infections, exposure to medications and toxins, thrombophilia, immunological disorders are genetic factors. In India it accounts for 30 percent of variceal bleeds in non-cirrhotic portal fibrosis. The characteristic portal hemodynamics include intrahepatic perisinusoidal portal hypertension, increased splenic and portal vein blood flow and increased intrahepatic portal resistance. Patients can be misdiagnosed as having liver cirrhosis, but awareness of this condition by looking at the face and general condition along with diligent search of non-cirrhotic portal hypertension can identify this subset of patients who have better prognosis then cirrhotic patients with similar symptoms.

Keywords: Portal fibrosis, Portal hypertension, Variceal bleeding, Non-cirrhotic portal fibrosis

INTRODUCTION

Non cirrhotic portal hypertension of unknown cause is a poorly understood condition attributed to obstructive portal venopathy.[1,2] The disease is diagnosed by the presence of unequivocal evidence of portal hypertension in the definite absence of liver cirrhosis and extra hepatic portal vein obstruction (EHPVO).[3] Though the etiology of INCPH can be of five types: chronic infections, exposure to medication or toxins, thrombophilia, immunological disorders, and genetic disorders and can be multifactorial etiology.[4] When portal hypertension in absence of cirrhosis occurs it generally indicates etiologically-extrahepatic portal venous bock.[5] Due to the fact that we have limited understanding of the disease pathogenesis, the treatment of Non cirrhotic portal fibrosis is still largely supportive. Recently, endothelial dysfunction has been documented during the development of portal hypertension, and its contribution to IPH is being analysed.[6] Management based on control and prophylaxis of variceal bleeding. In EHPVO, there are additional concerns of growth faltering, portal biliopathy, and liver parenchymal dysfunction and in addition to this surgical shunt are also used in different situations.[7]

CASE REPORT

Our patient 38-year-old male patient present to us with chief complaint of black tarry stool for last 3 days. He is a hawker by profession, father of three children with age of younger child being 1 year old and he came from a remote village of Murshidabad. He was free from presenting symptoms until 3 days before admission. When he started having black tarry stool. It was associated with dizziness, generalized weakness. He also reported floating of blood on flushing of his stool. On further interview he has no pain abdomen, no yellowish discoloration of eyes and urine, no abdominal distension, fully conscious, no history of blood vomiting, no history of bleeding from nose, no history of any other bleeding disorder, no history of

ingestion of any medicine (particularly pain medicine or complementary and alternative medicine). 5 years back he was admitted with similar history of black tarry stool with abdominal distension. He was diagnosed as a case of KOCH'S peritonitis with cirrhosis of liver based on his ascitic fluid ADA report of 44. No further investigation report could be found except for Upper GI endoscopy report showing Grade 3 varices.

Clinical examination shows-no icterus, mild pallor, BP: 100/60 mm of hg, PR: 80/ min, RR: 20/min, No other significant findings on general survey.

Abdominal examination

Scaphoid abdomen, slit like umbilicus in normal position, flanks resonant, no shifting dullness, no fluid thrill, no back veins, no spider naevi, spleen just palpable, liver span 14 cm, left lobe not palpable, no tenderness, no bruit, no rub.

Table 1: Biochemical parameters.

Parameters	Values
Haemoglobin	9.7
Total leukocyte count	2910
Platelets	1.35 Lakh
Sodium	136
Potassium	4.1
Urea	22
Creatinine	1.0
Total bilirubin	0.5
SGOT	32
SGPT	40
Alkaline phosphatase	120
Albumin	4.2
Prothrombin time	13.2
INR	11 Sec

Abdominal imaging

USG W/A (Figure 1): 1) Grade 1 fatty infiltration of liver, 2) Thickened gall bladder wall, 3) Mild splenomegaly and 4) No ascites.

Figure 1: USG w/a report.

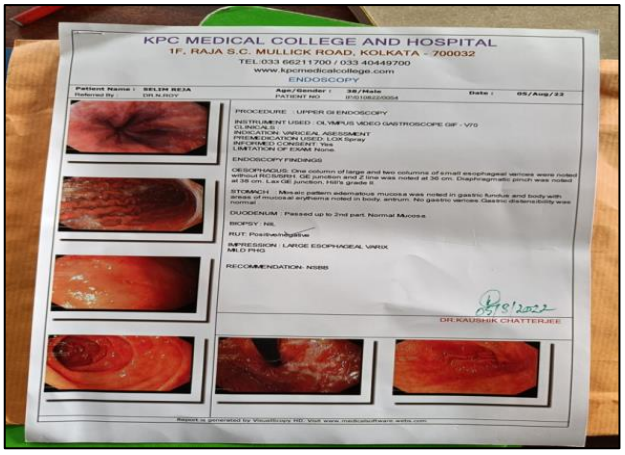

Figure 2: UGI endoscopy report.

Upper GI endoscopy (Figure 2) shows large esophageal varix.

Spleno portal doppler: suggestive of portal hypertension. On further course of treatment, we went for liver biopsy which showed evidence of portal fibrosis.

Figure 3: Difference between the patient in our case with a cirrhotic patient.

We diagnosed this patient as a case of non-cirrhotic portal fibrosis.

DISCUSSION

Non cirrhotic portal fibrosis is a disease of uncertain etiology characterized by periportal fibrosis and involvement of small and medium branches of the portal vein resulting in development of portal hypertension. Liver function and structure primarily remains normal.[8] As in India it accounts for 30 percent of variceal bleed in non-cirrhotic portal fibrosis patients thus constitute a considerable disease burden and affects young adults in prime of their lives.[9] Gastric varices are more common in non-cirrhotic portal fibrosis than oesophageal varices. The diagnosis is based on clinical features and general examination along with presences of varices, splenomegaly without features og chronic liver disease

with growth retardation may or may not be evident. The mortality is associated with other coexisting disorder and medical condition occurring at aged peoples and in comparison, to general population its survival is poor.[10] Management is primarily based on control and prophylaxis of variceal bleeding.

CONCLUSION

Non cirrhotic portal fibrosis is a heterogeneous group of liver disease and many times patients can be misdiagnosed as having liver cirrhosis, but awareness of this condition by looking at the face and general condition along with diligent search of non-cirrhotic portal hypertension can identify this subset of patients who have better prognosis then cirrhotic patients with similar symptoms. Our patients were previously falsely diagnosed as cirrhosis of liver with no general characteristics of chronic liver disease with normal liver enzyme and he also has three children. So, it's very important to look for facial and general features besides clinical features and investigations to diagnose non cirrhotic portal fibrosis in our population.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Masahiko O. Idiopathic Portal Hypertension and its pathology. Semin Liver Disease. 2002;22(1):59-72.
2. Hillaire S, Bonte E. Idiopathic non-cirrhotic intrahepatic portal hypertension in the west: a re-evaluation in28 patients. Gut. 2002;51(2):275-80.
3. Radha KD. Non-cirrhotic portal fibrosis (idiopathic portal hypertension): experience with 151 patients and a review of the literature, J Gastroenterol Hepatol. 2002;17(1):6-16.
4. Schouten JN, Garcia-Pagan JC, Valla DC. Idiopathic noncirrhotic portal hypertension. Hepatology. 2011;54(3):1071-81.
5. Mikkelsen WP, Edmondson HA, Peters RL, Redeker AG, Reynolds TB. Extra- and intra-hepatic portal hypertension without cirrhosis (hepatoportal sclerosis). Ann Surg. 1965;162:602-20.
6. Sato Y, Nakanuma Y. Role of endothelial-mesenchymal transition in idiopathic portal hypertension. Histol Histopathol. 2013;28(2):145-54.
7. Khanna R, Sarin SK. Non-cirrhotic portal hypertension - diagnosis and management. J Hepatol. 2014;60(2):421-41.
8. Sarin SK. Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment. J Gastroenterol Hepatol. 2002;17(5):526-34.
9. Sharma S, Kapoor VK. Non Cirrhotic Portal Fibrosis-Current Concepts. Gastroenterol Hepatol Open Access. 2016;5(1):00128.
10. Schouten JN, Nevens F, Hansen B. Idiopathic noncirrhotic portal hypertension is associated with poor survival: results of a long-term cohort study. Aliment Pharmacol Ther. 2012;35(12):142-33.

Cite this article as: Chanda A, Mukherjee M, Mandal M, Hossain MS, Roy N. Face says it all: a curious case of non-cirrhotic portal fibrosis. Int J Adv Med 2023;10:670-2.