Acta Scientific Gastrointestinal Disorders (ISSN: 2582-1091)

Volume 2 Issue 6 August 2019

Mini Review

A Short Review of Non Cirrhotic Portal Fibrosis

Anurag Lavekar*

Gastroenterology and Hepatology, India

***Corresponding Author:** Anurag Lavekar, Gastroenterology and Hepatology, India.

Received: June 24, 2019; **Published:** July 10, 2019

DOI: 10.31080/ASGIS.2019.02.0058

Non cirrhotic portal fibrosis (NCPF) is characterized by an increase in portal pressure, due to intrahepatic or prehepatic lesions, in the absence of cirrhosis of the liver. Various terminologies are used for describing same term i.e. NCPF in different parts of the world. E.g. in Japan, it is recognized by term idiopathic portal hypertension(IPH); though there are subtle differences between NCPF and IPH. Different terms used such as Hepatoportal sclerosis, non cirrhotic intrahepatic portal hypertension and idiopathic non cirrhotic intrahepatic portal hypertension signify same disease process i.e. NCPF [1-4].

NCPF is more commonly seen in developing countries as compared to developed countries [5-7]. It is difficult to comment on the incidence and prevalence of NCPF in India due to unavailability of national data registry system on NCPF. Peak age of incidence is between third and fourth decade of life which is almost a decade earlier than in Japan and there is no sex predilection for the disease in India [8-10]. Improved sanitation and hygiene, living conditions, changes in seroepidemiology of hepatitis A, reduction in frequency of umbilical sepsis, etc. are different proposed mechanisms for reduction of overall prevalence of NCPF in India.

Repeated septic embolization of portal circulation, xenobiotic exposure, immunologic abnormalities such as increased T4 to T8 ratio, chronic arsenic ingestion. Schistosomiasis, etc are a variety of mechanisms supposed to be involved in the pathogenesis of the NCPF [11]. On gross examination, liver may appear enlarged, shrunkened or normal in size. Surface may be smooth or nodular as in cirrhosis. Fibrous capsular thickening with increased vascularity may be seen. Deeper liver parenchyma is usually normal. Histological features reveal intimal fibrosis with subendothelial thickening of the wall of medium sized portal vein branches [12]. Increased portal collagenous connective tissue and sclerosis and obliteration of small branches of portal veins is reported. This is histological hallmark of the disease and is known as Obliterative portal venopathy [13].

GI bleed (hematemesis/melena) and splenomegaly are most common clinical presentations of the NCPF. Left upper quadrant pain can also occur as a result of perisplenitis and splenic infarction [14]. Anemia ensues as a result of hypersplenism. Jaundice, ascites and encephalopathy are relatively uncommon and transient features and usually follow an episode of GI bleed. Mortality secondary to variceal bleed is less common as the liver function is well preserved. Glomerulonephritis, hypoxemia, autonomic dysfunction are rare presentations [15-18]. Bleeding rate from the gastroesophageal varices in NCPF is very high (32-95%). Even though mortality due to GI bleed is less commonly seen due to preserved liver functions, GI bleed (variceal) is the most common cause of mortality in NCPF. Successful eradication of gastroesophageal varices ensures a 2 to 5 year survival rate of nearly 100% [19]. Development of ascites and portal venous thrombosis are indicators of clinical deterioration and progression of the disease process.

Absence of regenerative nodules with features of possible or definite cirrhosis in an adequate-sized liver biopsy is important for diagnosis of NCPF. Presence of moderate to massive splenomegaly, evidence of portal hypertension, varices, and/or collaterals, patent spleno-portal axis and hepatic veins on ultrasound Doppler, test results indicating normal or near-normal liver functions, normal or near-normal hepatic venous pressure gradient, and liver histology—no evidence of cirrhosis or parenchymal injury are six essential diagnostic criteria. Other features include absence of signs of chronic liver disease, no decompensation after variceal bleed except occasional transient ascites, absence of serum markers of hepatitis B or C virus infection, no known etiology of liver disease, and imaging with ultrasound or other imaging techniques showing dilated and thickened portal vein with peripheral pruning and periportal hyperechoic areas [11].

Liver function tests are normal or near normal in patients with NCPF. Patients do have microcytic hypochromic anemia (due to GI bleed) or normocytic normochromci anemia (due to hypersplenism). Hypersplenism is reflected in complete blood counts as pancytopenia. Significant prolongation of the prothrombin time and significantly reduced fibrinogen levels have been reported by some authors [19]. Esophageal varices are seen in 85-90% patients on endoscopy and varices are generally large at the time of diagnosis. Gastric varices in NCPF are more commonly seen than with cirrhosis and are usually associated with large esophageal varices [20]. Prevalence of anorectal varices is also reported to be significantly higher in NCPF than cirrhosis (89 Vs 56%) [21]. Hepatic venous pressure gradient (HVPG) is normal in NCPF and the hemodynamic studies indicate that the site of resistance is predominantly presinusoidal [11]. Whether HVPG increases in long run needs to be studied.

Management of acute variceal bleeding revolves around endoscopic therapy (Endoscopic sclerotherapy - EST or endoscopic variceal ligation - EVL).Endoscopic variceal ligation is the preferred modality due lesser complication rate. A combination of pharmacotherapy (Octreotide or terlipressin) plus endotherapy is superior to either pharmacotherapy or endothearpy alone in controlling acute bleed and preventing rebleed within 5 days [22,23]. Continued variceal bleed after two endoscopic treatments during single hospital admission for acute bleed is considered as failure of endoscopic therapy. Alternative treatment options such as rescue transjugular intrahepatic portosystemic shunt (TIPS) should be considered in such cases depending on the available expertise. Patients with transient ascites should undergo devascularization procedure.

EVL and beta blockers are routinely used therapies for primary prophylaxis of large esophageal varices in cirrhosis [24]. However, same treatment modalities may not be applicable in the setting of NCPF as majority patients of NCPF fall under Child Pugh Class A. In a study done by Sarin et. al, beta blockers and EVL was found to be equally effective in preventing first bleed [25]. A shunt surgery may be indicated for primary prophylaxis in patients with large esophageal varices with massive symptomatic splenomegaly, platelet counts less than 20,000/dl, non availability of GI endoscopy unit where variceal bleed can be managed. Patients with gastric varices of more than 2 cm could be taken up for surgical shunt or balloon-occluded retrograde transvenous obliteration (BRTO) if a splenorenal shunt is present. However, more studies validating this recommendation are needed.

Bibliography

1. Mikkelsen WP., *et al*. "Extra- and intra-hepatic portal hypertension without cirrhosis (hepatoportal sclerosis)". *Annals of Surgery* 162 (1965): 602-620.

2. Bioulac-Sage., *et al*. "Hepatoportal sclerosis". *Seminars in Liver Disease* 15 (1995): 329-339.

3. Kingham JGC., *et al*. "Non-cirrhotic intrahepatic portal hypertension: a long term follow-up study". *QJM* 50 (1981): 259-268.

4. Hillaire S., *et al*. "Idiopathic non-cirrhotic intrahepatic portal hypertension in the West: a re-evaluation in 28 patients". *Gut* 51 (2002): 275-280.

5. Dhiman RK., *et al*. "Non-cirrhotic portal fibrosis (idiopathic portal hypertension): experience with 151 patients and a review of the literature". *Journal of Gastroenterology and Hepatology* 17 (2002): 6-16.

6. Sarin SK. "Non-cirrhotic portal hypertension". In: Advances in therapeutic hepatology: a world view. Postgraduate Course, AASLD (1998).

7. Pande C., *et al*. "Non-cirrhotic portal fibrosis: a clinical profile of 366 patients [abstract]". *American Journal of Gastroenterology* 101 (2006): S191.

8. Bhargava DK., *et al*. "Efficacy of endoscopic sclerotherapy on long-term management of esophageal varices: a comparative study of results in patients with cirrhosis of the liver, non-cirrhotic portal fibrosis (NCPF) and extra hepatic portal venous obstruction (EHO)". *Journal of Gastroenterology and Hepatology* 6 (1991): 471-475.

9. Banghar PK., *et al*. "Profile of portal hypertension in Bombay". *Indian Journal of Gastroenterology* 11 (1992): A25.

10. Abraham P., *et al*. "Non-cirrhotic portal fibrosis in Bombay". *Indian Journal of Gastroenterology* 14 (1995): A97.

11. Sarin., *et al*. "Noncirrhotic portal fibrosis/idiopathic portal hypertension: APASL recommendations for diagnosis and treatment". *Hepatology International* 1.3 (2007): 398-413.

12. Fukuda K., *et al*. "Portal vein or hepatic vein? A curious aberrant vasculature in the liver with idiopathic portal hypertension". *Acta Pathology* 35 (1985): 885-897.

13. Nayak NC and Ramalingaswamy V. "Obliterative portal venopathy of the liver". *Archives of Pathology* 87 (1979): 359-369.

14. Vasia M., *et al*. "Idiopathic portal hypertension with splenic infarct. An unreported complication". *Acta Gastroenterology Latinoam* 31 (2001): 27-30.

15. Soma J., *et al*. "Membranoproliferative glomerulonephritis induced by portosystemic shunt surgery for non-cirrhotic portal hypertension". *Clinical Nephrology* 48 (1997): 274-281.

16. Kumar A., *et al*. "Glomerulonephritis complicating non-cirrhotic portal fibrosis". *Journal of Gastroenterology and Hepatology* 13.1 (1998): 271-275.

17. Babbs C., *et al*. "Non-cirrhotic portal hypertension with hypoxaemia". *Gut* 29 (1998): 129-131.

18. Rangari M., *et al*. "Prevalence of autonomic dysfunction in cirrhotic and noncirrhotic portal hypertension". *American Journal of Gastroenterology* 97 (2002): 707-713.

19. Sarin SK and Kapoor D. "Non-cirrhotic portal fibrosis: Current concept and management". *Journal of Gastroenterology and Hepatology* 17 (2002): 526-534.

20. Amarapurkar DN., *et al*. "Stomach in portal hypertension". *Journal of the Association of Physicians of India* 41 (1993): 638-640.

21. Chawla Y and Dilawari JB. "Anorectal varices—their frequency in cirrhotic and non-cirrhotic portal hypertension". *Gut* 32 (1991): 309-311.

22. de Franchis R., *et al*. "Diagnosis and treatment of portal hypertension". *Digestive and Liver Disease* 36 (2004): 787-798.

23. Dagher L and Burroughs A. "Variceal bleeding and portal hypertensive gastropathy". *European Journal of Gastroenterology and Hepatology* 13 (2001): 81-88.

24. Lay CS., *et al*. "Endoscopic variceal ligation versus propranolol in prophylaxis of first variceal bleeding in patients with cirrhosis". *Annals of Gastroenterology* 21 (2006): 413-419.

25. Sarin SK., *et al*. "Endoscopic variceal ligation plus propranolol versus endoscopic variceal ligation alone in primary prophylaxis of variceal bleeding". *The American Journal of Gastroenterology* 100 (2005): 797-804.

Volume 2 Issue 6 August 2019