VOLUME - 10, ISSUE - 10, OCTOBER - 2021 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Medicine

NON-CIRRHOTIC PORTAL FIBROSIS CAUSING PORTAL HYPERTENSION

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ABSTRACT Portal hypertension is more prevalent in patients with liver cirrhosis and occurs infrequently in those without liver cirrhosis. Non-cirrhotic portal fibrosis (NCPF) and extrahepatic portal venous obstruction (EHPVO) are the two most common causes of non-cirrhotic portal hypertension. Unlike EHPVO, NCPF does not cause thrombosis of the extrahepatic portal vein. Sclerosis of the portal vein's medium and small branches occurs in NCPF. In NCPF, the hepatic venous pressure gradient (HVPG) is normal, in contrast to cirrhosis, where it is increased. Additionally, NCPF is referred to as non-cirrhotic intrahepatic portal hypertension (NCIPH), idiopathic portal hypertension, hepatoportal sclerosis, and benign intrahepatic portal hypertension. It is a disease with an unknown etiology that primarily affects middle-aged males and females and manifests as hematemesis and massive splenomegaly.

KEYWORDS : Portal hypertension, non-cirrhotic portal fibrosis, massive splenomegaly

INTRODUCTION

Non-cirrhotic portal fibrosis (NCPF) is an etiologically unknown condition characterized by fibrosis of the portal vein's small and medium branches, resulting in portal hypertension (PHT). The structure and function of the liver remain normal. [1] It is a vascular condition of the liver that results in non-cirrhotic portal hypertension (NCPH), defined as a pressure gradient larger than 5mm Hg between the portal vein and the intraabdominal inferior vena cava in the absence of cirrhosis. [2] Increased portal pressure and resistance to prograde splanchnic flow finally results in the opening of the portosystemic collateral circulation, resulting in congestive splenomegaly. Varices, moderate to severe splenomegaly with or without symptomatic hypersplenism, intact liver function, and patent hepatic and portal veins are all present clinically. [2,3] It has been described worldwide, particularly in underdeveloped nations, and has been variously labelled as NCPF (India), idiopathic portal hypertension (Japan), hepatoportal sclerosis, incomplete septal cirrhosis, and nodular regenerative hyperplasia (West). [4–7] Each of these cases exhibits obliterative vascular lesions in the portal venous system and shares a similar clinical presentation. As a result, it has been proposed that they be treated collectively as a single entity with a variety of clinical manifestations, rather than as distinct clinicopathological entities.[8] According to the Asia Pacific Association for the Study of the Liver's consensus statement on NCPF, the disease accounts for between 10% to 30% of all instances of variceal bleed in numerous countries, including India.[1] A comparable illness process accounts for 3-5 percent of PHT patients in the Western world.[8] The demographic discrepancies in clinical manifestations associated with this disease between India, Japan, and the West are most likely due to changes in living conditions, ethnicity, average life expectancy, reporting bias, and diagnostic criteria utilized.

Case Presentation

A 39 year old female presented with chief complaints of abdominal discomfort for 6 months and abdominal pain for 3 months. Patient was apparently asymptomatic 6 months back when she developed gradual and progressive abdominal distension more in the left upper quadrant for 6 months. Later associated with abdominal discomfort, on and off in the form of mild dull pain aggravated on getting up from bed, bending down since 3 months. She had no history of jaundice, vomiting, oral ulcer, hematemesis, reflux, dysphagia, odynophagia, constipation, diarrhea, blood in stools, dark stools, anorexia, weight loss, nausea, fatigue, fever, pruritis, confusion, altered sleep, chest pain, palpitation and involuntary movement. She had past history of there been no known comorbidities. She has normal sleep, appetite, bowel movement and no history of substance abuse and addiction. Her Menarche was at 12yrs with regular cycle. She had a single boy with normal delivery and never undergone abortions.

During general examination, she was conscious, oriented, comfortable and cooperative. There were no pallor, icterus, clubbing, cyanosis and pedal edema whereas significant lymphadenopathy was observed. There were no external markers of liver cell failure in head to toe examination. The hemodynamic parameters such as pulse rate-72/min (regular, normal volume, character, all peripheral pulses felt bilaterally equal, no thickening of vessel wall), blood pressure-BP-110/70mmHg, respiratory rate-12'min, SpO2- 99% and she was afebrile.

The gastrointestinal tract examination revealed that no fetor hepaticus and oral ulcer in her oral cavity. While examining, the abdomen distended (L>R), Umbilicus normal, no dilated veins, no scars and Hernial orifice normal. A non-tender, swelling palpated in right hypochondrium 2 cm below the RCM in MCL which movies with respiration and is firm in consistency with regular surface. Not able to palpate the upper border; probably liver. A non-tender swelling is palpated in left hypochondrium enlarging towards right iliac fossa 8cm below the LCM in MCL crossing midline. Firm in consistency with smooth surface and notch along medical border. Not bimanually palpable or ballotable; probably enlarged spleen.

Routine complete blood count, renal function test, liver function test and coagulation profile was done and observed values were described in the table 1-4.

Table 1: Cbc Profile

CBC	Observed values
Hb	13.1
TC	3,400
DC	68/30/2
PCV	41%
Platelet	36,000
ESR	3/9

Table 2: Renal Function Test Profile

RFT	Observed values
Blood Glucose (Random)	78
Urea (mg/dl)	24
Creatinine (mg/dl)	0.9
Sodium (mEq)	134
Potassium (mEq)	3.8

Table 3: Liver Function Test Profile

LFT	Observed values
TB	0.8
DB	0.3
IB	0.5
SGOT	27
SGPT	28
ALP	52
TP	6.2
Albumin	4.1
Globulin	2.1
A/G ratio	1.9

Table 4: Coagulation Profile

Coagulation profile	Observed values
PT	14.9s
INR	1.2

The x-ray, ECG and urine routine were show normal. The serum ceruloplasmin was 25.0 mg/dL (the normal range 20-60 mg/dl). The ANA weak positive nucleolar 1:100. The oesophago-gastro-duodenoscopy (ODG) confirmed that presence of portal hypertensive gastropathy. The Bone marrow study was normal. Peripheral blood smear showed that erythrocyte-normocytic normochromic and thrombocytes were adequate. The leucocyte counts were P 63.58/ L 28.8/ E 4.5/ M 3.2.

The patient underwent the imaging studies. The USG abdomen with Doppler concluded that she had Massive splenomegaly, dilated portal vein (no thrombus), dilated splenic vein (Fig.1, 2 &3). Hepatic vein shows complete luminal filling with phasic fluctuation. The same clinical findings were confirmed by the MRI abdomen which showed that Massive splenomegaly, Dilated portal and splenic vein and Mild ascites. The liver biopsy was taken for histopathological analysis and it confirmed that patient had non-cirrhotic portal fibrosis (Fig.4 & 5). From all these diagnostic evaluations, the patient had Non cirrhotic portal fibrosis with portal hypertension, splenomegaly, hypersplenism (leucopenia, thrombocytopenia), minimal ascites with no other significant features of liver cell failure.

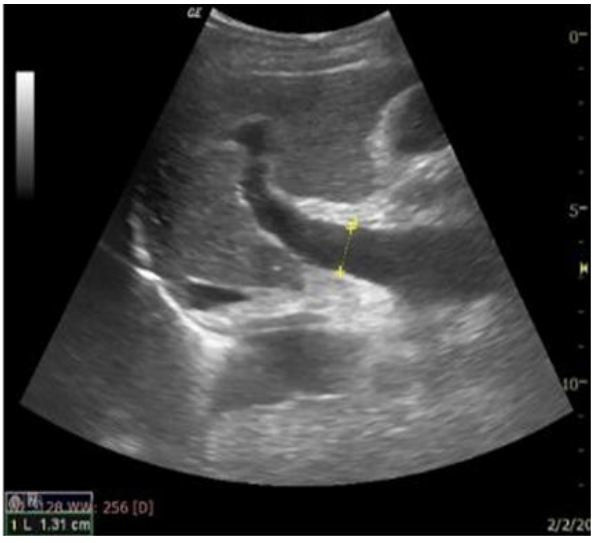

Figure 1: USG of Abdomen

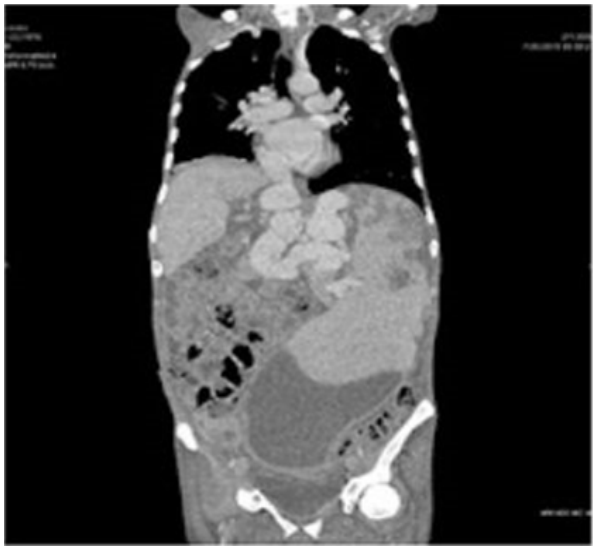

Figure 2: Ct Image Showing Large Collaterals, Massive Splenomegaly And Splenic Abscess At The Lower Pole Extending Up To The Pelvis

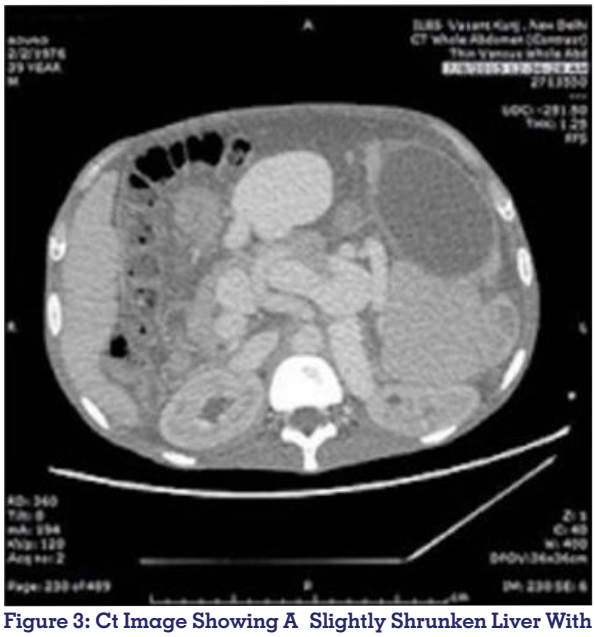

Figure 3: Ct Image Showing A Slightly Shrunken Liver With An 18 Mm Lieno-renal Shunt

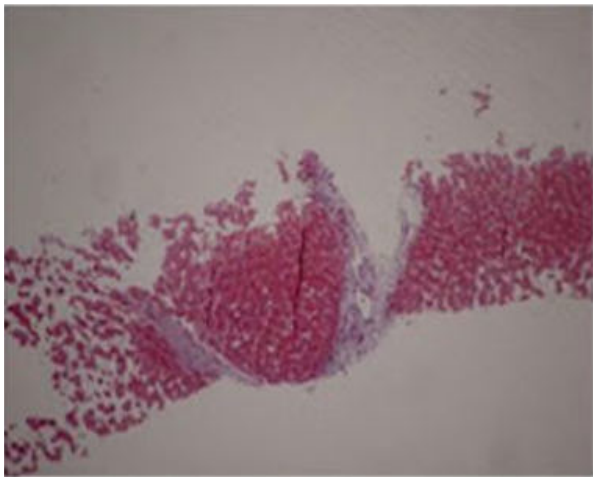

Figure 4: Trans Jugular Liver Biopsy Specimen Showing Normal Liver Parenchyma With Only Grade 1 Fibrosis Consistent With Ncpf; Massive Trichrome Stain 10x

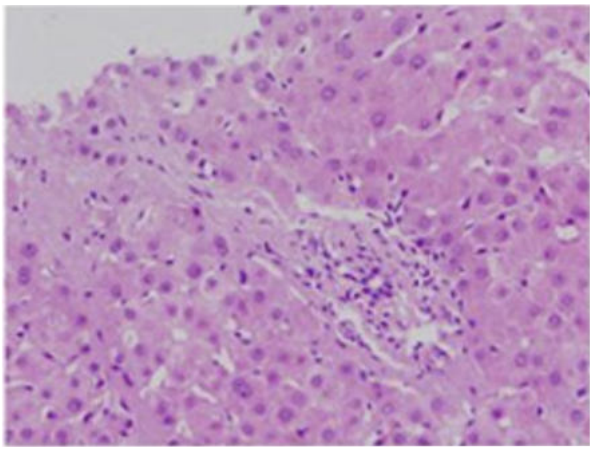

Figure 5: Liver Biopsy Specimen Showing Largely Preserved Parenchyma And A Normal Portal Tract. Hematoxylin And Eosin Stain:40x

DISCUSSION

In 27–87 percent of cases, hypersplenism is present, with anemia being the most common abnormality, followed by thrombocytopenia and leukopenia. Microcytic hypochromic anemia is typically associated with multiple variceal bleeds, hypersplenism, and iron deficiency.[9-12] Liver function tests are generally normal in NCPF/IPH, but abnormalities in liver enzymes, prothrombin time, and albumin are observed in a small proportion.[14,15] Additionally, in this case, a similar style of presentation was observed.

In both disorders, Doppler USG is the first line of radiological investigation. The liver is normal in size and echotexture in NCPF/IPH. In NCPF/IPH, the spleen is enlarged due to the presence of gamma-gandy bodies; the splenoportal axis is dilated and patent. PV is thickened (>3 mm) with echogenic walls and has smooth and regular intrahepatic radicles. There is a sudden narrowing or severance of intrahepatic second and third degree PV branches – a "withered tree" appearance accompanied by vascular channel approximation. The splenic index and PV inflow are both elevated.[15,16] 16 percent of patients have spontaneous shunts (paraumbilical and gastroadrenorenal).[16] Contrast-enhanced computed tomography (CT) features such as intrahepatic PV abnormalities (non-visualization, reduced caliber, occlusive thrombosis), focal nodular hyperplasia such as nodules, and perfusion defects aid in differentiating NCPF/IPH from cirrhosis.[17] Doppler USG of SPA has a sensitivity and specificity of greater than 95% for diagnosing EHPVO. PV undergoes cavernomatous transformation. Splenoportography and arterial portography have been phased out in favor of non-invasive methods such as computed tomography (CT) and magnetic resonance (MR) angiography and portography, which not only diagnose but also provide anatomical guidance prior to shunt surgery.[18] Although liver biopsy is not required to diagnose EHPVO unless the presence of underlying chronic liver disease is suspected, it is recommended in NCPF/IPH to rule out cirrhosis and other PHT etiologies.[19,20]

CONCLUSION

Non-cirrhotic portal fibrosis continues to be a common cause of portal hypertension. This condition's etiology and pathogenesis are likely multifactorial. Patients present clinically with splenomegaly and/or complications of portal hypertension. The patients' liver function is relatively well preserved. Clinical and endoscopic evidence of portal hypertension, as well as radiological and histological features, are used to make the diagnosis. If properly managed, these patients have a life expectancy comparable to that of the general population. Future research should focus on elucidating pathogenetic mechanisms in order to effectively prevent the condition.

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