ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 08 | August - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

TO STUDY THE ETIOLOGICAL, CLINICAL AND HISTOPATHOLOGICAL PROFILE OF PATIENTS WITH NON CIRRHOTIC PORTAL FIBROSIS IN A TERTIARY CARE CENTER : A HOSPITAL BASED OBSERVATIONAL STUDY

Gastroenterology

Dr. Rahul Garg* MBBS, MD (Medicine), Associate Professor, Medicine Department, FH Medical College and Hospital, Agra *Corresponding Author

ABSTRACT

Introduction: Non cirrhotic portal fibrosis is a syndrome characterized by obliterative portovenopathy leading to portal hypertension, massive splenomegaly and well tolerated episodes of variceal bleeding in young adults from low socioeconomic backgrounds, having near normal hepatic functions. **Objectives** : This study was designed to evaluate the patients of NCPF for clinical , etiological and histopathological profile and determine whether the source of drinking water could have a role in etiopathogenesis of NCPF. **Materials And Methods:** This retrospective study was conducted at FH Medical College and Hospital, Agra, involving 50 diagnosed NCPF patients. Distribution of cases was done according to age, sex, diet, source of drinking water, various clinical presentations on the basis of history and clinical examination, and histopathological findings. Statistical analysis was done using Chi square test and Fisher's exact test. **Results:** Study showed male preponderance. Most patients were in the age group of 25-35 years, 74% were vegetarian and 46% of the patients used tap water as the source of drinking water. Major presentation of patients with NCPF was with g.i.bleed and left upper quadrant mass on history and splenomegaly on clinical examination. Main histological findings were irregular intimal thickening of portal veins, and organizing thrombosis and/or recanalization of portal veins. p value was calculated for finding an association between source of drinking water and every histopathological finding on liver biopsy, and a significant association was found between source of drinking water as tap water and organizing thrombosis and/or recanalization of portal vein ($p = 0.0416$) which suggest that source of drinking water might have an impact on the etiology of NCPF. **Conclusion:** Etiology of NCPF is possibly multifactorial. Tap water (source of drinking water) could have a possible role in etiopathogenesis of NCPF, which needs to be further evaluated by case controlled studies.

KEYWORDS

Non Cirrhotic Portal Fibrosis (NCPF), Portal Hypertension (PTH)

INTRODUCTION

Non Cirrhotic Portal Fibrosis (NCPF) is one of the important disease entities, characterized by an increase in portal pressure, due to intrahepatic or prehepatic lesion, in the absence of cirrhosis of liver.

NCPF has been reported from all over the Indian subcontinent[1,2]. It is believed to account for nearly one-sixth to one-quarter of all cases of PTH seeking medical attention. The condition has been seen in people who are socioeconomically diasadvantaged. Most studies indicated a male predominance[1]. The mean age of patients varies from 25 to 35 years.[2]

The etiopathogenesis of NCPF is poorly understood. According to the multiple hypothesis proposed, abdominal infection at birth or early childhood[3], prolonged ingestion of arsenic[4] , inreased T4/T8 lymphocyte ratio[5] and ADAMST13 deficiency[6] has been implicated in the causation of NCPF.

Patients may present with one or more well tolerated episodes of gastrointestinal hemorrhage, long standing mass in the left upper quadrant(splenomegaly) and consequences of hypersplenism. Development of ascites, jaundice and hepatic encephalopathy is uncommon.

Histological changes in NCPF were aptly summarized by Nayak and Ramalingaswamy[1] as 'obliterative portovenopathy of liver'.[1] Major histological findings were irregular intimal thickening of portal veins and organizing thrombosis and/or recanalization of portal veins[1,7].

Our motive behind this study is to determine other clinical presentations, pathological changes and etiological factors which possibly could help to be used as a screening test for high risk patients.

MATERIAL AND METHODS:

This retrospective study was done at our center, a tertiary care center in Agra. 50 patients who were already diagnosed as a case of NCPF were included. Patients having Chronic Liver Disease due to cirrhosis, primary splenomegaly, and Extra hepatic portal vein obstruction were excluded. Chronic alcoholics were also excluded. A detailed history was taken regarding the following symptoms: hematemesis, a long standing mass in the left upper quadrant, yellowish discoloration of urine or sclera, pain in the left upper quadrant, abdominal distension, any episode of unconsciousness or altered behaviour. While performing physical examination, following signs of portal hypertension were seen: mass in the left or right upper quadrant, ascites, jaundice, hepatic encephalopathy. Patients were asked about their source of drinking water and nature of diet. Histopathological findings were recorded. Routine investigations were done. Statistical analysis was done using Chi square test and Fisher's exact test.

RESULTS:

Results observed are shown in the following tables and figures:

***Table* 1: Distribution of cases as per sex ,diet and source of drinking water**

Sex	N	%
Male	31	62
Female	19	38
Diet		
Vegetarian	37	74
Non vegetarian	13	26
Source of drinking water		
Ground	21	42
Tap	23	46
Pond	6	12

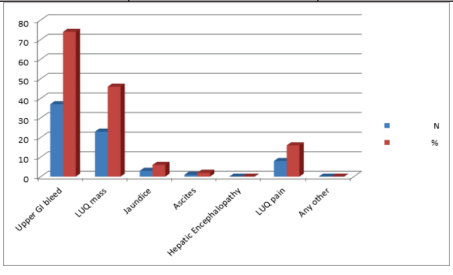

***Fig*.1:** Distribution of cases as per clinical presentation

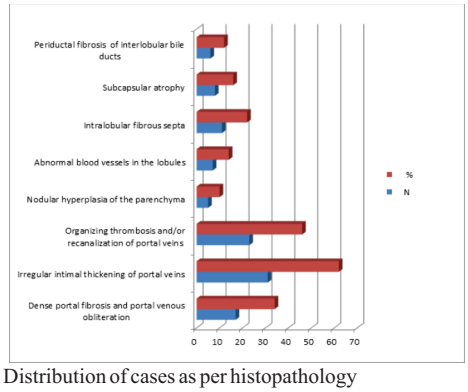

***Fig*.2 :** Distribution of cases as per histopathology

***Table* 2: Probability of occurence of a histopathological finding according to the source of drinking water**

Histopathological finding	Ground water (n=21)	Tap Water (n=23)	Pond Water (n=6)
Dense portal fibrosis and portal venous obliteration	p=0.3650	p=0.5133	p=0.0927
Irregualr intimal thickening of portal veins	p=0.4473	p=0.3462	p=0.1307
Organizing thrombosis and/or recanalization of portal veins	p=0.7492	p=**0.0416**	p=0.5851
Nodular hyperplasia of the parenchyma	p=0.3463	p=0.2286	p=0.4874
Abnormal blood vessels in the lobules	p=0.3651	p=0.1479	p=0.3836
Intralobular fibrous septa	p=0.3859	p=0.5023	p=0.1737
Subcapsular atrophy	p=0.0690	p=0.5023	p=0.2416
Periductal fibrosis of interlobular bile ducts	p=0.5009	p=0.5851	p=0.5557

DISCUSSION:

This retrospective study looked at the clinical, etiological and histopathological profile of 50 patients with non-cirrhotic portal fibrosis (NCPF) at our center. The results showed some important findings regarding the demographics, presentations, and potential etiological factors in this patient population.

In line with previous literature, there was a male preponderance with 62% of cases being male. The mean age range was 25-35 years, which is the typical age distribution reported for NCPF. An interesting finding was that 74% of patients were vegetarian, which has not been commonly highlighted in prior studies on NCPF. (Table 1)

The major clinical presentations were gastrointestinal bleeding (74%) and left upper quadrant mass representing splenomegaly (46%), consistent with the usual manifestations of portal hypertension in NCPF. Development of jaundice, ascites and hepatic encephalopathy was uncommon, reflecting the preservation of liver synthetic functions in these patients. (Fig.1)

On histopathological examination, the key findings were irregular intimal thickening of portal veins (62%), organizing thrombosis and/or recanalization of portal veins (46%), and dense portal fibrosis with portal venous obliteration (34%). These pathological changes correlate with the "obliterative portovenopathy" that characterizes NCPF. (Fig.2)

An important finding was the significant association between using tap water as the source of drinking water and the presence of organizing thrombosis and/or recanalization in portal veins on biopsy (p=0.0416) as shown in Table 2. This suggests that the source of drinking water could potentially be an etiological factor contributing to NCPF, possibly through contaminants or toxins. However, this association needs to be interpreted cautiously given the retrospective study design. The author acknowledges that the etiology of NCPF is likely multifactorial, with previous hypotheses implicating factors like abdominal infections, arsenic exposure, ADAMST13 deficiency and immunological mechanisms. This study highlights a novel potential association with drinking water source that merits further investigation through well-designed prospective case-control studies.

Overall, this study adds to the existing literature on NCPF by providing insight into the clinico-pathological profile and raising an important question about the role of drinking water in disease pathogenesis in this endemic region of India. Unraveling the complex etiology is crucial for developing preventive strategies against this disabling condition predominantly affecting young adults.

Limitations:

Limitations of the study include its retrospective design, which has inherent limitations. Small sample size may limit the generalizability of findings to broader populations. Possibility of recall bias regarding source of drinking water and dietary habit could be a factor. The study did not include a control group of healthy individuals or patients without NCPF for comparison, which would have strengthened the analysis of potential risk factors, such as the source of drinking water.

CONCLUSION:

This study has shown that the etiopathogenesis of this condition is possibly multifactorial. Tap water (source of drinking water) could have a possible role in etiopathogenesis of NCPF, which needs to be further evaluated by case controlled studies. Common clinical presentation is with hematemesis and splenomegaly. If managed suitably, these patients have a life expectancy similar to that of the population at large.

***Conflict of interest*:**

This research is conducted independently and authors declare no conflict of interest with pharmaceutical companies or other entities.

REFERENCES:

1. Nayak NC, Ramalingaswami V. Obliterative portal venopathy of the liver. A study of 49 surgical specimens in Indians. *Arch. Path*. 1969;87(4):359-69.
2. Sarin SK. Non-cirrhotic portal fibrosis. Gut. 1989 Mar;30(3):406-15.
3. Sarma MS, Seetharaman J. Pediatric non-cirrhotic portal hypertension: Endoscopic outcome and perspectives from developing nations. World J Hepatol. 2021 Oct 27;13(10):1269-88.
4. Datta DV, Mitra SK, Chhuttani PN, Chopra B. Arsenic content in drinking water and noncirrhotic portal fibrosis. *Gastroenterol*. 1976;70(4):677-8.
5. Sarin SK, Nayyar AK, Malhotra P, Sharma BK, Kumar R, Broor SL. Immunological profile of patients with non-cirrhotic portal fibrosis. J Gastroenterol Hepatol. 1990 Jul-Aug;5(4):425-31.
6. Mackie I, Eapen CE, Neil D, et al. Idiopathic noncirrhotic intrahepatic portal hypertension is associated with sustained ADAMTS13 deficiency. Dig Dis Sci. 2011;56:2456–65.
7. Chougule A, Rastogi A, Maiwall R, et al. Spectrum of histopathological changes in patients with non-cirrhotic portal fibrosis. Hepatology International. 2018 Mar;12(2):158-66.