References


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Idiopathic Noncirrhotic Intrahepatic Portal Hypertension Is an Ongoing Problem in India

To the Editor:

Schouten et al. describe the need for histological confirmation of idiopathic noncirrhotic portal hypertension and its contrasting incidence between the West and India.1 We prefer the term idiopathic noncirrhotic intrahepatic portal hypertension (NCIPH) to distinguish it from extrahepatic portal vein thrombosis—the most common cause of pediatric portal hypertension at our center.2

After the report from Chandigarh in 2002,3 there is scarce literature on the incidence of biopsy-proven NCIPH in India. We herein report on our recent experience with NCIPH.

From 2005 to 2007, retrospective analysis of 227 portal hypertensive patients who underwent liver biopsy at our center showed that of 62 patients labeled as having "cryptogenic cirrhosis," 30 (48%) were diagnosed as having NCIPH after liver biopsy.4

We prospectively studied the prevalence of NCIPH among all new portal hypertensive patients in our unit from July 2009 to July 2010 (after institutional ethics committee approval). NCIPH was diagnosed as per the previously described criteria.4 The need for liver biopsy in each patient was decided on a case-by-case basis, based on the clinical scenario.

Of 610 consecutive new portal hypertensive patients studied, cryptogenic cirrhosis (210 patients) was the most common cause of portal hypertension identified after noninvasive tests. Of 44 cryptogenic cirrhosis patients who underwent liver biopsy, 17 (39%) had NCIPH and 8 had "true cryptogenic cirrhosis." NCIPH and true cryptogenic cirrhosis patients were 27 (range, 14-59) and 42 (range, 25-67) years old, respectively; 10 and 4 patients, respectively, were males. Hepatic venous pressure gradient measured in 15 NCIPH and 4 true cryptogenic cirrhosis patients was 7 (range, 1-21) and 18 (range, 10-27) mmHg, respectively (P = 0.012).

Liver biopsies were performed percutaneously in 4 NCIPH patients and transjugularly in 13. Number of cores in percutaneous biopsies was 3 per patient and 3 (range, 1-6) in transjugular biopsies; length of the largest core was 13 (range, 12-15) in percutaneous and 12 mm (range, 6-16) in transjugular biopsies. The number of portal tracts in liver biopsies was 10 (range, 5-20).

Liver biopsies showed no significant fibrosis (6 patients), mild portal/periportal fibrosis (10), moderate fibrosis (1), mild perisinusoidal fibrosis (1), abnormal portal venous ectasia (6), and mild diffuse sinusoidal dilatation (8); no patient had cirrhosis or severe fibrosis.

In summary, in 2009-2010 and 2005-2007,4 39%-48% of patients with clinical diagnosis of cryptogenic cirrhosis who underwent liver biopsy at our center had NCIPH.

References


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Reply:

We would like to thank Goel et al. for their reply to our review of idiopathic noncirrhotic portal hypertension (INCPH).1,2

The authors studied the prevalence of INCPH among all new portal hypertension patients from July 2009 until July 2010. Eventually, 17 (39%) patients were diagnosed with INCPH. The authors described the morphological features observed in liver specimens from these 17 INCPH patients. Interestingly, neither
the presence of nodular regenerative hyperplasia nor obliteration of portal venules (phlebosclerosis)—both the most frequently observed morphological features in liver specimens of Western INCPH patients—were found. The apparent absence of these features in Indian INCPH patients could imply differences in pathophysiological mechanisms between patients from different continents.

As reported by Goel et al., liver biopsy is mandatory in the diagnosis of INCPH. Its main role is the exclusion of liver cirrhosis, which may be very difficult to discern by radiological examinations only. In addition, the diagnosis of INCPH can be suggested or supported by the presence of morphological features such as nodular regenerative hyperplasia, phlebosclerosis, increased number of portal vascular channels, paraportal shunt vessels, and sinusoidal dilatation. However, one must take into account that these features can also be observed in patients without clinical signs of portal hypertension.

References


4. Speliotes EK, Butler JL, Palmer CD, Voight BF;GIANT Consortium; MIGen Consortium; NASH CRN, Hirschhorn JN. PNPLA3 variants specifically confer increased risk for histologic nonalcoholic fatty liver disease but not metabolic disease. HEPATOLOGY 2010;52:904-912.

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