Portal Cavernoma Cholangiopathy – History, Definition and Nomenclature

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Biliary changes secondary to portal hypertension, especially in portal cavernoma secondary to extrahepatic portal vein obstruction have long been described in literature under different names by various authors. Most of the times these changes are asymptomatic and discovered on imaging, but can occasionally cause obstructive jaundice. There is no consensus on the appropriate nomenclature and definition of this entity. This article reviews the history of portal hypertensive biliopathy and the Indian Association for the Study of Liver Working Party consensus definition and nomenclature for it. (J Clin Exp Hepatol 2014;4:S15–S17)

The relationship between jaundice and extrahepatic portal vein obstruction (EHPVO) was first reported by Gibson et al in 1965.[1] Out of 28 cases they found jaundice in 5 patients. In 4 of these cases the jaundice was attributed to the etiology of the EHPVO like inflammatory portal mass and hilar cholangiocarcinoma, or due to associated chronic cholecystitis. In only one case a large collateral vessel was seen lying across a narrowed segment of the bile duct during shunt surgery and possibly compressing it. This was the first report of biliary system involvement as a result of the carvernomatous transformation of the portal vein. After that several reports of jaundice or varices along the bile duct in patients of EHPVO are available in the literature.[2–4] The first description of the cholangio graphic changes in EHPVO was by Williams et al in 1982 who demonstrated varices along the bile duct.[5] The first prospective case series describing biliary abnormalities on endoscopic retrograde cholangiography (ERC) in patients of EHPVO was published by Dilawari and Chawla in 1992.[6] All 20 patients were found to have biliary abnormalities similar to that seen in sclerosing cholangitis, mainly involving the left hepatic duct and mid common bile duct. Since then, several case series have described biliary abnormalities on ERC among patients with portal hypertension.[7–15] Subsequently biliary changes were also demon strated in other causes of portal hypertension like non-cirrhotic portal fibrosis (NCPF) and cirrhosis of liver, albeit with less frequency.[11,16,17] Biliary changes are seen on cholangiography in 81–100% of patients with EHPVO[6–15] compared with 0–33% in cirrhosis of liver[10,11,17] and 9–40% in NCPF.[11,17] This is probably due to the longer duration of portal hypertension in cases of EHPVO in many of whom the obstruction to the portal vein occurs in infancy or childhood compared to cirrhosis wherein the duration of portal hypertension is less. This is supported by also a significantly higher frequency of anorectal varices, large oesophageal varices and variceal bleeding in patients with EHPVO compared with those having cirrhosis.[18,19] However, despite the high frequency of cholangiographic changes these same studies found that only a minority of these patients are symptomatic, and symptoms are more likely in patients with long standing portal hypertension.

DEFINITION

Sarin et al have defined portal biliopathy broadly as biliary ductal and gallbladder wall abnormalities seen in patients with portal hypertension which though simple is vague and would include biliary changes due to any other disease in a patient who happens to have portal hypertension also.[7] Dhiman et al have proposed a more descriptive definition as abnormalities of the entire biliary tract including intrahepatic and extrahepatic bile ducts, cystic duct and gallbladder in patients with portal hypertension.[20] This definition also does not specify what changes are to be included and in which portions of the biliary tree the changes are mandatory. The need was felt to have a definition which would be specific to this condition so as to have reproducibility between studies from different authors on this subject. Definition of this entity of biliary changes in patients with portal hypertension was debated by the working party on two fronts: 1. Of the many biliary changes described on cholangiography, which ones have been consistently reproduced in the literature and are

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Abbreviations: EHPVO: extrahepatic portal vein obstruction; ERC: endoscopic retrograde cholangiography; NCPF: non-cirrhotic portal fibrosis
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specific for this condition; 2. Is the mere presence of portal hypertension of any etiology enough to produce these changes, or is the presence of a cavernoma mandatory?

Review of description of cholangiographic findings in portal biliopathy revealed that the extrahepatic biliary ducts were almost invariably involved, and that changes had been described in the gallbladder and cystic duct as well. Changes have also been described upto the 2nd generation of biliary ducts. Although portal biliopathy is seen in other causes of portal hypertension as well, majority of the typical changes have been described in relation with EHPVO and portal cavernoma is present in most of these patients. Also the natural history, prognosis and response of the biliopathy to shunt surgery will vary between various causes of portal hypertension. Another difference is that the veins involved in cavernoma formation in EHPVO are portoportal collaterals whereas in other settings of portal hypertension like cirrhosis the collaterals are usually portosystemic. Hence, to maintain uniformity between studies and to ensure a homogenous patient population the presence of a portal cavernoma was considered to be a part of the definition.

The working party arrived at a consensus definition, stating that the entity would be defined as abnormalities in the extrahepatic biliary system including the cystic duct and gallbladder with or without abnormalities in the 1st and 2nd generation biliary ducts in a patient with portal cavernoma. For the diagnosis to be established all of the following three criteria would have to be fulfilled: 1. The presence of a portal cavernoma, 2. Cholangiographic changes on ERC or magnetic resonance cholangiography consistent with typical changes described for this entity (Table 1) and 3. Absence of other causes of these biliary changes like bile duct injury, primary sclerosing cholangitis, cholangiocarcinoma etc. (Table 2).

NOMENCLATURE

The biliary changes secondary to portal hypertension have been described by a multitude of names in published literature. Dilawari and Chawla described findings similar to sclerosing cholangitis, especially in the left hepatic duct.

Table 1 Cholangiographic Abnormalities of Portal Cavernoma Cholangiopathy.

Extrinsic impressions/indentations
Shallow impressions/indentations
Irregular ductal contour
Stricture
Filling defects
Bile duct angulation
Upstream dilatation
Ectasia

Table 2 Differential Diagnosis of Portal Cavernoma Cholangiopathy.

Primary sclerosing cholangitis
Bile duct neoplasms
Biliary tract surgery
Recurrent pyogenic cholangitis
AIDS cholangiopathy
Biliary parasitosis
Choledocholithiasis
Congenital abnormalities of the biliary tract
Ischemic bile duct stricture
Toxic bile duct strictures
Strictures due to autoimmune and chronic pancreatitis

However, as only 1 of the 20 patients had evidence of cholangitis or even biochemical cholestasis, and unlike primary sclerosing cholangitis, biliary strictures in patients with EHPVO were smooth rather than irregular they termed it as "pseudosclerosing cholangitis".[6] At about the same time Bayraktar et al described it as the "pseudocholangiocarcinoma sign" due to ERC similarities with cholangiocarcinoma.[9] Sarin et al named it as "portal biliopathy" to reflect the biliary changes secondary to portal hypertension.[7] Similarly Malkan et al called it "cholangiopathy associated with portal hypertension", which while descriptively accurate was cumbersome to use.[11] Subsequently the entity was called "portal cavernoma associated cholangiopathy" as almost all patients had portal cavernoma at surgery.[13] Dhiman et al named it "portal hypertensive biliopathy" as the entire or part of the biliary tract is involved in these patients, and these abnormalities can be due to portal hypertension of any etiology and not necessarily portal cavernoma.[15] Other terminology that have been used include "extrahepatic portal biliopathy"[21] "vascular biliopathy",[22] "ischemic cholangiopathy",[23] "portal ductopathy",[24] and "portal cholangiopathy".[25]

In light of the new definition adopted by the working party the term "Portal Cavernoma Cholangiopathy" was agreed upon as the consensus nomenclature as it implied the presence of a portal cavernoma resulting in abnormalities of the biliary tree including extra- and intrahepatic bile ductular system, gallbladder and cystic duct.

CONFLICTS OF INTEREST

All authors have none to declare.

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