Pseudosclerosing cholangitis in extrahepatic portal venous obstruction

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Abstract

Biliary changes secondary to portal hypertension have rarely been described in published reports. Twenty consecutive patients with extrahepatic portal venous obstruction, all of whom showed a variable degree of abnormalities in the biliary tract suggestive of sclerosing cholangitis, are described. These biliary abnormalities were: focal narrowing, dilatations, and cholangitic changes affecting the main bile ducts and hepatic ducts. The left hepatic duct and its branches were affected in all patients. Only one patient had clinical or biochemical evidence of cholestasis. The mechanism of these abnormalities in the biliary tract of these patients is perhaps the development of portal collaterals.

There are only a few case reports of biliary changes in patients with portal hypertension,¹⁻⁴ and most are of patients with extrahepatic portal venous obstruction. We studied the frequency of biliary changes in patients with extrahepatic portal venous obstruction by endoscopic retrograde cholangiopancreatography (ERCP). To the best of our knowledge this is the first report describing biliary abnormalities in such patients.

Methods

ERCP

ERCP was performed by the standard technique using a side viewing duodenoscope (Olympus JF 10) to delineate the intra- and extrahepatic biliary system and pancreatic duct. Patients received premedication with intravenous diazepam (10 mg) and hyoscine hydrochloride (40 mg) before ERCP. All the patients gave informed consent. They were told about the

procedure and what we were looking for and advised that the ERCP findings would probably have little bearing on their management.

SPLENOPORTOVENOGRAPHY

Percutaneous splenoportovenography was done under fluoroscopic control using 40 ml of Conray 420 (sodium iothalamate BP) by the standard technique⁵ (Fig 1). Six films at the rate of 1/second were taken. Spontaneous splenorenal shunt was defined when contrast, injected into the splenic pulp, was seen entering the inferior vena cava through collaterals between the splenic and renal veins.

SPLENIC PULP PRESSURE

Splenic pulp pressure was measured using an electronic pressure recorder (Siemens mingo-graph) at the time of splenoportovenography.

OESOPHAGOGASTRODUODENOSCOPY

Upper gastrointestinal endoscopy was undertaken to look for any mucosal lesion in the oesophagus, stomach, or duodenum. Oesophageal varices, if present, were graded according to the criteria of Paquet⁶ (grade I-IV).

PATIENTS

Two groups of patients were included in the study.

Control group

The control group comprised 15 age and sex matched patients with a history of abdominal pain suggestive of pancreatic disease but with a normal upper gastrointestinal endoscopy and ERCP. Patients in both the groups were clinically assessed for any biliary symptoms. Liver function tests were done in all.

Extrahepatic portal venous obstruction

Twenty consecutive patients with an established diagnosis of extrahepatic portal venous obstruction by splenoportovenography and splenic pulp pressure were included in this group. All had a previous history of massive upper gastrointestinal bleed, large varices (grades III-IV) on endoscopy, and a moderately enlarged spleen. All patients, except one, underwent repeated sessions of endoscopic sclerotherapy. Obliteration of varices was achieved in 17 patients at the time of ERCP. Successful ERCP was performed in all 20. There were 16 men and four women, and the average age was 22 years (Table I).

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Figure 1:
Splenoportovenogram showing portal cavernoma (PC) instead of a well defined portal vein in a patient with portal hypertension SV = splenic vein.

TABLE I Parameters studied in 20 patients with extrahepatic portal venous obstruction

Age sex	DOI (yrs)	SPL (cm)	SOB	SPP (mm Hg)	EST (no)	EST Res	Interval OBL-ERCP (mths)	MBD max diam (mm)/findings	LHD max diam (mm)/findings	RHD max diam (mm)/findings
38M	7	5	PVT (D)	44.5	3	OBL	2	6 IND	5 SEV	5 MOD
24M	4	3	PVT (D)	28.5	5	OBL	42	9 IND, LNR	9 MOD	7 MOD
21F	11	8	PVT (D)	36.1	6	OBL	10	7 IND	6 SEV	IF
21F	10	8	PVT	31.6	0	IV	—	5 IND, LNR	8 SEV	7 SEV
18M	2	5	PVT	30	6	OBL	20	4 IND	4 MOD	2 NML
18M	6	16	PVT	36.1	3	III	—	8 IND	4 MOD	3 MOD
27F	7	12	PVT	27	5	OBL	22	6 IND	11 SEV	4 NML
20M	4	10	PVT	44.6	2	III	—	6 IND, LNR	6 SEV	IF
16M	1	3	PVT	30	3	OBL	49	5 IND	6 MOD	2 NML
19M	3	7	PVT	30	9	OBL	4	5 IND	5 MOD	IF
13F	4	6	PVT	33	5	OBL	10	11 IND	7 MOD	8 SEV
15M	1	5	CB	27	4	OBL	9	7 IND, LNR	4 MOD	3 NML
23M	1	4	CB	37	6	OBL	28	10 IND	16 SEV	9 MOD
18M	2	12	CB	35	5	OBL	34	8 STONE	2 MOD	4 MOD
21M	14	3	CB (SRS)	35	3	OBL	29	8 IND, LNR	7 SEV	4 NML
25M	8	6	CB (SRS)	34.6	6	OBL	18	7 IND	8 SEV	7 SEV
34M	5	3	CB (SRS)	20	5	OBL	17	7 IND	4 SEV	3 SEV
24M	1	SPLX	SS (T)	—	3	OBL	38	2 IND	7 SEV	IF
29M	10	SPLX	SS (T)	—	6	OBL	27	7 IND	5 MOD	5 MOD
17M	11	13	SS (T)	38	6	OBL	20	5 IND	7 MOD	2 NML
Mean (S/D)	22 (6)	5.6 (3.8)	7.1 (3.8)	33.2 (5.9)	5.0 (1.5)	—	22.3 (12.9)	6.7 (2.1)	6.5 (3.0)	4.7 (2.3)

DOI=duration of illness; SPL=spleen; SPLX=splenectomy; SOB=site of block; PVT(D)=portal vein thrombosis (distal); CB=complete block of splenoportalaxis; (SRS)=splenorenal shunt; SS(T)=surgical shunt (thrombosed); SPP=splenic pulp pressure; EST=endoscopic sclerotherapy; Res=Result; OBL=obliteration of varices; MBD=main bile duct; IND=indentations; LNR=localised narrowing; LHD=left hepatic duct; RHD=right hepatic duct; SEV=severe changes; MOD=moderate changes; NML=normal; IF=inadequate filling.

Results

The papilla and the pancreatic duct were normal in both the groups of patients.

CONTROL GROUP

None of the patients gave any history of biliary disease and their liver function tests were normal. The main bile duct in this group was almost of uniform diameter in the upper, middle, and lower segments. The mean (SD) maximum diameter of the main bile duct was 4.8 (1.3) mm, which was significantly greater than the mean diameters of the left and right hepatic ducts (Table II). The left hepatic duct was wider than the right hepatic duct. The walls of the main bile duct, left hepatic duct, and right hepatic duct were smooth and regular. The intrahepatic ducts on both sides were smooth and had a regular branching pattern.

EXTRAHEPATIC PORTAL VENOUS OBSTRUCTION GROUP

Nineteen of 20 patients did not have any history of biliary disease. Only one patient had biliary colic, with a bilirubin concentration of 1.6 mg and an alkaline phosphatase activity of 219 IU (normal range 70–140 IU). This patient had two small stones in the bile duct. All other patients had normal liver function tests.

Main bile duct

Eighteen of 20 (90%) patients had abnormalities

TABLE II Mean maximum diameters (mm) of biliary ducts

	Control group (n=15)	Patients (n=20)
Main bile duct	4.8 (1.3) ^a	6.71 (2.03) ^d
Left hepatic duct	3.0 (1.08) ^b	6.5 (3.0) ^c
Right hepatic duct	2.6 (1.2) ^a	4.7 (2.3) ^e

a v b<0.001 d v e NS a v d<0.01
a v c<0.001 d v f<0.01 b v e<0.001
b v c<0.05 e v f<0.01 c v f<0.01

Figure 2: Indentations in the main bile duct (arrowed) due to choledochal varices and dilatation of the left hepatic duct simulating sclerosing cholangitis in a patient with extrahepatic portal venous obstruction.

confined to the mid portion of the main bile duct (Fig 2). All 18 had indentations suggestive of external compression by choledochal varices. Five patients had localised narrow segments in the main bile duct (Fig 3). The mean (SD) maximum diameter of the main bile duct was 6.7 (2.1) mm, which was significantly greater than in controls (Table II). In one patient the main bile duct was normal looking but there were two small stones in it, while another patient had abnormalities (narrowing and irregularity) confined to the lower portion.

Left hepatic duct and its branches

All patients had an abnormal looking left hepatic duct and branches. The abnormalities were as

Figure 3: Narrowed mid-segment of the main bile duct (arrowed) with irregular and dilated left hepatic duct in extrahepatic portal venous obstruction.

Figure 5: Indentations of the main bile duct (arrowed) with moderate changes in the left and right hepatic ducts in extrahepatic portal venous obstruction.

follows: focal narrowing, dilatations, irregular walls, and clustering of intrahepatic branches. All these changes were suggestive of sclerosing cholangitis. Severe abnormalities (Figs 2-4) were seen in 11 of 20 (55%) while nine (45%) patients had moderate changes (Fig 5). The mean (SD) maximum diameter of the left hepatic duct was 6.5 (3.0) mm, which was significantly more than in controls. Unlike in control subjects, the left hepatic duct in patients with extrahepatic portal venous obstruction was as wide as the main bile duct (Table II).

Right hepatic duct and its branches

The abnormalities were similar to the ones seen

in the left hepatic duct but with a lesser degree of severity. Moreover, these were not present in all patients. Four of the 20 (20%) did not have adequate filling of the right hepatic duct and hence results could not be analysed. Of the remaining 16 patients, severe changes were seen in three (19%), moderate changes in six (37.5%), and seven (43.5%) were normal. The mean (SD) maximum diameter was 4.7 (2.3) mm, which was significantly greater than in controls. However, it was significantly less compared with the left hepatic duct in these patients (Table II).

Cystic duct and gall bladder

Because of natural tortuosity of the cystic duct and odd mixing of contrast with the bile of the gall bladder, it was difficult to assess mild to moderate abnormalities. No major or obvious abnormalities were detected. None of the patients had gall stones in the gall bladder.

Figure 4: Dilated and irregular right and left hepatic ducts in extrahepatic portal venous obstruction.

Other features in patients with extrahepatic portal venous obstruction (Table I)

Other features in these patients were as follows:

- (a) Splenomegaly. The spleen was moderately enlarged and was palpable a mean (SD) of 7.16 (3.83) cm below the left costal margin.
- (b) Site of block in splenoportovenous axis. Three patients had distal portal vein thrombosis – that is, thrombosis of the portal vein at the junction of the left and right portal veins. Eight patients had complete portal vein thrombosis. Six patients had block at the splenoportal axis, and three of these had a spontaneous splenorenal shunt. Three patients had a thrombosed surgical splenorenal shunt. The extent of the block in the portal venous axis did not correlate with the severity of the biliary abnormalities seen in the main bile duct and left and right hepatic ducts. All patients had massive collateral veins

(cavernoma) at the porta hepatitis because of portal vein obstruction (Fig 1).

(c) Splenic pulp pressure. This was recorded in 18 patients. All patients had a raised splenic pulp pressure (normal values up to 15 mm Hg), the mean (SD) being 33.2 (5.9) mm Hg (range 20–44.6 mm Hg). The severity of biliary changes did not correlate with splenic pulp pressure.

(d) Oesophageal varices. All patients, except one, had undergone sclerotherapy. Oesophageal varices resolved in 17 of the 20 patients and required a mean (SD) of 5.05 (1.5) sessions of sclerotherapy. Two patients in whom varices did not resolve had received only two and three sessions of sclerotherapy respectively. The mean (SD) interval between resolution of varices and ERCP was 22.29 (12.9) months. The varices were either absent or thrombosed in the 17 patients in whom obliteration had been achieved, while they were large (grades III–IV) in three patients who still had them.

Discussion

Extrahepatic portal venous obstruction is a frequent cause of portal hypertension in India. In our recent analysis of 521 patients with portal hypertension, three main types were identified: 41% had cirrhosis, 40% had extrahepatic portal venous obstruction, and 18% had non-cirrhotic portal fibrosis.⁷ In over 90% of our patients with extrahepatic portal venous obstruction, the aetiology was not known.

We have, for the first time, shown biliary changes suggestive of sclerosing cholangitis in 100% of 20 consecutive patients with portal hypertension caused by extrahepatic portal venous obstruction. These changes were most noticeable in the left hepatic duct and its branches. They were seen in this duct in all patients, whereas only 56% had changes in the right hepatic duct. The mid portion of the main bile duct was affected in 90% of the patients. The biliary changes observed in this study did not relate to the degree of portal hypertension, the site of the block in extrahepatic portal venous obstruction, on splenomegaly. Resolution of varices probably had no influence on biliary abnormalities, as three patients without resolution (varices grades III–IV) had biliary changes similar to those in patients whose oesophageal varices had been successfully treated.

Portal hypertension secondary to biliary disorders such as primary cirrhosis and primary or secondary sclerosing cholangitis is well known. Biliary changes secondary to portal hypertension are extremely rare, however, and have been described only in case reports.^{1–4} Most of these reports are of patients with extrahepatic portal venous obstruction, because varices as a consequence of portal hypertension are much more common, prominent, and of longer duration in these patients than in those with cirrhosis.⁸ In support of this observation we have recently shown a significantly increased frequency of anorectal varices, large oesophageal varices, and variceal bleeding in patients with extrahepatic portal venous obstruction compared with those with cirrhosis.⁹ The finding of biliary changes possibly caused by choledochal varices in patients

with extrahepatic portal venous obstruction further supports this observation.

The biliary abnormalities can be explained by the presence of two types of venous system along the biliary tract. Firstly, the paracholedochal veins seen along the bile duct¹⁰ joining the gastric and pancreaticoduodenal veins to the portal veins in the liver. Secondly, the epicholedochal veins which form a network around the bile duct.¹¹ With obstruction in the main portal vein, as happens in extrahepatic portal venous obstruction, the paracholedochal veins may become prominent and cause indentations on the bile duct. The enlargement of epicholedochal veins may cause irregularity in the walls of the bile duct which may look like cholangitis.

The type of abnormalities seen in the bile ducts, particularly in the left hepatic duct, are similar to the radiological findings in sclerosing cholangitis. Only one of these patients, however, had any evidence of either cholangitis or biochemical abnormalities of cholestasis. The radiological abnormalities were noticeable in the branches of left hepatic duct and this may be due to the formation of prominent collateral veins where the umbilical vein joins the left branch of the portal vein.

One study has reported somewhat similar changes caused by regenerating nodules and severe fibrosis in the intrahepatic bile ducts of patients with cirrhosis.¹² In their study, however, the common bile ducts was normal, unlike that in our patients almost all of whom showed indentations. Moreover, intrahepatic bile changes in extrahepatic portal venous obstruction are certainly not caused by regenerating nodules and severe fibrosis, as these are almost always absent in this disorder. Abnormalities in the common duct similar to the ones seen in extrahepatic portal venous obstruction may be caused by lymphoma, biliary tumours, and parasitic infestations.^{2,3} None of our patients had these conditions.

Choledochal varices in a patient with portal hypertension are very important from a surgical point of view. They prevent satisfactory exploration of the common bile duct, which at the time may even be difficult to recognise, and these varices can be mistaken for the bile duct.³ Patients like these who have bile duct stones may be candidates for non-surgical therapy. Rarely, the varices may cause haemobilia after liver biopsy or transhepatic cholangiography.^{13,14} Jaundice secondary to the obstruction of the bile duct by enlarged choledochal varices occurs rarely. This phenomenon was first described by Fraser and Brown in 1944,¹⁵ and quite recently by Choudhary *et al*,¹⁶ who showed improvement of jaundice and the cholangiogram after successful shunt surgery in a patient with extrahepatic portal venous obstruction. Of the 213 patients with extrahepatic portal venous obstruction seen in our centre, only two had jaundice in addition to the problems of portal hypertension. Both these patients had multiple stones in both the intra- and extrahepatic bile ducts.

None of our patients, except one who had stones in the common bile duct, had raised bilirubin or alkaline phosphatase values despite narrowing of the bile ducts. We presume that

this could be a result of the intermittent forceful contractions of gall bladder in response to the ingestion of food which cause an increase in the intraluminal pressure above that of the pressure in the choledochal varices and thus prevent stasis in the flow of the bile. Moreover, the biliary abnormalities are patchy and mainly confined to the left duct, which may allow the bile to flow through other biliary channels.

In conclusion, biliary abnormalities simulating sclerosing cholangitis are very frequent in patients with extrahepatic portal venous obstruction. We believe these changes are caused by portal cavernoma/choledochal varices. These observations are important in the differential diagnosis of sclerosing cholangitis and also before considering these patients for biliary tract surgery.

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