Bile Duct Varices "Pseudo-Cholangiocarcinoma Sign" in Portal Hypertention Due to Cavernous Transformation of The Portal Vein

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Summary: *During the period 1986-1991, a total of eight hundred thirty two patients with portal hypertention resulting from different etiology were studied by sonography as a secreening test. In 17 patients, cavernous transformation of the portal veins was detected by means of ultrasonography, and the diagnosis was confirmed by arterial portography or splenoportography with DSA. 6 of seventeen patients underwent surgical operation for splenectomy because of severe hypersplenism and bleeding from esophageal varices.*

To evaluate how much biliary tract has been effected from cavernous transformation of the portal vein which is a rare condition characterized by portal hypertension and portal vein occlussion with recanalization and/or collateral vein formation to bypass the obstruction, and to explain the cause of mildly increased alkaline phosphatase and biluribin levels too, a 17 patients with portal vein cavernous transformation were studied. ERCP was performed in sixteen patients, there were narrowing, irregularity and nodular extrinsic defects mimicking cholangiocarcinoma spreading along the common bile duct (CBD) resulting from compression of thrombosis of the portal vein and collateral circulation on biliary tract, but there were not found same findings in 6 patients with portal hypertension due to liver cirrhosis.

Key Words Portal vein, cavernous transformation, varices, Behçet's disease, pseudo-cholangiocarcinoma.

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Cavernous transformation of the portal vein is a rare condition resulting from extrahepatic portal vein thrombosis or obstruction with recanalization and/or collateral vein formation to by pass the obstruction. Despite this disease is heterogenous in etiology (1-3), the etiology remains obscure in about a half of the patients. It is prevalant in developing countries (4). It is obvious that cavernous transformation around the occluded portal vein or in the hepatic hilus is seen in patients with portal venous occlusion of any etiology.

The normal portal vein is amenable to be studied by ultrasound scanning (5). Ultrasonography has been used to evaluate the portal venous system for along time and ultrasonographic recognition of the portal vein is achieved in 97% of cases (5-7). Sonography is the procedure of choice for the detection of many abnormalities of the portal vein system, mostly the diagnosis of this disease has been made by sonography and confirmed by angiography, CT or surgery (8-10). There are a few studies in the literature about biliary tree system's involvement by cavernous transformation of the portal veins (11,12).

During the past 6 year, we have studied different modalities of 832 patients with portal hypertension due to different etiologic factors and cavernous transformation of the portal vein secondary to portal vein thrombosis was diagnosed in seventeen patients. The diagno-

Table I: Sex and Age Distrubition and The Results of Imaging Modalities of The with Cavernous Transformation of The Portal Vein

Cas.No.	Sex/Age	Follow-up(yr)	US	portography*	ERCP
1.	F/28	1	+	+	+
2.	M/32	3	+	+	+
3.	M/38	3	+	+	+
4.	M/35	5	+	+	+
5.	F/28	2	+	+	+
6.	F/29	4	+	+	+
7.	F/30	4	+	+	+
8.	M/34	5	+	+	+
9.	M/30	15	+	+	+
10.	F/27	3	+	+	+
11.	M/25	1	+	+	
12.	M/35	1	+	+	+
13.	F/28	3	+	+	+
14.	F/27	8	+	+	+
15.	F/27	4	+	+	+
16.	M/20	2	+	+	+
17.	M/17	3	+	+	+

US; ultrasonography,*; either with DSA or splenoportography
ERCP; endoscopic retrograte cholangio pancratography

sis was confirmed by angiography and surgery.

To evaluate how much biliary system has been effected by cavernous transformation and to explain mildly increased alkalen phosphatase and biluribin levels, ERCP was performed in the patients with cavernous transformation of the portal vein. ERCP revealed nodular extrinsic defects, narrowing and irregularity of the CBD, resulting from compression of thrombosis and periportal collaterals and suggesting segmental variety of primary sclerosing cholangitis, sclerosing carcinoma of the common bile duct, and chronic cholangitis.

PATIENTS and METHODS

During the period November 1986 to December 1991, we evaluated 832 patients with portal hypertension by using ultrasonography as a screening test. Splenoportography, CT, surgery and peritonoscopy were done for further evaluation. Scans were obtained by using a commercially available, real-time sonograph (Toshiba SAL-90) at 3.5 MHz. Patients were fasted and examined in the supine position during normal respiration. The portal vein system, the patency of the intrahepatic division of portal vein, common bile duct, pancreas and vascular structure of the portal hepatis were evaluated carefully. In seventeen patients (9 male, 8 female; mean age 30 years) cavernous transformation of the portal vein was diagnosed by sonographically. To confirm the diagnosis, either splenoportography or arterial portography with DSA was performed to all 17 patients, by using a Phillips DVI divice. A "side winder" catheter was introduced via the femoral artery. DSA of the celiac and superior mesenteric artery were obtained following selective injection of 35ml of 50% diluted contrast medium (Omnipaque 300) at a rate of 8cc/second. Digital images were recorded for 25 second at a rate of one frame/second during the arterial, arteriocapillar and venous phase.

6 of the seventeen patients underwent splenectomy because of the hypersplenism and excessive bleeding from esophageal varices.
Sonography guided liver biopsy was done in all patients for the evaluation of paranchimal liver diseases. Beside the routine investigation of blood and urine analyses, serial liver function tests including serum alkalen phosphatase and bilirubin levels and tumor markers such as alpha feto protein (AFP), Carcinoembryonic antigen (CEA) were detected. HBV (Hepatitis B virus) and HDV (Hepatitis D virus) markers and all the coagulation parameters including protein c activity were tested too. After confirming the diagnosis of cavernous transformation, ERCP was performed to 16 patients by using a side-viewing duodenoscop (Olimpus JF-IT 20). After cannulation of the papilla of Vater, a contrast material, conray-60 was injected through the cannula under flouroscopic control and the radiograms of the biliary and pancreatic ducts in different position were taken. When the tip of the cannula was introduced into the papilla, both ducts were filled simultaneously in most pa-

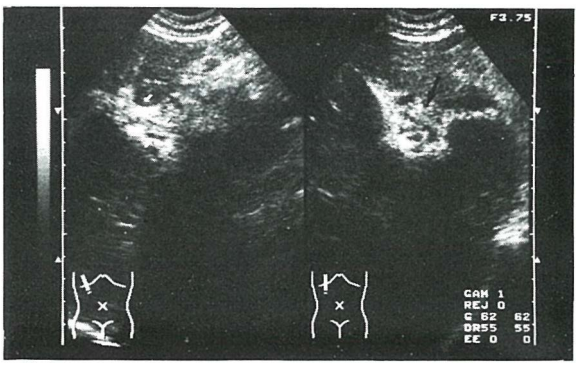

Fig. 1. Real-time sonogram shows at the level of porta hepatis, multiple echogenic bands and anechoic replacing the main portal vein (arrows).

Fig. 2. Arterial portography reveals on venous phase, many worm-like tortuous veins extending along the portal vein to portal vein radicles in the liver (arrows).

tients. Repositioning of the cannula was required for demonstration of the duct which was not filled initially. The following data were recorded: the appearance of the CBD and left and right hepatic ducts, narrowing or irregularity of biliary system, any sign related to compression. ERCP was also performed in 6 patients with portal hypertension due to liver cirrhosis.

RESULTS

All cases showed on US absence of a normal portal vein lumen, no demonstrable intrahepatic branches, multiple tortuous channels surrounding an hyperechoic cord, which run the course of a normal portal vein (Fig. 1). Sonographic pattern of the liver paranchima was normal and there wasn't any findings related to chronic pancreatitis. All the liver biopsies demonstrated no paranchimal abnormalities, but one. Arterial portography with DSA (in 7 patients) and splenoportography (in 10 patients) demonstrated extrahepatic portal vein occlusion with filling of multiple tortuous collateral vessels which replaced the main portal veins. In all seven patients, performed arterial portography at the venous phase revealed that veins over the head of pancreas drain into many tortuous, wormlike veins extending along the portal vein to portal veins radicles in the liver (Fig 2).

ERCPs demonstrated numerous nodular extrinsic defects, narrowing and irregularity along the CBD, mimicking cholangiocarcinoma (Fig. 3-8) without any evidence of intraluminal filling defect. There was not any evidence of ampullary or biliary tract neoplasm on this examination. We like to propose to call these findings "pseudo-cholangiocarcinoma sign". In 6 patients who underwent surgical operation, multiple collateral venous channels were seen adjacent to the common bile duct which confirmed the findings seen on ERCPs, US and portography. ERCP findigs were normal concerning CBD and left and righ hepatic duct in patients with portal hypertension due to liver cirrhosis.

There was no history of pancreatitis, umbilical vein catheterization during perinatal period, previous abdominal infection, trauma and alcohol abuse and other diseases known to cause portal hypertension. But all patients' social levels were lower or middle class. Umbilical sepsis might be an important factor to create portal vein occlusion. Two patients had Behçet's disase (BD): one has been followed-up and doing well, the other died of vascular complication of BD and we could not perform ERCP. One had liver cirrhosis in stage A.

Liver function tests except serum alkalen phosphatase and bilurubin AFP and CEA levels were normal. HBV and HDV markers were negative. There was a mild increase in serum alkaline phosphatase and bilurubin levels in all patients during course of the disease. The tests showed a fluctuation from normal to mildly increased levels. All coagulation tests were normal.

DISCUSSION

The etiology of the cavernous transformation of the portal veins is not clear. Majority believes that it represents recanalization of the occluded portal vein caused by great variety of diseases those are associated with periportal collateral developement. Some belive that it may be a rarely congenital anomaly. The occlusion has a vide variety of etiologies such as omphalitis, pancreatitis, alcoholic cirrhosis, and hepatic carcinomas (13-15). Although any underlying cause couldn't be determined in 14 of the seventeen patients, the history of splenomegaly since childhood imposed that umbilical sepsis or omphalitis might be an etiologic factor. In the two of the remaining three cases, Behçet's disease (BD) was the main etiologic factor, in which vasculitis is the major finding on histologic studies. One of them died of severe vascular complication of BD (16). Finally last one had liver cirrhosis.

Sonographic features of the disease have been well described (9,17-21). In the previous studies, the diagnostic findings are the following basicly: Failure to visualize the extrahepatic portal vein, the demonstration of high-levels echoes in the area of the porta hepatis, and finally direct visualization of multiple tubular structures in the porta hepatis. Although all of our patients fulfilled these three criteria, the diagnosis was confirmed angiographically. In our all 17 cases, sonographic diagnosis was confirmed by portography. It proves that sonography has a high accuracy in the diagnos-

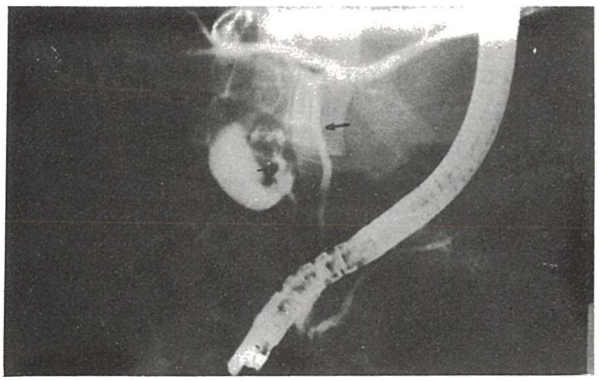

Fig.3. ERCP demonstrates narrowing and extrinsic compression along the common bile duct (arrows). Note; intrahepatic ducts are larger than CBD.

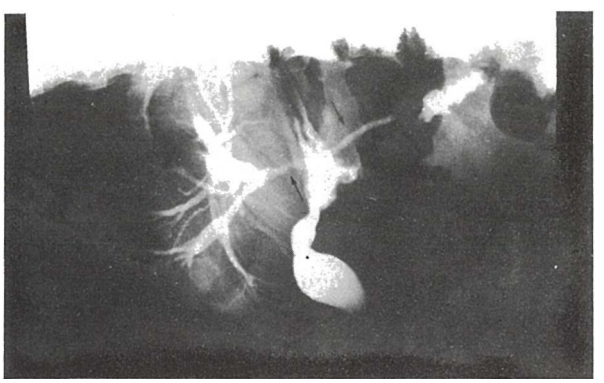

Fig.4. ERCP reveals narrowing, irregularity and ondulation along the CBD (arrows).

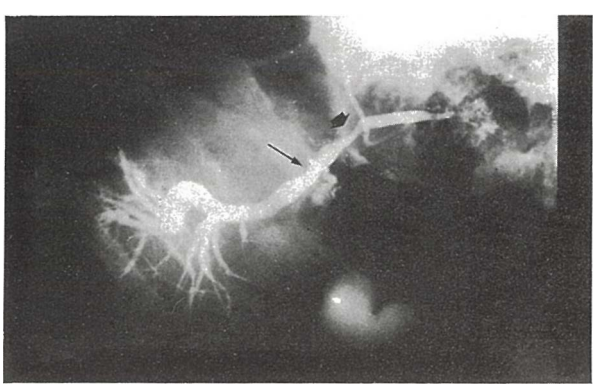

Fig. 5. ERCP demonstrates narrowing and irregularity and asymetric compression on CBD (arrows).

ing of portal vein pathology.

Extensive collateral veins due to portal vein occlusion may compress and narrow the biliary tract and may cause obstructive jaundice occasionally (11,22-23). There are two venous systems along the extrahepatic biliary tract. The paracholedochal veins (24) are separate structures from the bile ducts, and the epicholedocal venous system (25) is a fine reticular mural structure in intimate contact with the outer surface of the common bile duct. The veins of the plexus are relatively small in size, not larger than 1mm. Dilatation or varices of the plexus due to portal vein obstruction alter the normally smooth intraluminal surface of the common duct and produce irregular mural changes. On the other hand, the varices of the paracholedocal veins cause extrinsic impressions (Fig. 8). The cholangiographic demonstration of choledocal varices was first described by Williams et al (26) in 1982; since then, six additional cases have been reported (12,27-28). In these reports, main findings were narrowing, irregularity and nodular extrinsic compression. It is obvious that these findings resulted from the compression of paracholedocal varices and the presence of the varices of epicholedocal venous plexus. As seen in figures, (Fig. 3-8) the narrowing and the irregularity are very prominent along the extrahepatic biliary tract in our cases, suggesting not only bile duct varices but also cholangiocarcinoma which spreads along the bile duct and through its wall. For that reason, we proposed a name such as "pseudocholangiocarcinoma sign". In the literature, two of the seven cases with choledocal varices, (12,26-29), had cavernous transformation of the portal vein. We reported here seventeen additional cases in which the diagnosis were proved angiographically, six of them anatomically.

Beside cholangiocarcinoma, these rontgenographic findings of the common bile duct may

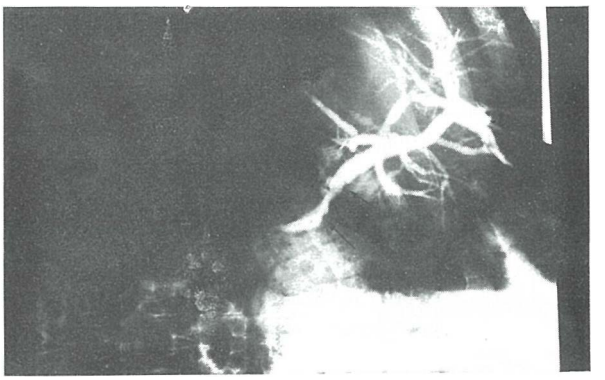

Fig. 6. ERCP demonstrates narrowing and irregularity on the distal part of CBD (arrows).

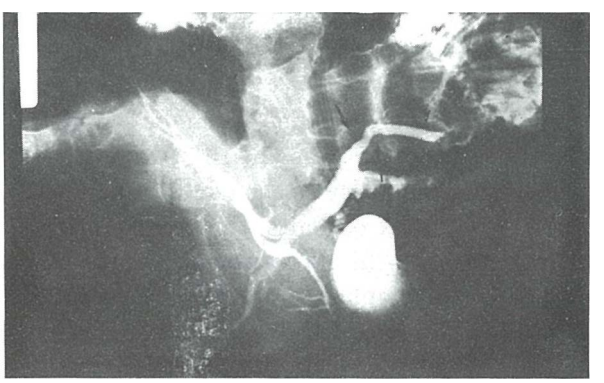

Fig.7. ERCP shows irregularity, narrowing and ondulation at the distal part of CBD (arrows).

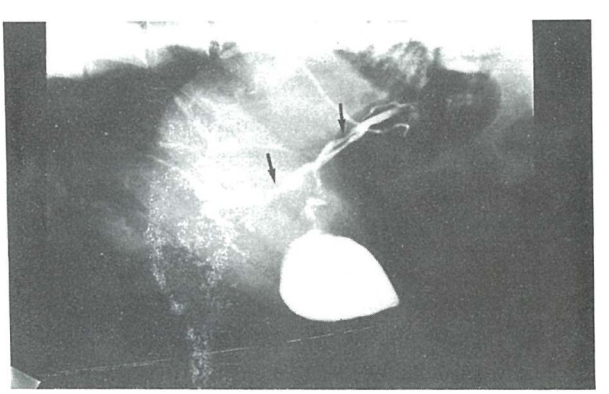

Fig.8. ERCP reveals narrowing irregularity and nodular extrinsic defects along the CBD (arrow).

be caused, by lymphoma, or by any submucosal tumors. Extrinsic lesions such as lymph nodes, vessels crossing the bile duct may also cause similar defects. Rarely calculi may become adherent to the wall and may simulate intraluminal lesions. Parasitic organisms (30) may inhabit in the biliary tree and cause nodular filling defects in the common bile duct.

In most patients, there were mild to moderate alkaline phosphatase and bilirubin elevation during their natural course. Chronic incomplete obstruction might be the main reason for these laboratory findings. The same laboratory findings and cholangiographic appearances can be seen in chronic cholangitis, benign strictures, chronic pancreatitis, segmental variety of primary sclerosing cholangitis (31-32) and especially in cholangiocarcinoma which spreads along the bile duct (33). Mean follow-up period was about 3yr for our cases. During this period, there was not any attack of cholangitis or any other disease. Additonally, serial liver function tests and liver biopsies were normal.

We conclude that ERCP findings of our thirteen patients with cavernous transformation of the portal vein on extrahepatic biliary system are related to compression of choledochal varices and mimick many common bile duct diseases particularly cholangiocarcinoma spreading along the bile duct, and considering our cases of chronic portal vein cavernous transformation we propose a name "pseudo-cholangiocarcinoma sign" to explain these radiologic appearance.

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