CASE REPORT

Aneurysmal dilatation of the portal vein: A rare cause of portal hypertension

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Abstract

Aneurysmal dilatation of the portal vein (ADPV) is a rare cause of portal hypertension. We described a case of ADPV in a female patient who presented with ascites. Imaging studies revealed tortuosity and dilatation of the main portal vein with turbulent flow. Endoscopy revealed oesophageal varices. A liver biopsy showed no abnormalities in liver histology. This is the first case of ascites as a complication of ADPV in the absence of liver cirrhosis, arteriovenous fistula or documented portal vein thrombosis. Hyperdynamic circulation and increased portal vein flow could be implicated in the pathogenesis of ascites in this setting.

Key Words: Aneurysmal dilatation of the portal vein, ascites, portal hypertension

The diameter of the portal vein ranges from 0.64 to 1.21 cm (mean 0.89 cm) [1]. A diameter >2 cm can therefore be considered as an aneurysm [2]. Aneurysmal dilatations of the portal vein (ADPV) represent only 3% of all aneurysmal dilatations of the venous system [3]. This is a rare finding occurring in 1 in 1500 patients with hepatobiliary disease and in 1 in 150 referred with portal hypertension [1]. ADPV has also been described in association with hereditary hemorrhagic telangiectasia [4].

Case report

A 45-year-old female presented with abdominal distension. Physical examination revealed ascites and splenomegaly but no other stigmata of chronic liver disease. Twelve years ago, cholecystectomy was done because of acute cholecystitis. She had no previous history of liver disease or excessive alcohol consumption. Serology for hepatitis B and C was negative. Antinuclear, anti-smooth muscle and antimitochondrial antibodies were negative. Standard blood tests, except for anemia (Hb 9 g/dl), were

Figure 1. Color Doppler ultrasonography: aneurysmal dilatation of the portal vein aneurysm with turbulent flow.
within normal limits, including normal liver function, coagulation studies as well as serum electrophoresis and albumin. Paracentesis of ascitic fluid revealed a transudate with sterile cultures. Endoscopy visualized grades I and II esophageal varices. Thyroid function was normal and tuberculin test was negative.

Color-coded Doppler ultrasonography revealed tortuosity and dilatation of the main portal vein with turbulent flow (Figure 1). A biphasic spiral computed tomography of the upper abdomen demonstrated homogeneously enhanced dilated portal vein in vein phase with tortuosity. The size of dilatation was 2 cm (Figure 2A, B). No mass in the ovaries and pancreas was visible. The same findings were seen in contrast-enhanced magnetic resonance angiography and digital subtractive angiography (Figure 3A, B). A guided liver biopsy revealed no fibrosis, granulomata or other abnormalities in liver histology. An aneurysmal dilatation of the main portal vein causing portal hypertension was diagnosed and the patient started on diuretics and beta-blockers with disappearance of ascitic fluid. Three months thereafter diuretics were withdrawn without recurrence of ascites.

Comment

ADPV or portal vein aneurysms are classified as extrahepatic and intrahepatic. Extrahepatic aneurysmal dilatations are the largest and the most frequent. The most common locations are the main portal vein, the splenomesenteric venous confluence and the intrahepatic portal branches at bifurcation sites [3]. The ADPV in the current case was localized in the main portal vein and was fairly small (2 cm), while the mean diameter of extrahepatic cases was larger (37.3 ± 7.7 mm) [3].

Portal aneurysms may be of acquired or congenital origin. Portal hypertension secondary to liver dis-
ease, necrotizing pancreatitis, abdominal surgery or trauma have been considered predisposing factors for ADPV [5]. Our patient had a past medical history of cholecystectomy that would have induced portal vein malformation. However, there are no medical reports available about the procedure itself and the possible complications of the postoperative period. Alternatively, a congenital origin of the aneurysm could not be excluded.

Most ADVPs are asymptomatic and are discovered incidentally. Complications of ADPV include abdominal pain, thrombosis, rupture, compression of the common bile duct and portal hypertension [5]. Altered flow patterns induced by the aneurysm may be the cause of portal hypertension, which, in the absence of cirrhosis, portal vein obstruction or arteriovenous fistula, was found in 2 of 11 cases (18%) in the literature [1]. Because portal hypertension is seldom found in a rare condition such as ADVP, the mechanism behind this complication is not well studied. Increased values of portal flow volume and flow velocity have been measured by Doppler ultrasonography in a patient with ADVP [6]. However, no hemodynamic study of portal venous pressure in ADVP has been reported in the literature so far.

No cause of secondary portal hypertension was found in our case, since histological examination of the liver was normal and no portal vein obstruction was shown. Thus, pre-hepatic portal hypertension should be considered as a complication of ADPV. The presence of ascites in the present case could not easily be elucidated in the absence of cirrhosis or portal vein occlusion. Hyperdynamic circulation due to tortuosity of the portal dilatation and turbulent portal vein flow which was further deteriorated by anemia was the most plausible explanation for the presence of ascites. Besides lowering the portal pressure, beta-blocker treatment ameliorates hyperdynamic circulation [7] and could account for the improvement in the clinical condition of our patient.

In conclusion, ADPV should be considered a rare cause of portal hypertension. Ascites is not a well-recognized complication of ADPV. However, this report shows that ascites may be associated with ADPV in the absence of liver cirrhosis, arteriovenous fistula or documented portal vein thrombosis. Hyperdynamic circulation and increased portal vein flow could be implicated in the pathogenesis of ascites in this setting.

References