Endoscopic ultrasound diagnosis of an ERCP-related portobiliary fistula

A 69-year-old man, who had undergone a laparoscopic cholecystectomy 5 years previously, presented with episodic right upper quadrant pain accompanied by vomiting and sweating. Computed tomography (CT) showed two stones in a mildly dilated common bile duct (CBD). At endoscopic retrograde cholangiopancreatography (ERCP), wire-guided biliary cannulation was rendered challenging by a large periampullary diverticulum but was eventually achieved, revealing two stones in the distal CBD. A small biliary sphincterotomy was undertaken that was followed by a small, self-limiting bleed. An attempt to extract the stones using a balloon was unsuccessful owing to increasing patient restlessness and was complicated by dislodgment of the cannulation wire from the CBD. Biliary recannulation proved difficult and the procedure was terminated after deployment of a 4-cm, 7-Fr, double pigtail, plastic biliary stent (Fig. 1). The patient experienced troublesome abdominal pain and sweating after the procedure, requiring a brief hospital admission 3 days later. However, physical examination and a plain X-ray were unrevealing, laboratory investigation showed only a C-reactive protein of 61 mg/L, and the pain settled with conservative management. The patient underwent a further ERCP 5 weeks later during which removal of the biliary stent was complicated by a modest, spontaneously resolving bleed. Wire-guided cannulation at the previous sphincterotomy site revealed only faint, rapidly dissipating, biliary-like opacification despite several contrast injections (Fig. 2). The procedure was abandoned and, although the patient remained entirely asymptomatic thereafter, an urgent CT scan was performed. This disclosed gas shadowing in the portal vein extending to its peripheral intrahepatic branches, a thrombus in the main portal vein, and a small amount of peripancreatic and periportal free gas in keeping with a localized perforation (Fig. 3). A pancreaticobiliary endoscopic ultrasound (EUS) performed 8 days later demonstrated evidence of a communication between the portal vein and CBD, the two structures being inseparable at the level of the mid-duct in keeping with a portobiliary fistula (Video 1). However, no blood flow could be detected within the CBD, possibly due to the presence of a thrombus, albeit nonocclusive, within the portal vein. In addition, multiple CBD stones were seen (not shown). A CT scan carried out 2 months later confirmed spontaneous resolution of all portal vein abnormalities and extraluminal free gas. Complete biliary clearance was accomplished at subsequent ERCP (Fig. 4).

Portal vein opacification is an exceedingly rare event during ERCP with an estimated incidence of 1 in 6000–8000 [1]. First reported by Huibregts et al. in 1988 [2], it has since been described in several case reports, mostly, but not exclusively, in the setting of attempted biliary cannulation following needle-knife precut or biliary sphincterotomy [3]. However, to our knowledge, the existence of a portobiliary fistula per se has hitherto been demonstrated at ERCP in only three cases, two of which having been thought to be secondary to multiple liver abscesses [4,5] and the third occurring in the setting of a presumed portal vein tumor thrombus [6]. The case reported here represents the first to be documented by EUS. Other, more common, causes of portobiliary fistulas include percutaneous transhepatic cholangiography, choledocholithiasis, biliary surgery, trauma, and percutaneous liver biopsy [6]. Demonstration of a portopancreatic fistula at ERCP has also been described in the setting of pancreatic pseudocyst or malignancy [7]. The more general finding of hepatic portal venous gas on plain X-rays, CT or doppler ultrasound has a wide differential diagnosis including, in addition to portobiliary fistula, necrotizing enterocolitis (in neonates), mesenteric ischemia, intra-abdominal sepsis (particularly diverticulitis), acute gastric dilatation, gastric emphysema, inflammatory

Fig. 1 Endoscopic retrograde cholangiopancreatography (ERCP) view showing a deployed pigtail stent.

Fig. 2 Endoscopic retrograde cholangiopancreatography (ERCP) view showing portal vein opacification (arrows).

Video 1
Linear pancreato-biliary endoscopic ultrasound scan showing a portobiliary fistula (PBF) and a nonocclusive portal vein (PV) thrombus. The common bile duct (CBD), main pancreatic duct (PD), and head of pancreas are also shown.

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bowl disease, liver transplantation, chemotherapy, and a variety of other endoscopic procedures [8].

Clinically, hemobilia and right upper quadrant pain are the most common manifestations of portobiliary fistulas [6] although previously reported cases of portal vein opacification at ERCP, with or without proven portobiliary fistulas, have all been associated with surprisingly little bleeding [9]. Other potential clinical features include those of sepsis, portal vein thrombosis, and air embolism.

Management may entail surgical repair, thrombin injection, coil occlusion, simple or balloon tamponade, or transhepatic biliary or portal vein stenting [10, 11]. Typically, however, portal vein opacification in the setting of ERCP tends to have a favorable prognosis in the absence of further intervention [9] with spontaneous clearance of hepatic portal venous gas within as little as 4 hours [12], although this may merely reflect reporting bias. Indeed, a single fatality related to air embolism as a consequence of overlooking the diagnosis has been described [13]. Thus, immediate termination of the procedure is recommended and, to this end, prompt recognition of portal vein opacification as a radiographic pattern that could closely mimic that of biliary underfilling (with striking similarities between the two structures in location, orientation, and branching pattern) is imperative; perseverance in the event of misinterpretation may lead to potentially deleterious therapeutic misadventures such as transduodenal portal vein diathermy or stenting. Aspiration of blood rather than bile upon cannulation may facilitate a prompt diagnosis.

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