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Mind the Sump! – Diagnostic Challenge of a Rare Complication of Choledochoduodenostomy

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Key Words

Sump syndrome · Biliary tract disease · Cholangitis · Choledochoduodenostomy · Endoscopic retrograde cholangiopancreatography · Complication · Pneumobilia

Abstract

Sump syndrome is a rare long-term complication of side-to-side choledochoduodenostomy (CDD), a common surgical procedure in patients with biliary tract disease in the era before endoscopic retrograde cholangiopancreatography (ERCP). Frequently only pneumobilia, serving as sign for functioning biliary-enteric anastomosis, is reminiscent of the former surgery. We present the case of an 81-year-old patient with sump syndrome who presented with clinical signs of ascending cholangitis, decades after the initial CDD procedure. Finally the detailed medical history that was taken very thoroughly in combination with the presence of pneumobilia led to the suspicion of sump syndrome. Sump syndrome was diagnosed by ERCP, and after endoscopic debris extraction and antibiotic treatment the patient recovered quickly. In the ERCP era little is known about CDD and its long-term complications, especially by young colleagues and trainees. Therefore this report provides an excellent opportunity to refresh the knowledge and raise awareness for this syndrome.

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Case Presentation

We report the case of an 81-year-old female patient with a rare cause of ascending cholangitis. Initially she presented with loss of appetite, recurrent vomiting (2–3 times per

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week) along with epigastric pain and a large abdominal mass measuring 7.7 × 8.0 × 7.6 cm located in the transverse colon with contact to the ascending colon. Her past medical history included type 2 diabetes mellitus treated with premixed 70/30 insulin (neutral protamine Hagedorn insulin/regular insulin) twice daily, cholecystectomy and revision of the common bile duct (CBD) in the 1970s. Colonoscopy revealed an exulcerating, near-circumferential lesion in the transverse colon without stricture formation. Biopsy of the abdominal mass revealed diffuse large B-cell non-Hodgkin lymphoma (DLBCL). Widespread mesenteric, peritoneal and retroperitoneal nodes seen on positron emission tomography/computed tomography (PET/CT) scans as well as extranodal manifestations in the liver, left adrenal gland and pulmonary involvement finally led to the classification of DLBCL stage IV E. In order to attenuate the risk of perforation in case of rapid tumor shrinkage, a 'prephase' with vincristine (1 mg) and prednisone (1 mg/kg/day for 5 days) was initiated before immunotherapy with bendamustine (90 mg/m²/day, d1–d2) and rituximab (375 mg/m², d1) was scheduled. Two weeks after the first chemotherapy cycle, which she initially tolerated well, she complained of increasing right upper quadrant abdominal tenderness and vomited occasionally. A few days later she developed the full clinical picture of acute cholangitis with fever of 39.0°C, right upper abdominal pain, nausea and liverish vomiting and increasing serum gamma-glutamyl transpeptidase 536 U/l (normal <35), alanine aminotransferase 129 U/l (normal <55), aspartate aminotransferase 128 U/l (normal <40), alkaline phosphatase 421 U/l (normal 42–98), but normal serum bilirubin. Immediately empiric intravenous antibiotic therapy with ceftriaxone and metronidazole and fluid resuscitation were commenced. Abdominal ultrasonography revealed a slightly dilated CBD of 8 mm (within the normal range after cholecystectomy) as well as marked pneumobilia throughout the liver, but no signs of intrahepatic cholestasis. There was no free intra-abdominal gas as indirect sign for gastrointestinal perforation on CT scans, and retrospectively pneumobilia had already been present on the initial staging scan (fig. 1a).

Assuming that a side-to-side choledochoduodenostomy (CDD) had been performed several decades before in the context of revision of the CBD due to complicated cholecystectomy, an endoscopic retrograde cholangiopancreatography (ERCP) was performed to evaluate biliary drainage. The thorough endoscopic examination of the duodenum revealed a small orifice proximal to the papilla of Vater. Air insufflation into this orifice induced distinct pneumobilia, whereas contrast injection showed a normal proximal CBD and drainage of the contrast dye via the anastomosis, proving the presence of CDD and its functional efficiency. In addition, filling defects of the distal CBD (fig. 2a) revealed impaction of abundant debris. After sphincterotomy an organic, 'fruit skin'-like material could be extracted using a Dormia basket (fig. 2b).

To summarize, the findings of ascending cholangitis in the context of a former CDD procedure together with the endoscopic picture of filling defects in the distal CBD led to the diagnosis of sump syndrome.

After endoscopic debris extraction and antibiotic treatment the patient recovered quickly and immunochemotherapy could be continued.

Discussion

CDD in the Past and in the Present

In the pre-ERCP era CDD was a common surgical procedure in patients with biliary tract disease. In the setting of a side-to-side CDD the bile does not drain through the distal CBD anymore. Therefore the part of the CBD distal from the CDD anastomosis consequently

transforms into a poorly drained reservoir, making this so-called ‘sump’ prone to accumulation of debris (fig. 3).

Nowadays CDD is widely replaced by ERCP [1], but we are still faced with its consequences and long-term complications in elderly patients. Sump syndrome was first described in 1976, and a diverse prevalence ranging from 2.5 to 15.7% after CDD has been reported [2–5]. Sump syndrome is not precisely defined, but it results from accumulation of lithogenic bile, debris or calculi as well as refluxed duodenal contents in the distal CBD, leading to biliary and/or pancreatic complications. According to Marbet et al. [6] reduced filling pressure as well as reduced peristalsis and drainage of the distal CBD caused by the upstream anastomosis play an important role in the pathophysiology of sump syndrome. It can present with a variety of symptoms and clinical pictures such as recurrent pancreatitis, colicky-like pain, jaundice and cholangitis. Therefore the diagnosis of sump syndrome is challenging because no characteristic clinical or laboratory finding is highly specific. Taking a detailed past medical history and having a high degree of vigilance appear to be the most important elements for diagnosing sump syndrome. To increase the challenge, sump syndrome often becomes clinically manifest only several decades after the initial CDD procedure [2, 6]. At that point of time, generally the original medical records may no longer be available and patients may not know the details of the procedure anymore.

Nowadays in the ERCP era this kind of bile duct surgery is rarely performed and its complications, including sump syndrome, are almost forgotten, especially due to the long interval until clinical manifestation of sump syndrome occur after CDD [1]. Also we are faced with a comeback of this rare syndrome due to global migration of people from developing countries where CDD is still performed nowadays, with ERCP not being a standard procedure.

Gas in the Liver: Differential Diagnosis

In addition to a thorough past medical history, pneumobilia is another key element in establishing the diagnosis of sump syndrome. Generally the presence of gas in the liver may be a result of local gas formation by aerogenic bacteria [7] or can result from an abnormal biliary or portovenous connection to the intestines.

Gas formation by aerogenic bacteria may show diffuse parenchyma penetration or can be localized within an abscess. Hepatic portal venous gas (HPVG), a condition that was first described in 1955 [8], may result from intestinal ischemia, intra-abdominal abscess or inflammatory bowel disease. In HPVG the gas is localized predominantly in the periphery of the liver parenchyma because gas assumingly enters via the portomesenteric venous circulation and is randomly flushed into the liver by the hepatopetal blood flow (fig. 1b). On imaging the diagnostic features include portovenous air extending within 2 cm of the liver capsule [9]. Pneumobilia, defined as air in the biliary tree, occurs mainly due to carcinoma, mechanical or iatrogenic causes such as sphincterotomy and biliary-enteric anastomosis. In contrast to HPVG, which is located peripherally, pneumobilia is predominantly located in the central portion of the liver as a consequence of ‘hepatofugal’ bile flow preventing the ascending gas from migrating peripherally (fig. 1a). Therefore, pneumobilia after CDD serves as a sign for functioning biliary-enteric anastomosis.

Conclusion

To summarize, the diagnosis of sump syndrome is challenging because of a variety of symptoms and findings resulting from an unphysiological, artificial anatomy generated by

former CDD. Nowadays CDD as a common surgical procedure for benign biliary tract disease has been widely replaced by endoscopic procedures. In the ERCP era little is known about CDD and its long-term complications, especially by young colleagues and trainees. However, the incidence may increase again, especially due to migration of patients from developing countries where ERCP has not been widely available in the past. Therefore this case report and discussion provides an excellent opportunity to refresh the knowledge and raise awareness for this syndrome.

Disclosure Statement

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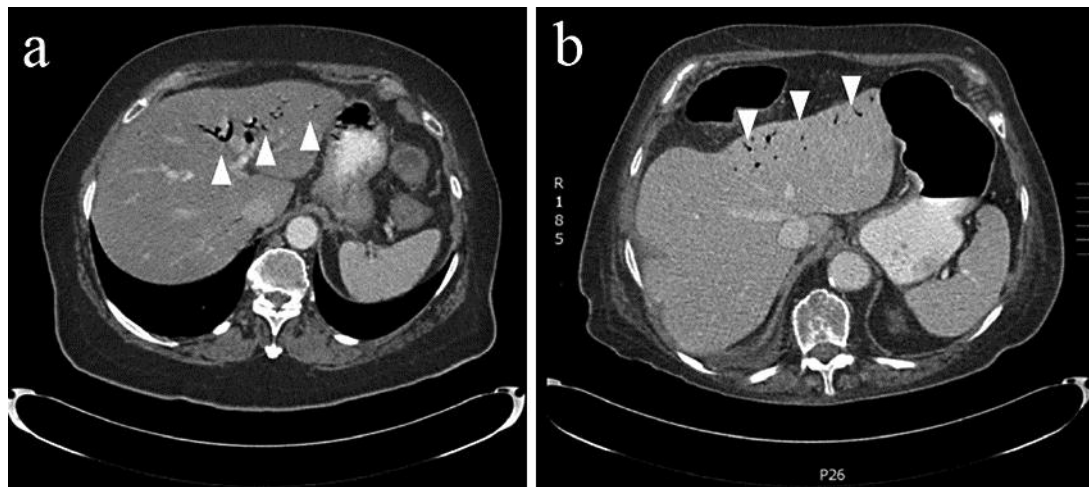


Fig. 1. CT scan of the abdomen. **a** Arrowheads show pneumobilia. **b** Arrowheads show HPVG.

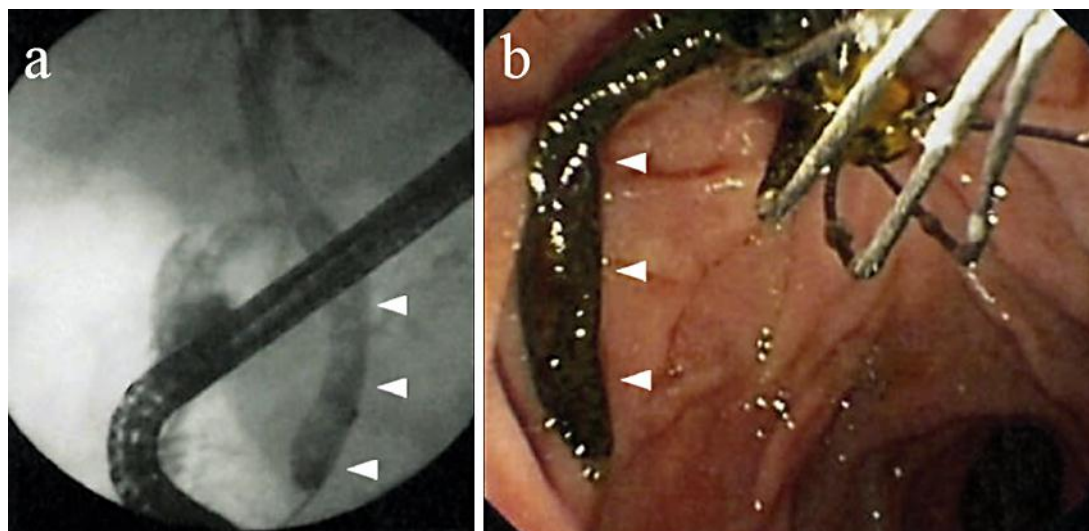


Fig. 2. **a** ERCP: Filling defects in the distal CBD (arrowheads) reveal impaction of abundant debris (for a schematic account see fig. 3). **b** Endoscopy: Dormia basket with 'fruit skin'-like material (arrowheads) extracted after sphincterotomy.

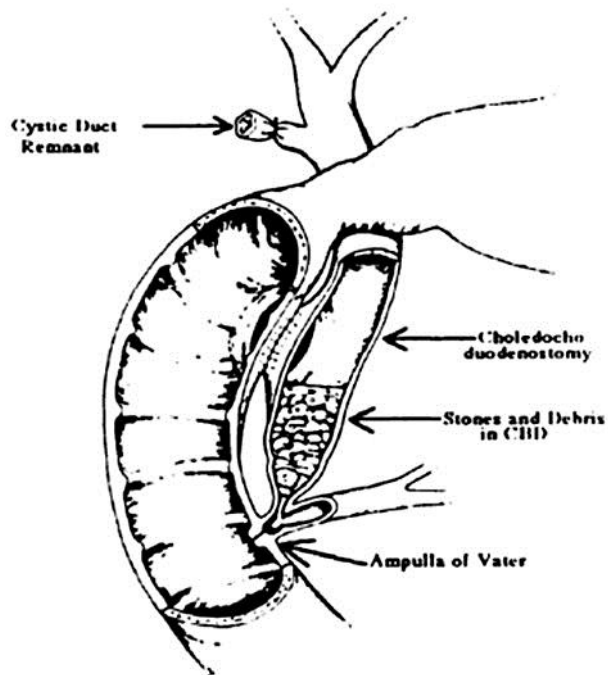


Fig. 3. Schematic drawing of CDD. The distal CBD is excluded from draining bile flow and consequently transforms into a poorly drained reservoir, functioning as a so-called 'sump'. When the sump gets symptomatic due to collected stones and debris it provides the basis for sump syndrome. Reprinted with permission from the American Journal of Roentgenology [10].