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CASE REPORT

Unusual cause of gastric outlet obstruction mimicking superior mesenteric artery syndrome: A case of infiltrative duodenal cancer arising from a choledochocele

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Abstract

Choledochocele is a rare subtype of choledochal cyst and is associated with increased prevalence of periampullary cancers. Here, we report an unusual manifestation of infiltrative duodenal cancer arising from a choledochocele, involving superficial spreading (muscularis mucosae) of cancer cells along the duodenum causing gastric outlet obstruction, which clinically mimicked superior mesenteric artery syndrome. Histologically, wide spread of cancer cells was confirmed from periampullary region to duodenojejunal junction showing mismatch with radiologic findings, in which the cancer segment was mainly located in the distal duodenum. Clinical, radiologic, and pathologic findings are discussed with literature reviews.

Introduction

Choledochal cysts (CCs) are rare congenital anomalies. It is more common in Asia (incidence of 1/13 000 births) than in western countries (incidence of 1/100 000 births).[1] Recently, increasing cases of CC in asymptomatic adults have been observed with the universal use of computed tomography (CT). Only 10% of CC cases are diagnosed in adults, and they most commonly present with jaundice or symptoms of cholangitis.[1] Pancreaticobiliary maljunction (PBM) in CC is associated with a higher incidence of biliary malignancy.[2] Choledochocele, type III of the five variants of CC, is the least common, comprising 1.4–4.5% of CCs. Of all choledochoceles, 14.3–27% harbor biliary malignancy.[3] We hereby report an unusual manifestation of diffuse infiltrative duodenal cancer arising from a choledochocele, causing gastric outlet obstruction, which clinically mimicked superior mesenteric artery (SMA) syndrome.

Case report

A 68-year-old man presented with nausea, vomiting, and epigastric discomfort lasting 2 days. The patient had no significant medical history except for hypertension. He had undergone screening endoscopy approximately 5 months ago, and the results were normal. Immediate CT was indicated and revealed a markedly distended stomach and proximal duodenum with abrupt narrowing of the third duodenal portion, no obstructing mass was visible (Fig. 1a). These findings mimicked those of SMA syndrome. However, left hydronephrosis due to peritoneal seeding with left periureteral metastasis was suspected. Therefore, subsequent magnetic resonance imaging (MRI) was performed. MRI

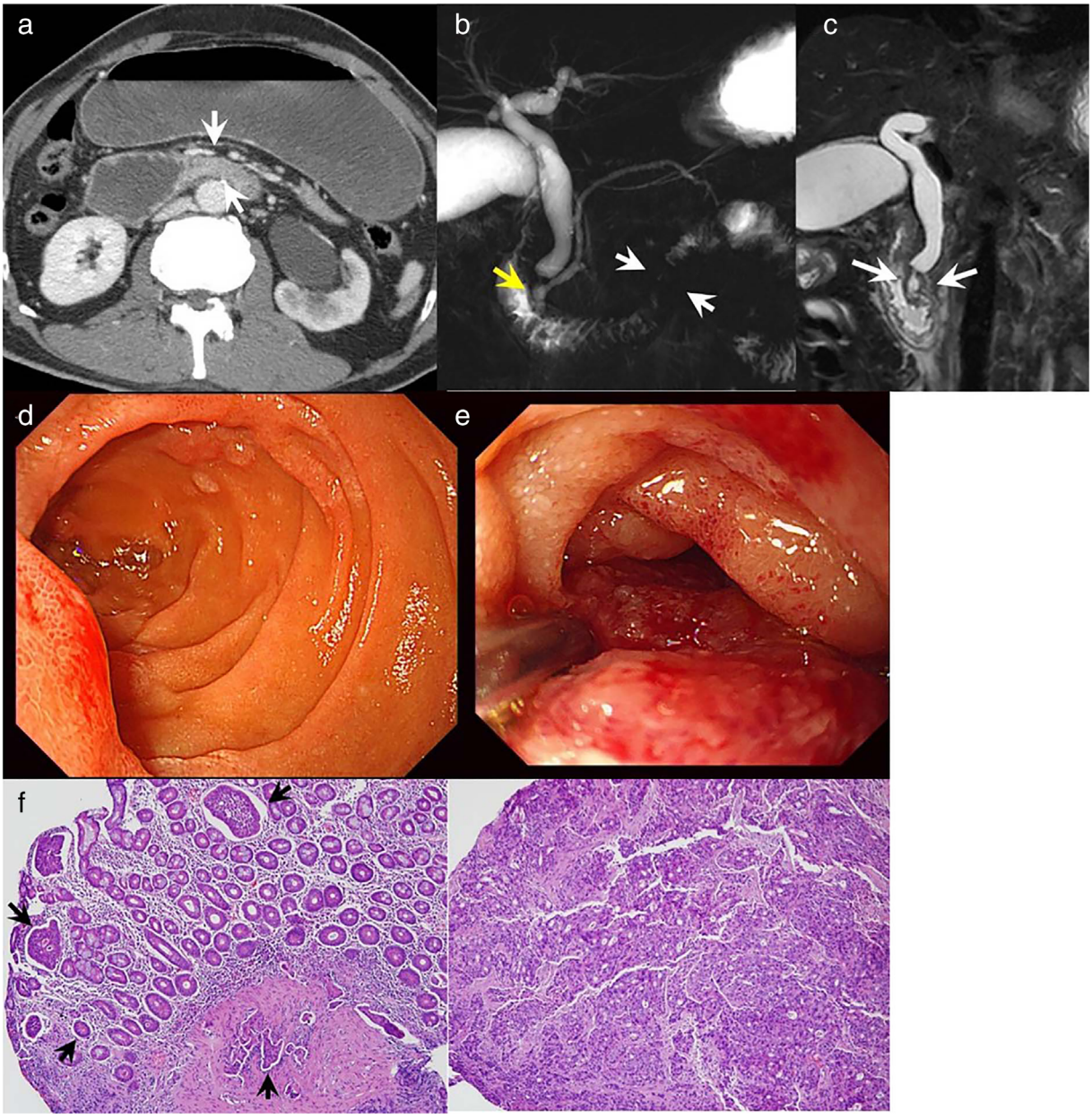

Figure 1 A 68-year-old man with choledochocele-associated duodenal cancer. (a) Axial contrast-enhanced CT image shows a markedly distended stomach and abrupt narrowing of duodenal at the horizontal portion in the aortomesenteric space (arrows). (b) Magnetic resonance cholangiopancreatography (MRCP) shows segmental wall thickening with stenosis of distal duodenum, which was indicative of cancer (white arrows) and reveals a choledochocele associated with a pancreaticobiliary maljunction (yellow arrow). (c) Coronal T2-weighted fat-suppressed image shows the thickened wall of the choledochocele, which was highly suggestive of cancer (arrows). Endoscopic images show (d), several mucosal nodular lesions scattered in the second duodenal portion and the ampulla of Vater (AOV) and (e), diffuse infiltrative mucosal lesions with stenosis in the distal duodenum. (f) Pathologic specimen (hematoxylin & eosin, ×100) shows cancer cells infiltrating the muscularis mucosa, submucosa, and muscle layers in duodenal second (right) and third portions (left) with frequent lymphovascular invasions (black arrows).

revealed segmental wall thickening between the third duodenal portion and the duodenojejunal junction with diffusion restriction, which was indicative of infiltrative duodenal cancer (Fig. 1b). Furthermore, a choledochocele, with a PBM was observed. The walls of the choledochocele at the common channel was thickened, which further raised suspicions of cancer (Fig. 1c). Endoscopy revealed sparse mucosal nodular lesions and prominent ampulla of Vater (AOV) in the second duodenal portion (Fig. 1d), as well as an infiltrative mucosal lesion with stenosis in the third duodenal portion (Fig. 1e). The distal

duodenum was not visible because the gastroscope could not pass through the stenotic portion. Multiple biopsies were performed in the periampullary area and the distal duodenum, which confirmed moderately differentiated adenocarcinomas. The tumors were mainly located in the muscularis mucosae, and frequent lymphovascular invasions were observed in the biopsy specimens (Fig. 1f). Despite extensive cancer infiltration in the duodenum, the distal duodenum was the epicenter of cancer, thereby causing a stenosis which initially mimicked SMA syndrome. The patient underwent a palliative bypass gastrojejunostomy and is currently undergoing chemotherapy.

Discussion

The malignant potential of all CCs is age-related, ranging from 1% in those less than 10 years of age, to 15% in those older than 20 years.[1] A malignancy risk of about 6–30% in adults has been reported. Furthermore, a lifelong elevated risk of up to 4% for cancer development persists even after cyst excision.[4] Patients with CCs are typically predisposed to cholangiocarcinoma and gallbladder carcinoma. In a multicenter study on Korean patients with CCs, about 10% of cases presented with biliary malignant tumors. Majority of the malignancies were of the bile duct and/or the gallbladder, and only three cases were of the periampullary region.[5] CCs are closely associated with PBM, through which pancreatic juice refluxes into the common bile duct.[6] Changes in biliary phospholipids as a result of the reflux may be responsible for inducing malignant transformation.[7]

Choledochocele is the least common and rare type of CC and is associated with increased prevalence of periampullary cancer, although lower than other types of CC.[8] A choledochocele may be lined by duodenal or biliary epithelium and the presence of biliary epithelium is believed to be a risk factor for malignant transformation.[9] Predisposition to cancer in choledochocele can be explained by the same mechanism as that leading to carcinogenesis in patients with PBM; bile containing amylase may stagnate in the choledochocele, causing mucosal irritation, metaplasia, and eventually malignant change in the inner epithelium of the choledochocele.[10] Reported malignant changes of choledochoceles include adenocarcinoma and superficial spreading cancer of the bile duct.[11]

Choledochocele in this case showed cystic dilatation of common channel (intraampullary region) that drains both bile duct and pancreatic duct, therefore, associated with PBM. Our case showed irregular wall thickening of the choledochocele, which was highly suggestive of malignancy and the origin of the superficial spreading tumor cells, which invaded almost the entire duodenum from the periampullary area to the duodenojejunal junction.

The treatment of a choledochocele depends on its size and lining epithelium. Uncomplicated choledochoceles with intestinal mucosa can be treated with endoscopic sphincterotomy and only require limited surveillance. However, choledochoceles lined with biliary epithelium warrant pancreaticoduodenectomy or cyst excision with indefinite surveillance.[11] The prognosis of patients with CC-originated malignancy is poor because of limited resectability.[5] In line with this, our case presented with symptoms of gastric outlet obstruction and found to have unresectable duodenal cancer with peritoneal seeding.

Choledochocele-originated duodenal cancer can be a rare cause of gastric outlet obstruction in elderly patients. Superficial spreading of cancer in the duodenum may present as gastric outlet obstruction without visible obstructing mass upon imaging, which clinically mimick SMA syndrome. Although the epicenter of tumor was the distal duodenum, wide range of cancer infiltration was from the periampullary area to the duodenojejunal junction. A diagnosis of duodenal cancer with periampullary infiltration was confirmed, and this was an unusual manifestation of malignancy arising from a choledochocele.

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