Endoscopic Corner

Thanapirom K
Angsuwatcharakon P
Miranda DA
Kimtrakool S
Rerknimitr R

CASE 1

An 86-year-old woman presented with progressive painless jaundice for 1 month. Physical examination revealed palpable gallbladder. Liver function test showed markedly elevated serum bilirubin (18.2 mg/dL). The computed tomography of the abdomen showed an enhancing soft tissue lesion occupying at the periampullary region with upstream bile duct dilation (Figure 1). The side-viewing duodenoscopy found a large ampullary mass. Endoscopic retrograde cholangiopancreatography (ERCP) revealed distal CBD stricture, 2 cm. in the length, causing upstream biliary dilatation (Figure 2A). A self-expandable metallic stent was placed across the stricture (Figure 2B). The histopathology of biopsy specimen from the ampullary mass revealed moderately differentiated adenocarcinoma. Because of her age, we did not offer her for any further treatment.

Diagnosis:

Ampullary adenocarcinoma

Discussion:

Ampullary tumors are rare, with an approximate 5% incidence of all gastrointestinal neoplasms[1]. Primary malignant ampullary tumors are adenocarcinoma, lymphoma, neuroendocrine, and signet ring cell carcinoma. Metastatic neoplasms include malignant melanoma, hypernephroma, and lymphoma. Of these malignant tumors, adenocarcinoma is the most common malignant ampullary lesion[2]. Side-viewing duodenoscopy and ERCP are important tools for an evaluation of all ampullary tumors including size, margin and extent of intraductal growth. There are several endoscopic appearances of ampullary tumors, endoscopic assessment alone sometimes may not be sufficient for

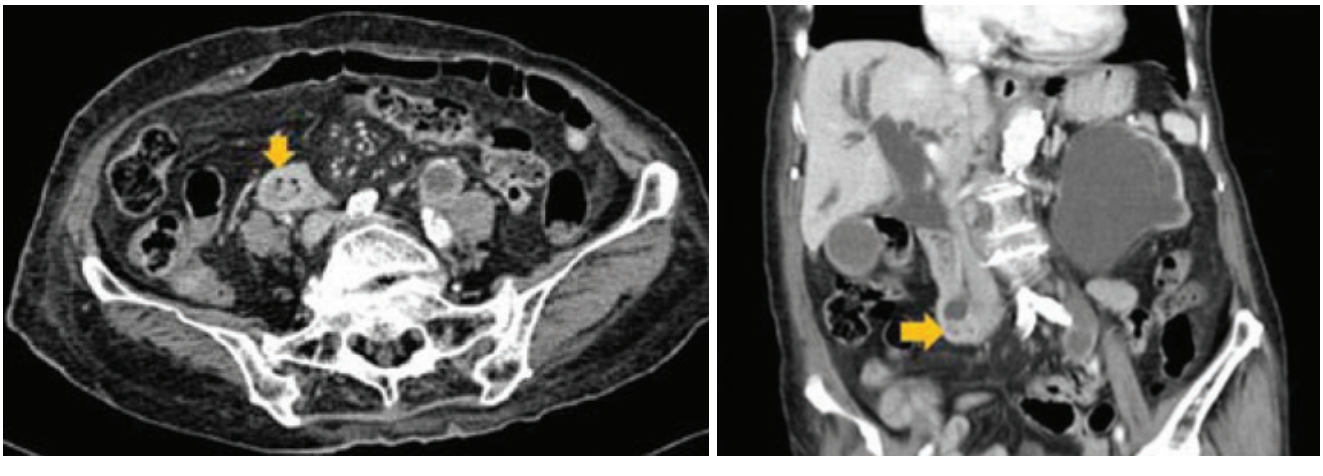

Figure 1. Computed tomography of the abdomen revealed an enhancing soft tissue mass at the peri-ampullary area (arrow) causing biliary obstruction.

Address for Correspondence: *Rungsun Rerknimitr, M.D., Division of Gastroenterology, Faculty of Medicine, Chulalongkorn University, Bangkok 10330, Thailand.*

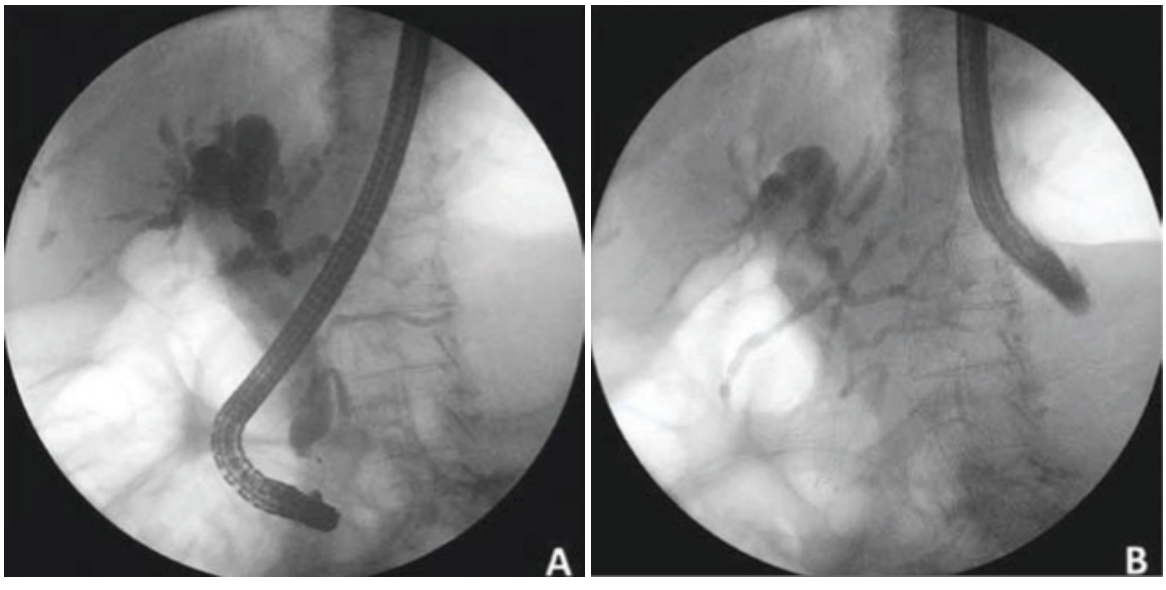

Figure 2. ERCP showed distal CBD stricture, 2 cm. in the length with upstream biliary dilation (A). A self expandable metallic stent was inserted across the stricture (B).

diagnosis. Tissue sampling is always required to establish the diagnosis. However, obtaining biopsy after endoscopic sphincterotomy may be difficult to interprete due to coagulated and necrotic tissue[3]. Endoscopic ultrasonography are helpful in tumor (T) and nodal (N) staging. The overall accuracy of T and N staging are 56-91% and 50-81%, respectively[4,5].

REFERENCES

1. Scarpa A, Capelli P, Zamboni G, *et al*. Neoplasia of the ampulla of Vater. Ki-ras and p53 mutations. Am J Pathol 1993;142:1163-72.
2. Huibregtse K, Tytgat GN. Carcinoma of the ampulla of Vater: the endoscopic approach. Endoscopy 1988;20 Suppl 1:223-6.
3. Bourgeois N, Dunham F, Verhest A, *et al*. Endoscopic biopsies of the papilla of Vater at the time of endoscopic sphincterotomy: difficulties in interpretation. Gastrointest Endosc 1984;30:163-6.
4. Artifon EL, Couto D, Jr., Sakai P, *et al*. Prospective evaluation of EUS versus CT scan for staging of ampullary cancer. Gastrointest Endosc 2009;70:290-6.
5. Tio TL, Sie LH, Kallimanis G, *et al*. Staging of ampullary and pancreatic carcinoma: comparison between endosonography and surgery. Gastrointest Endosc 1996;44:706-13.

CASE 2

A 22-year-old man presented with recurrent jaundice for one year. This time, jaundice recurred for a week but accompanied with fever and abdominal pain for two days. Liver function test revealed elevated transaminases, alkaline phosphatase and direct hyperbilirubinemia. CT scan of the abdomen revealed fusiform dilatation of the common bile duct and left intrahepatic ducts, no stone was noted (Figure 1). Cholangiography showed choledochal cyst type 4A (Todani classification) without filling defect, but bile sludge was extracted during balloon sweeping. Double pig tail 10F was inserted to provide temporary drainage. He was then referred to surgeon for definitive management.

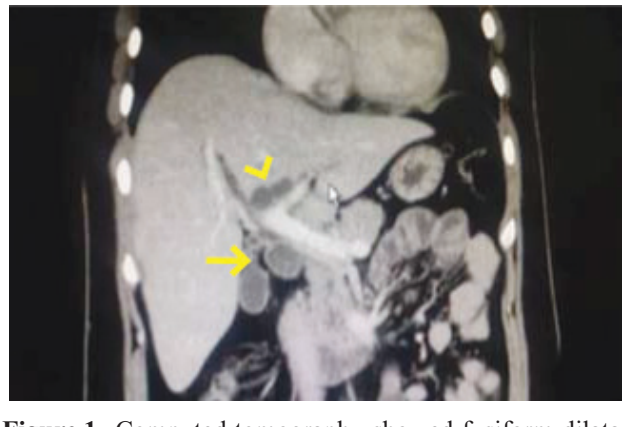

Figure 1. Computed tomography showed fusiform dilatation of the common bile duct (arrow), and left intrahepatic duct (arrow head).

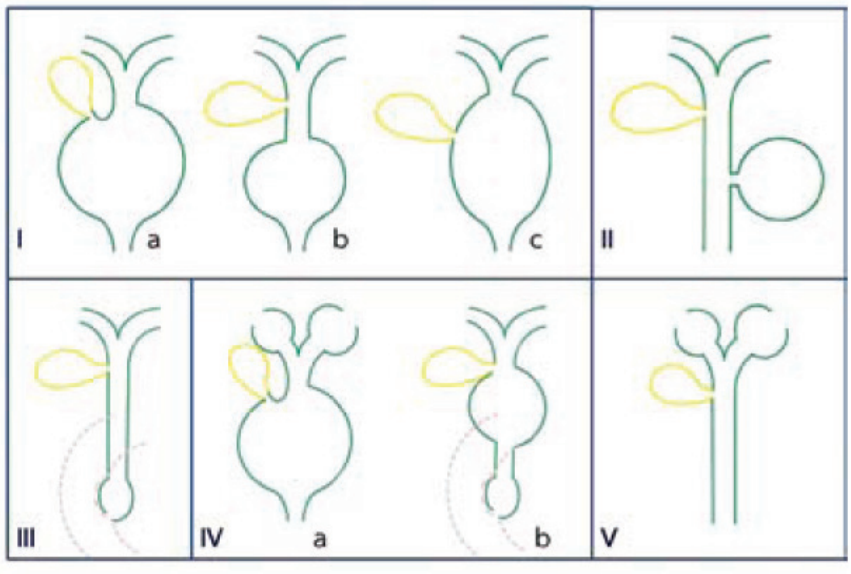

Figure 2. Type Ia, common type; Ib, segmental dilatation; Ic, diffuse or cylindrical dilatation; II, diverticulum; III, choledochocele; IVa, multiple cysts (intra- and extrahepatic); IVb, multiple cysts (extrahepatic); V, single or multiple dilatations of the intrahepatic ducts (Caroli's disease)[4].

Diagnosis:

Cholangitis, Choledochal cyst type 4A

Discussion:

Choledochal cysts are congenital anomalies of the biliary tract that are manifested by disproportionate dilatations of the biliary system[1]. The incidence of choledochal cysts shows significant geographic variation, being higher in the Asian population and reaching up to 1 in 1000[2]. Choledochal cysts are not familial; female children are affected more commonly than male children. Cases have been described in utero and in older adult patients, but approximately two thirds of patients seek medical attention before the age of 10[3]. The most common type based on Todani and colleagues' classification is type 1 accounting 80-90%[4]. This case presented with multiple left intrahepatic and common bile duct cysts which was classified as type 4A, the second most common with the incidence of 12-25% based on Gadelhak *et al* 20 years single center experience[5] (Figure 3). The cause of choledochal cysts has not been well established[3]. Cysts are composed of a fibrous wall; there may be no epithelial lining or a low columnar epithelium. The infantile form of choledochal cyst disease often presents first month of life and as many as 80% with jaundice and acholic stools[6]. In a 2008 series, adults were more likely to exhibit abdominal pain (97% vs. 63%, $p < 0.001$), and children were more likely to experience jaundice (71% vs. 25%, $p = 0.001$). The classic triad of abdominal pain, jaundice, and a palpable abdominal mass is observed in less than 20% of patients[7]. The diagnosis of a choledochal cyst can be established by ultrasonography[8]. In the older patients, percutaneous transhepatic

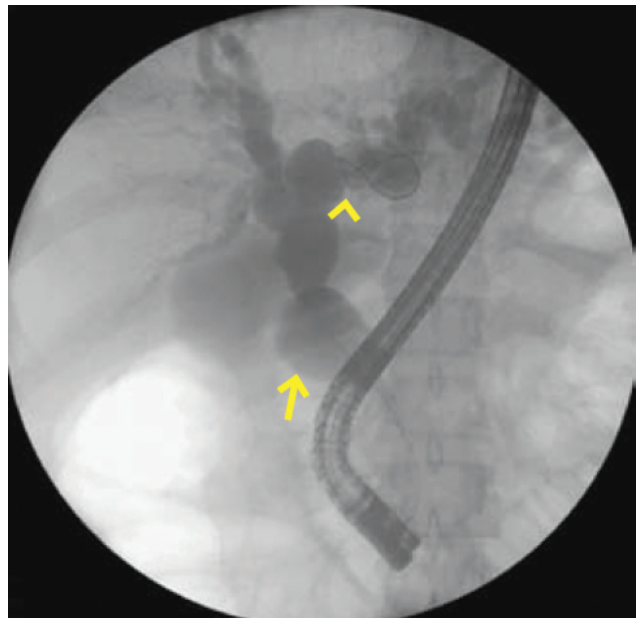

Figure 3. Cholangiogram revealed saccular dilatation of the common bile duct (arrow) and saccular dilatation of the left intrahepatic duct (arrow head), compatible with Todini type 4A.

cholangiography or ERCP may help define the anatomic features of the cyst. MRCP and CT scan is less effective than ERCP for detecting minor ductal abnormalities and small choledochoceles in adults[9]. Complete excision of the cyst with reconstruction of the extrahepatic biliary tree is the best treatment strategy to avoid long-term complications especially malignant transformation, recurrent cholangitis and gallstones[10,11].

REFERENCES

1. Lee HK, Park SJ, Yi BH, *et al*. Imaging features of adult choledochal cysts: a pictorial review. Korean J Radiol 2009;10:71-80.
2. Hung MH, Lin LH, Chen DF, *et al*. Choledochal cysts in infants and children: experiences over a 20-year period at a single institution. Eur J Pediatr 2011;170:1179-85.
3. Miyano T, Yamataka A. Choledochal cysts. Curr Opin Pediatr 1997;9:283-8.
4. Todani T, Watanabe Y, Narusue M, *et al*. Congenital bile duct cysts: Classification, operative procedures, and review of thirty-seven cases including cancer arising from choledochal cyst. Am J Surg 1977;134:263-9.
5. Nabil G, Ahmed S, Hosam H. Diagnosis and management of choledochal cyst: 20 years of single center experience, World J Gastroenterol 2014;20:7061-6.
6. Todani T, Urushihara N, Morotomi Y, *et al*. Characteristics of choledochal cysts in neonates and early infants. Eur J Pediatr Surg 1995;5:143-5.
7. Edil BH, Cameron JL, Reddy S, *et al*. Choledochal cyst disease in children and adults: A 30-year single-institution experience. J Am Coll Surg 2008;206:1000-5.
8. Paltiel HJ. Imaging of neonatal cholestasis. Semin Ultrasound CT MR 1994;15:290-305.
9. Shaffer E. Can MRCP replace ERCP in the diagnosis of congenital bile-duct cysts? Nat Clin Pract Gastroenterol Hepatol 2006;3:76-7.
10. O'Neill JA Jr. Choledochal cyst. Curr Probl Surg 1992;29:361-410.
11. Liu SL, Li L, Hou WY, *et al*. Laparoscopic excision of choledochal cyst and Roux-en-Y hepaticojejunostomy in symptomatic neonates. JPediatr Surg 2009;44: 508-11.

CASE 3

A 49-year-old Thai woman presented with recurrent cholangitis. The CT of upper abdomen revealed left intrahepatic duct (IHD) dilatation, multiple stones in IHD and common hepatic duct (CHD), and atrophy of the left lobe of liver (Figure 1). ERCP confirmed markedly dilation of left IHD with numerous round-shaped filling defects in left IHD. Multiple attempts of balloon extraction were done, however the complete stone clearance could not be achieved because of the associated stricture (Figure 2). Then, the surgical team was consulted for left hepatic resection.

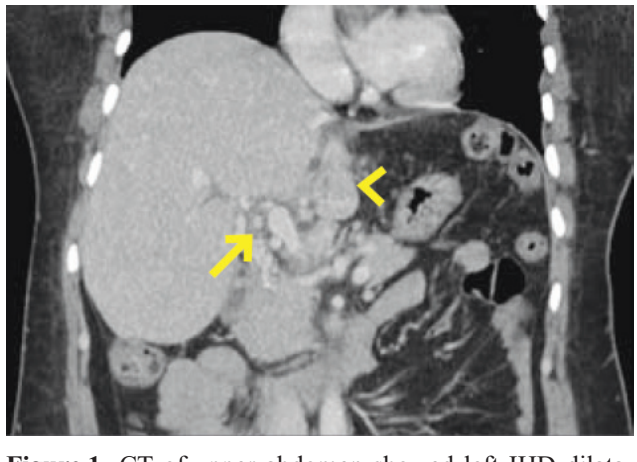

Figure 1. CT of upper abdomen showed left IHD dilatation with multiple IHD stones (arrow) and left lobe atrophy (arrow head).

Diagnosis:

Oriental cholangiohepatitis

Discussion:

Oriental cholangiohepatitis was also known as recurrent pyogenic cholangitis, intrahepatic pigmented stone, and Hong Kong disease[1]. The pathological changes were primarily in the bile ducts such as proliferation, inflammatory cells infiltration and fibrous changes at bile ducts wall[1]. Occasionally, the lateral segment of left lobe liver might be shrunken. Chronic infestations at bile ducts such as Clonorchis sinensis, Opisthorchis species, Fasciola hepatica and Ascaris lumbricoides were the most described in pathogenesis of oriental cholangiohepatitis[3]. Chronic infestations result in recurrent cholangitis, bile duct injury, bile stasis and leading to stone formation[2]. The management of oriental cholangiohepatitis is multidisciplinary approach including endoscopists, intervention radiolo-

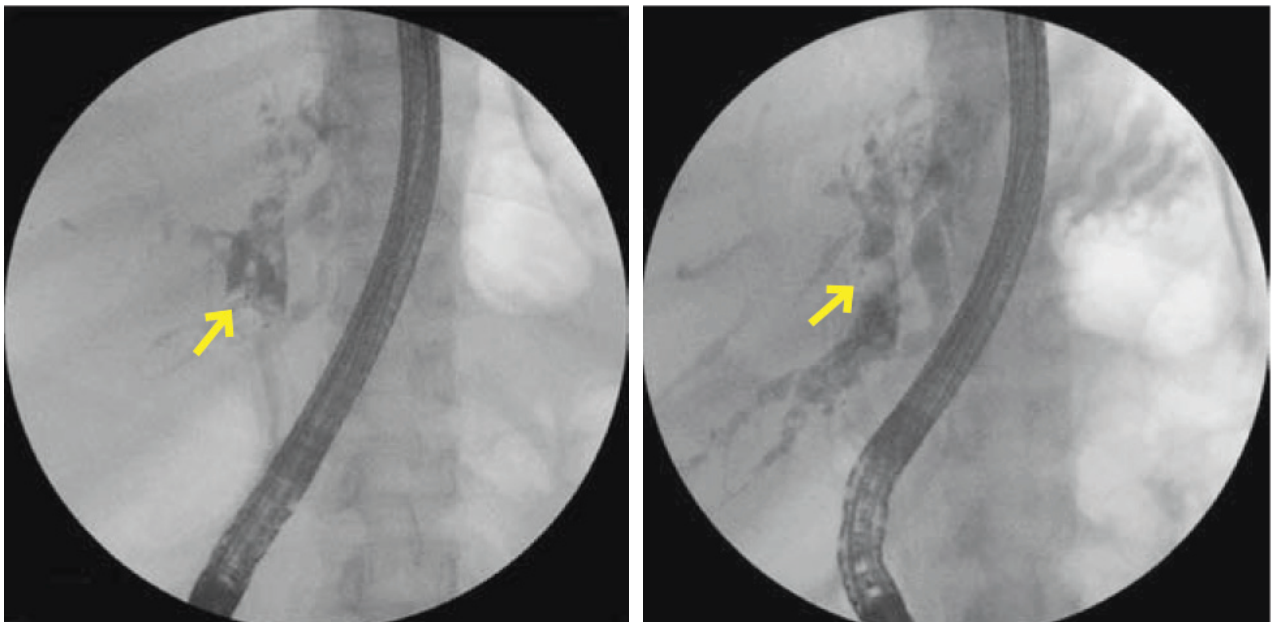

Figure 2. ERCP demonstrated markedly dilated left IHD with numerous round-shaped filling defects (arrow).

gist, and hepato-biliary surgeon[4]. Hepatic resection is preferred in patients with hepatic segment atrophy, focal disease, or failure of less invasive therapy[3].

REFERENCES

1. Al-Sukhni W, Gallinger S, Pratzer A, *et al*. Recurrent pyogenic cholangitis with hepatolithiasis—the role of surgical therapy in North America. J Gastrointest Surg 2008;12:496-503.
2. Lim JH. Oriental cholangiohepatitis: pathologic, clinical, and radiologic features. AJR Am J Roentgenol 1991;157:1-8.
3. Singla S, Warner AH, Jain A, *et al*. Oriental cholangiohepatitis masquerading as cholangiocarcinoma: A rare presentation that surgeons need to know. Int J Surg Case Rep 2012;3:235-7.
4. Tsui WM, Chan YK, Wong CT, *et al*. Hepatolithiasis and the syndrome of recurrent pyogenic cholangitis: clinical, radiologic, and pathologic features. Semin Liver Dis 2011;31:33-48.