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Two Consecutive Cases of Ampulla of Vater Cancer Combined with Annular Pancreas and Unusual Anatomic Variation

Annular pancreas is a rare congenital anomaly that consists of a ring of pancreatic tissue partially or completely encircling the descending portion of the duodenum. Coexisting ampullary carcinoma in annular pancreas combined with anomaly of hepatic artery or bile duct are thought to be extremely rare. Two consecutive cases of ampullary carcinoma in annular pancreas with bile duct or hepatic artery anomaly are described. In addition, English literature reports of coexisting ampullary carcinoma in annular pancreas are summarized. Clinical symptoms of the two patients were jaundice and abdominal discomfort. The two ampullary cancers were early adenocarcinomas in the ampulla of Vater that were curatively treated by pylorus preserving pancreaticoduodenectomy. Ampullary carcinoma associated with annular pancreas is rare. Its combination with an additional biliary or hepatic artery anomaly make our cases extremely unique. Certain aberrant events in the overall stages of the development of the liver, bile duct, and pancreas may have occurred in these patients. Surgeons need to note preoperatively these possible associated anatomic variations.

Key Words : Annular pancreas, Ampulla of vater cancer

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Introduction

Annular pancreas is a rare congenital anomaly that consists of a ring of pancreatic tissue partially or completely encircling the descending portion of the duodenum. Tiedemann described this pancreatic anomaly in 1818 and the term "annular pancreas" was named by Ecker in 1862.[1] Its incidence has been quoted as between 1～3 per 20,000 autopsies.[2] The Korean study group for pancreatobiliary disease recently reported only 2 cases (0.05%) out of 4097 endoscopic retrograde cholangiopancreatogram (ERCP).[3] Variety of variations in anatomy of the bile duct and vascular system have been recognized in the hepatobiliary system. Retrospective reviews of intraoperative cholangiograms have shown that up to 20% of patients have anatomical variations in the biliary tree.[4] About 25～75% of hepatic artery variations have been known to occur.[5] Very few studies have been published in the English literature about the coexistence of annular pancreas with ampullary malignancy. Therefore, those cases

combined with anomaly of hepatic artery or bile duct are thought to be extremely rare. Herein, we report two consecutive cases of ampullary adenocarcinoma with annular pancreas combined with the bile duct or hepatic artery anomaly.

Case Report

Case 1

A 61-year-old woman was admitted to our hospital because of dark urine and jaundice with fever. She had been in good health without any previous medical history. Laboratory investigations showed leukocytosis (13,310/ μl, neutrophil, 87.2%) and raised total bilirubin (4.4 mg/dl; reference range, 0.2～1.2), direct bilirubin (3.2 mg/dl; reference range, 0.1～0.4) serum AST (130 IU/L; reference range, 13～34 IU/L), serum ALT (116 IU/L; reference range, 5～46), alkaline phosphatase (1,304 IU/L; reference range, 38.0～115), and gamma-GT (537 IU/L; reference range, 7～35). Serum amylase and lipase were within normal limits. CA 19-9 and CEA were 295 U/ml (reference range, <37) and 1.15 ng/ml (reference range, <5), respectively. Percutaneous transhepatic biliary drainage

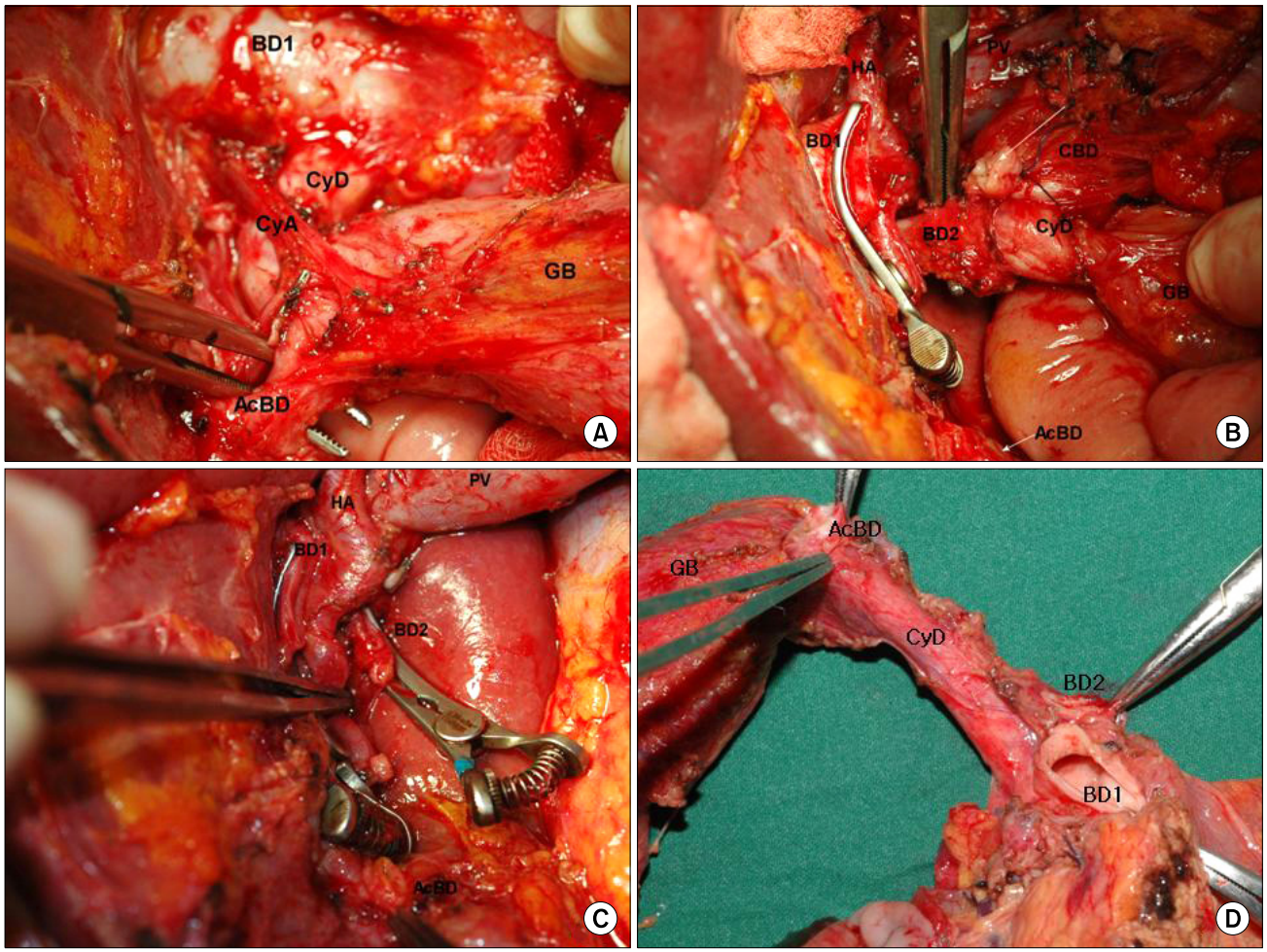

Fig. 1. Intraoperative view of bile duct variation. An AcBD is draining into GB. Unusually long cystic duct and artery are noted (A). A BD2 is identified after division of biliary structures (BD1) that were thought to be CBD. Note CyD draining into BD1, not CBD (B). Three bile ducts were identified after removal of specimen (C). Bile duct variation noted in resected specimen. (AcBD=accessory bile duct; BD1=bile duct 1; BD2=bile duct 2; GB=gallbladder; CyD=cystic duct; CBD=common bile duct; HA=hepatic artery; CyA=cystic artery; PV=portal vein).

(PTBD) was done for decompression of biliary obstruction since endoscopic retrograde cholangiography failed due to a large ampullary mass about 3 cm in size. Tissue biopsy was successfully conducted to report tubulovillous adenoma with focal area of well-differentiated adenocarcinoma transformation. Abdominal CT scan showed periampullary mass with marked dilatation of the bile duct system, suggesting periampullary malignant mass. There was no evidence of distant metastasis or contraindications to curative surgery. She underwent pylorus-preserving pancreaticoduodenectomy. Interestingly, the right lobe of the liver was unusually enlarged to the level of the right iliac crest in the operative field. Complete annular pancreas wrapping the second portion of the duodenum and combined anomaly of the biliary system were noted during operation (Fig. 1). Three separate choledochojejunostomies were performed to complete biliary drainage. The patient was discharged on the 15th operative day after uneventful recovery. Follow-up PTBD cholangiogram and MRCP confirmed unusual draining of the biliary system without evidence of surgical complications (Fig. 2). Gross surgical pathology showed presence of complete annular pancreas without pancreatitis. An ill-defined polypoid mass is noted in the ampulla of Vater, measuring 3.5×3×3.5 cm (Fig. 3). The tumor was revealed to be well-differentiated adenocarcinoma arising from ampulla of Vater with focal extension to the duodenal wall (T2N0M0, stage IB). She will be followed up without adjuvant treatment.

Case 2

A 64-year-old male was admitted for abdominal discomfort. He had an episode of pancreatitis and cerebral infarction 10 years ago. About 7 years ago, he underwent exploratory laparotomy due to biliary complication after laparoscopic cholecystectomy in other hospital. Initial laboratory investigations revealed elevated total bilirubin (2.3 mg/dl; reference range, 0.2～1.2), direct bilirubin (1.2 mg/dl; reference range, 0.1～0.4) serum AST (32 IU/L; reference range, 13～34), serum ALT (150 IU/L; reference range, 5～46), alkaline phosphatase (632 IU/L; reference range, 38.0～115), and gamma-GT (1,772 IU/L; reference range, 7～35). Serum amylase and lipase were also raised (300 U/L, 905 U/L, respectively). CA 19-9 was checked as 132 U/ml (reference

Fig. 2. MRCP and percutaneous transhepatic cholangiogram of case 1. Follow-up biliary images confirmed intrahepatic biliary anatomy. There are separate right posterior hepatic bile duct (arrow) (A), right anterior hepatic bile duct (long arrow), and left hepatic bile duct (short arrow) (B).

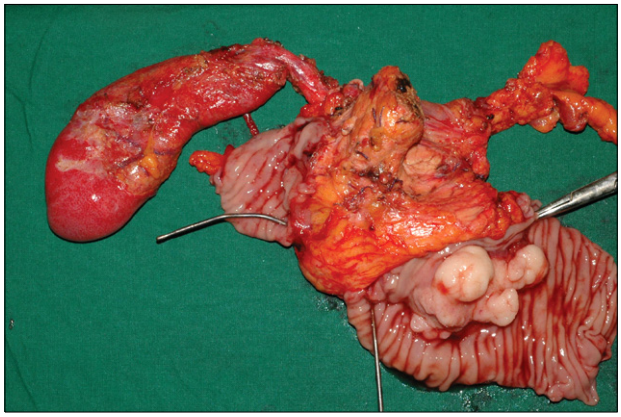

Fig. 3. Surgical pathology of case 1. Resected specimen shows definitive annular pancreas and polypoid ampullary mass.

range, <37) and CEA were within normal limits. Endoscopic retrograde cholangiography was done and tissue biopsy of ampullary mass w as successfully performed to report adenocarcinoma of well differentiation. Abdominal MRI scan showed periamullary mass with well visualization of annular pancreas. Aberrant pancreatic duct is extending to the lateral side of the duodenum with small size of pancreatic parenchyma accompanying it suggesting annular pancreas (Fig. 4A). He underwent pylorus-preserving pancreaticoduodenectomy under the impression of ampullary cancer. Total replaced hepatic artery arising from the origin of the superior

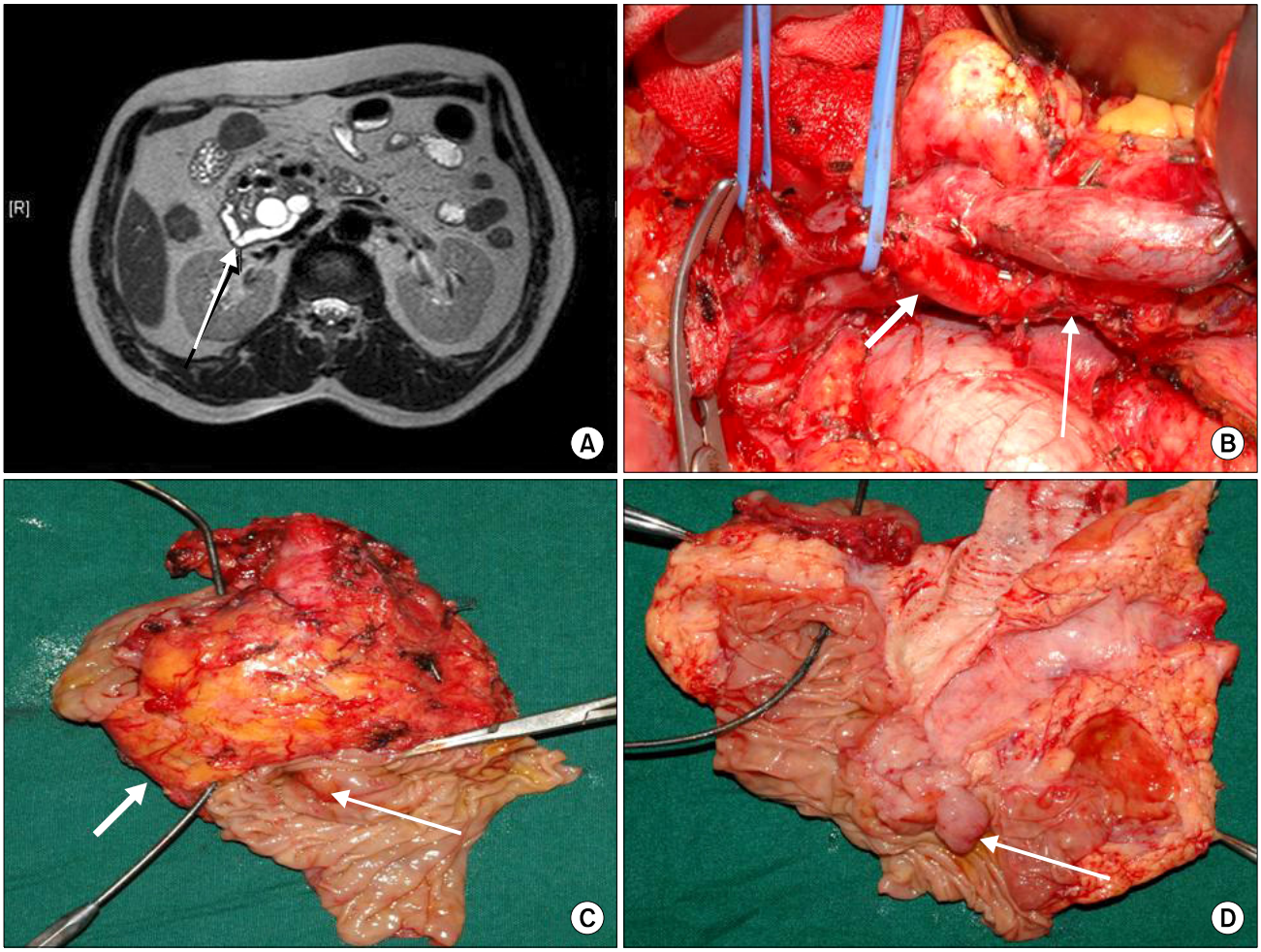

Fig. 4. Image finding, operative finding, and surgical pathology of case 2. MRCP typically indicates presence of annular pancreas. Aberrant pancreatic duct is extending to the lateral side of the duodenum with small size of pancreatic parenchyma accompanying it suggesting annular pancreas (A). Total replaced right hepatic artery (wide short arrow) is arised from superior mesenteric artery (narrow long arrow) (B). Gross findings of resected specimen are shown (C, D).

Table 1. Characteristics of ampulla of vater cancer associated with annular pancreas

No.	Age	Gender	Symptoms	Tumor	Operation	Tumor pathology
1[10]	80	Female	Jaundice, weight loss, steatorrhea	"Large"	PD	Adenocarcinoma, moderate diff (TxN0M0)
2[16]	66	Male	Jaundice, anorexia, weight loss	"Small"	PD	Adenocarcinoma, moderate diff (T2N0M0)
3[17]	55	Male	Jaundice, pruritus, clay-colored stool, high-colored urine	2×2 cm	PD	Adenocarcinoma (T1NXM0),
4[18]	40	Female	Nausea, vomiting, abdominal pain and distention, jaundice	"Fragile mucosa"	PD	Malignancy (TXN0M0)
5[18]	45	Male	Epigastric pain, jaundice	"Small"	PPPD	Adenocarcinoma (TXNXM0)
6[9]	78	Female	Epigastric pain, back pain, jaundice	1.5×3.5 cm	PD	Adenocarcinoma well-diff (T1N0M0)
Case 1	61	Female	Dark urine, jaundice	3.5×3 cm	PPPD	Adenocarcinoma well-diff (T2N0M0)
Case 2	64	Male	Chill, fever, abdominal discomfort	2.5×2 cm	PPPD	Adenocarcinoma well-diff (T1N0M0)

PD=pancreaticoduodenectomy, PPPD=pylorus preserving pancreaticoduodenectomy

mesenteric artery was noted and carefully preserved during operation (Fig. 4B). Gross surgical pathology showed presence of complete annular pancreas without overt pancreatitis. An irregular-shaped polypoid mass is noted in the ampulla of Vater, measuring 2.5×2 cm (Fig. 4C, D). The tumor was revealed to be well-differentiated adenocarcinoma arising from ampulla of Vater limited to only ampulla (T1N0M0, stage I). He will be followed up without adjuvant treatment.

Discussion

Annular pancreas is a very rare but well-known congenital anomaly. It is generally accepted that the ring formation originates from a ventral pancreas but the exact development of annular pancreas is not clear. Nobukawa et al.[6] recently suggested that an annular pancreas originates from paired ventral pancreata with a ring formation originating from the left lobe of paired ventral pancreata. Clinical presentation can be different according to the age. In the infant period, it includes symptoms of gastric outlet obstruction and associated anomalies, such as Down syndrome, intestinal malrotations, duodenal stenosis, duodenal diverticulum, tracheoesophageal fistula, imperforated anus, and cardiac defects. However, half of patients with annular pancreas do not have symptoms until adulthood.[7] The case 1 patient had jaundice with mild fever and case 2 patient had abdominal discomfort with a previous episode of pancreatitis. In general, clinical manifestation of annular pancreas is rare in adulthood but it can include duodenal obstruction, peptic ulcer, pancreatitis, and jaundice. In particular, there are several causes for jaundice in patients with annular pancreas,[8] but the possible coexistence of periamullary malignancy should not be overlooked. Foo et al.[9] recently reviewed 14 cases of pancreaticobiliary malignancy cases associated with annular pancreas (1 case of insulinoma, 2 chollangiocarcinomas, 5 pancreatic cancers, and 6 ampullary adenocarcinomas). It is believed that the first case of annular pancreas with coexistence of ampullary carcinoma was reported in 1982.[10] Until now, ampullary carcinoma is most commonly associated with periampullary malignancy in annular pancreas. Here, we summarized the characteristics of reported cases of ampullary carcinoma associated with annular pancreas (Table 1). Although this case study is too small to generalize the characteristics of ampullary carcinoma

associated with annular pancreas, it seems to have no gender predilection (4 males, 4 females) with a median age of 61 years (range, 40～80 years). Ampullary carcinomas usually is polypoid mass with relatively early cancerous lesions, which means curative surgery may play a great role in managing this malignancy. We could not determine whether our case 1 patient had annular pancreas preoperatively. The volume of annular pancreas was so thin that it was very difficult to identify the presence of this pancreatic anomaly preoperatively; however, a retrospective review of abdominal CT scan could delineate the annular pancreas in the second portion of the duodenum. In contrast, we noted preoperatively that the case 2 patient had annular pancreas due to a dilated pancreatic duct resulting from obstruction of ampullary mass (Fig. 4A). Currently, annular pancreas is recognized with increasing frequency in clinical settings since the advent of abdominal CT,[11] ERCP,[12] and MRCP.[13] Therefore, frequent and long- term follow-up measures must be taken in patients with annular pancreas. In addition, the patients had combined bile duct or hepatic artery variation. This anatomical variability of the bile duct system results from aberrations of embryological development.[4] In the case 1 patient, intraoperative finding, follow-up MRCP, and PTBD cholangiogram confirmed that the patient had separate right posterior hepatic bile duct (AcBD) draining into the gallbladder and right anterior hepatic bile duct (BD1) joining the cystic duct before the left hepatic bile duct (BD2) communicates right anterior hepatic bile duct. This variation is believed to be a very unusual biliary anomaly.[14] In the case 2 patient, total replaced right hepatic artery arsing from SMA was also noted. One anatomic study of hepatic arteries in donor liver shows 2 out of 1,000 (0.2%) donor livers have that type of hepatic artery variation.[15] In other reports, there were no comments about associated anatomic variations in coexisting ampullary adenocarcinoma with annular pancreas. Considering the interesting operative findings of the enlarged hepatic right lobe, combined bile duct variation, replaced hepatic artery, and annular pancreas, these patients might have experienced certain aberrant events in the overall stages of the development of the liver, bile duct, and pancreas. Hence, surgeons must note preoperatively these possible associated anatomic variations in order to avoid unnecessary damage during operation.

Ampullary carcinoma associated with annular pancreas is rare. Its combination with additional biliary or hepatic artery anomaly made our cases extremely unique. Complete curative surgeries could be performed in all reported cases of coexisting ampullary carcinoma with annular pancreas. The identification of annular pancreas is increasing, and careful follow up and proper investigation is recommended in patients with jaundice and abdominal pain.

References

1. Yogi Y, Shibue T, Hashimoto S. *Annular pancreas detected in adults, diagnosed by endoscopic retrograde cholangiopancreatography: report of four cases. Gastroenterol Jpn 1987;22:92-99.*
2. Ravitch MM, Woods AC Jr. *Annular pancreas. Ann Surg 1950;132:1116-1127.*
3. Kim MH, Lim BC, Park HJ, et al. *A study on normal structures, variations, and anomalies of the Korean pancreaticobiliary ducts: cooperative multicenter study. Korean J Gastrointest Endosc 2000;21:624-632.*
4. Hayes MA, Goldenberg IS, Bishop CC. *The developmental basis for bile duct anomalies. Surg Gynecol Obstet 1958; 107:447-456.*
5. Nelson TM, Pollak R, Jonasson O, Abcarian H. *Anatomic variants of the celiac, superior mesenteric, and inferior mesenteric arteries and their clinical relevance. Clin Anat 1988; 1:75-91.*
6. Nobukawa B, Otaka M, Suda K, Fujii H, Matsumoto Y, Miyano T. *An annular pancreas derived from paired ventral pancreata, supporting Baldwin's hypothesis. Pancreas 2000; 20:408-410.*
7. Lloyd-Jones W, Mountain JC, Warren KW. *Annular pancreas in the adult. Ann Surg 1972;176:163-170.*
8. Morrell MT, Keynes WM. *Annular pancreas and jaundice. Br J Surg 1970;57:814-816.*
9. Foo FJ, Gill U, Verbeke CS, Guthrie JA, Menon KV. *Ampullary carcinoma associated with an annular pancreas. JOP 2007;8:50-54.*
10. Transveldt E, Keith RG, Fonger J, Fisher MM. *Annular pancreas with coexistent ampullary carcinoma in an elderly woman. Can J Surg 1982;25:687-688.*
11. Inamoto K, Ishikawa Y, Itoh N. *CT demonstration of annular*

pancreas: case report. Gastrointest Radiol 1983;8:143-144.

12. Itoh Y, Hada T, Terano A, Itai Y, Harada T. *Pancreatitis in the annulus of annular pancreas demonstrated by the combined use of computed tomography and endoscopic retrograde cholangiopancreatography. Am J Gastroenterol 1989; 84:961-964.*
13. Yamaguchi Y, Sugiyama M, Sato Y, et al. *Annular pancreas complicated by carcinoma of the bile duct: diagnosis by MR cholangiopancreatography and endoscopic ultrasonography. Abdom Imaging 2003;28:381-383.*
14. Lamah M, Karanjia ND, Dickson GH. *Anatomical variations of the extrahepatic biliary tree: review of the world literature. Clin Anat 2001;14:167-172.*
15. Hiatt JR, Gabbay J, Busuttil RW. *Surgical anatomy of the hepatic arteries in 1,000 cases. Ann Surg 1994;220:50-52.*
16. Benger JR, Thompson MH. *Annular pancreas and obstructive jaundice. Am J Gastroenterol 1997;92:713-714.*
17. Rathnaraj S, Singh SK, Verghese M. *Annular pancreas associated with carcinoma of papilla. Indian J Gastroenterol 1998;17:110.*
18. Shan YS, Sy ED, Lin PW. *Annular pancreas with obstructive jaundice: beware of underlying neoplasm. Pancreas 2002;25: 314-316.*