Annular pancreas in a patient with malignant obstructive jaundice: A case report

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Abstract

Majority of cases with annular pancreas are asymptomatic. Common presentations in symptomatic patients are abdominal pain, nausea and vomiting. Jaundice is very rare presentation of annular pancreas. We are presenting a case of 40 years male with annular pancrea. He presented with jaundice, anorexia and weight loss. Contrast enhanced computed tomography shows annular pancreas with extrahepatic as well as intrahepatic bile duct dilatation. Magnetic resonant cholangiopancreatography showed abrupt narrowing of distal common bile duct with upstream dilation of biliary duct suggestive of distal cholangiocarcinoma. This case illustrates the important of evaluation for malignant cause of obstructive jaundice in patients with annular pancreas presented with jaundice, anorexia and weight loss

Key words: Annular pancreas, Distal cholangiocarcinoma, Jaundice

Introduction

Annular pancreas (AP) is a rare congenital anomaly. Majority of AP are asymptomatic and found incidentally. Usual symptoms in adults are abdominal pain in 70%, nausea and vomiting in 60%, and hematemesis in 10% of patients¹. Most symptoms are secondary to duodenal obstruction, pancreatitis (acute or chronic or recurrent) or peptic ulcer disease. It may rarely present with obstructive jaundice and even rarely malignancy². We are presenting one case of annular pancreas with malignant obstructive jaundice.

Case Report

Fortyyears gentleman presented in gastrointestinal surgery clinic with complain of yellowish discoloration of eyes, dark color urine and itching of whole body for 2 months. He also had anorexia and lost weight (15kg) during this illness. He did not complain pain abdomen, nausea, vomiting, black tarry stool, fever or myalgia. Onphysical examination, he was jaundiced and hepatomegaly was present. Rest of the physical examination was normal. His haemoglobin, total bilirubin, conjugated bilirubin, alanine aminotransferase,

aspartate aminotransferase, and alkaline phosphatase level were 10.8 gm%, 263 Umol/l, 203 Umol/l, 48 U/L, 37 U/L, and440 U/L respectively. Tumor markers(CEA and CA 19-9) were normal. Others haematological and biochemical investigations were with in normal limit. Upper gastroduodenoscopy did not show growth at ampullary region. Ultrasonography showed mild hepatomegaly, distended GB with dilated intrahepatic and extrahepatic bile duct (common bile duct 14.5mm). Magnetic resonance cholangio pancreatography (MRCP) showed marked dilatation of the intrahepatic and extrahepatic bile duct with abrupt narrowing and beaking of the distal common bile duct however no definite mass was identified (Figure 1). Contrast enhanced computed tomography (CECT) of abdomen showed pancreatic tissue completely encircling the second part of duodenum without proximal duodenal dilatation, abrupt narrowing of distal common bile duct with upstream biliary dilatation (common bile duct 15.7mm). There was presence of duct of Santorini separately opened into minor papilla in CECT, suggestive of pancreatic divisum (Figure 2). Thus, with the suspicious of distal cholangiocarcinoma, classic pancreaticoduodenectomy was performed. Intraoperative findings were presence

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of annular pancreas completely encircling second part of duodenum, 1cm x 1 cm mass at distal common bile duct. There was grossly normal pancreatic parenchyma, dilated common bile duct and distended gallbladder (Figure 3). Histopathological examination showed pancreatic tissue all around the second part of duodenum, small whitish nodule measuring 0.5 cm x 0.5 cm in distal common bile duct. Microscopic examination showed that the tumor is seen arising from the lining epithelium of distal common bile duct and infiltrating into the subepithelial tissue close to but not infiltrating the pancreas features consistent with moderately differentiated adenocarcinoma (Figure 4). There was presence of single metastatic lymph node. Thus, AJCC 7th edition TNM stage was distal cholangiocarcinoa, T1N1Mo. Intraoperative and postoperative period was uneventful other than superficial surgical site infection, which was the cause of prolonged hospital stay. Patient was discharged on 15th postoperative day.

Figures



Figure 1. MRCP showing abrupt tapering of distal CBD with proximal biliary dilatation.



Figure 2. CECT abdomen showing annular pancrease

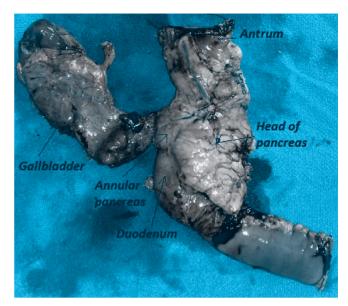


Figure 3. Resected specimen after Whipple Operation.



Figure 4. Histopathology showing pancreatic tissue encircling duodenum.

Discussion

Annular pancreas is a rare congenital abnormality (1/20,000), which results from the failure of ventral pancreas to rotate with the duodenum during embryogenesis³. Leeco's and Baldwin's theories of pathogenesis of APare the most acceptable. Lecco postulated that adhesion of the distal tip of the ventral primordium to the duodenal wall before its migration results in annular pancreas. Baldwin stated that persistence and further development of left component is responsible for the formation of pancreatic ring around the duodenum⁴. Annular pancreas affects both sexes equally; however, a recent review has found that symptomatic adult AP is mainly concentrated in males⁵. Annular pancreas has a bimodal pattern of presentation, the first peak is in infancy and a later peak occurs in the

fourth decade of life. However, both have very different clinical pictures. Paediatric cases usually present with duodenal obstruction and may be associated with other congenital anomalies⁶. Adult annular pancreas is associated with duodenal obstruction (60%), pancreatitis (15-50%) and peptic ulceration (26-48%)⁷. Most adults become symptomatic between the ages of 20 and 50⁸. Pancreatitis due to annular pancreas is generally confined to the annulus and to the adjoining pancreatic head, preserving the body and tail of the gland. Its pathogenesis is most likely related to the inability of pancreatic secretions to flow through the annular duct (Santorini duct)⁹.

Obstructive jaundice is an uncommon feature and not usually a direct result of the AP. Morrell and Keynes reported on 15 AP patients with jaundice and causes included pancreatitis (10 cases), choledocholithiasis (4 cases) and alcoholic liver disease (1 case)¹⁰. A review by Yogi et al. of 105 annular pancreas patients revealed nine with obstructive jaundice due to ampullary carcinoma (5 cases), pancreatic carcinoma (2 cases) and cholangiocarcinoma (2 cases)¹¹. A subsequent collection of 151 cases of annular pancreas by Ogawa et al. revealed 15 had pancreaticobiliary malignancy, including 5 cholangiocarcinomas, 4 gallbladder carcinomas, 4 duodenal and 2 pancreatic carcinomas

Annular pancreas can be diagnosed on the basis of CECT and magnetic resonance imaging(MRI) findings that reveal pancreatic tissue and an annular duct encircling the descending duodenum ¹³.Endoscopic retrograde cholangiopancreatography (ERCP) has been considered the gold standard method in the diagnostic workup, but it is an invasive method and associated with morbidity, including acute pancreatitis.MRCP has overtaken ERCP as it is non-invasive and is in demonstrating anomalous pancreatic duct and where ERCP fails especially in duodenal obstruction¹⁴. Study by Kumaresan S et al shows different type of annular pancreas according to configuration of annular pancreas based on CECT/MRI findings, that is 1) Pancreas extending in anterolateral direction (10- to 12-o'clock position) to second part of duodenum, 2) Pancreas extending in posterolateral direction (6- to 8-o'clock position) to second part of duodenum, 3) Pancreas extending around second part of duodenum but not completely and 4) Complete annulus. More than one third (37.5%) of patients had a radiologically incomplete annulus on images¹⁵.

Once Symptoms develops, AP needs surgical treatment. Dissection of the pancreatic ring should be avoided due to a high incidence of complications, including duodenal leak, pancreatic fistula, and postoperative pancreatitis. Surgical bypass is required. Duodenoduodenostomy is preferred because it is most physiological and prevent to form blind loop. Other options are duodenojejunostomy and gastrojejunostomy 16. Periampullary malignancy should be suspected in adult patient with annular pancrease presenting with obstructive jaundice. Pancreaticoduodenectomy needs to be considered in case of annular pancrease associated with proven or suspected periampullary malignancy 17 Another indication for pancreaticoduodenectomy ispancreatolithiasis and localized chronic pancreatitis 18.

Conclusion

Presence of jaundice and/or weight loss in patient with annular pancreas need to be evaluated to exclude associated malignancy which might be the cause of obstructive jaundice.

Conflict of interest: None declared.

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