Diagnosis of Annular Pancreas with Endoscopic Retrograde Cholangiopancreatography

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Two patients with annular pancreas are described. The diagnosis was established unequivocally with endoscopic retrograde cholangiopancreatography before operation. In both patients there were pancreatitis of the annular pancreas. The first patient also had congenital absence of the ventral pancreas and pancreatic insufficiency. The second patient subsequently developed gastric outlet obstruction. The literature is reviewed.

The annular pancreas is a congenital anomaly which may cause duodenal obstruction in the neonatal period or remain silent throughout life. In an adult, symptoms from annular pancreas usually present between the ages of 20-50 yr as duodenal obstruction. Peptic ulcer disease and pancreatitis may be found in association with annular pancreas. Biliary tract obstruction has also been reported. Although an upper GI series is considered the diagnostic test, recognition is not always easy preoperatively. In this report we describe 2 patients in whom the diagnosis of annular pancreas was clearly established preoperatively by endoscopic retrograde cholangiopancreatography (ERCP).

Case Reports

Case 1. G.O. is a 38-yr-old white woman with diabetes for 10 yr, requiring insulin. An elevated alkaline phosphatase (500 units, normal <85 units) and 5'-nucleotidase were noted 8 yr before; however, a liver biopsy demonstrated only pericholangitis. Calcifications were noted in the head of the pancreas, and fecal fat excretion was 15.6 g/24 hr (normal <6 g/24 hr). An upper gastrointestinal and small bowel series was normal. Chronic pancreatitis of unknown etiology with secondary pericholangitis was the diagnosis. In 1975, weight loss, diarrhea, greasy stools, and a persistently elevated alkaline phosphatase led to a repeat upper gastrointestinal series which showed a constriction of the second portion of her duodenum which appeared to be extrinsic and was compatible with an annular pancreas (Figure 1). An ERCP clearly showed the pancreatic duct encircling the proximal descending duodenum, suggesting an annular pancreas (Figure 2). The main pancreatic duct was not visualized. The common bile duct was normal. Exploratory laparotomy revealed an annular pancreas with pancreatitis and absence of the remainder of the pancreas. The sphincter of Oddi was felt to be mildly stenotic, and a 2-mm gallstone was found. A cholecystectomy and sphincteroplasty were performed. She has done well since surgery on Viokase and insulin, although her alkaline phosphatase has remained elevated.

Case 2. F. P. is a 48-yr-old white male with a history of diabetes for 6 yr. He had a 30-yr history of heavy drinking. In 1976, he had acute pancreatitis which rapidly improved, but his serum amylase remained elevated at the time of discharge. Although he stopped drinking, epigastric pain and vomiting recurred 1 mo later. Serum amylase was initially 2,000 units and remained persistently elevated at 500 units despite clinical improvement. An upper gastrointestinal series showed a constricted area in the second portion of the duodenum (Figure 3). An ultrasound examination and a computerized tomographic evaluation showed an enlarged head of the pancreas (Figure 3). An ultrasound examination and a computerized tomographic evaluation showed an enlarged head of the pancreas (Figure 4). An ERCP demonstrated a constricted area in the second portion of the duodenum just below the duodenal bulb and a dilated pancreatic duct (1 cm in diameter) encircling the duodenum at the same level as the constriction on the upper gastrointestinal series (Figures 5 and 6). The main pancreatic duct was displaced medially (Figure 6). These findings were interpreted as pancreatitis in an annular pancreas. At exploratory laparotomy, the pancreatic head

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Figure 1. Localized narrowing of the postbulbar portion of the duodenum is evident on the upper gastrointestinal series.

was markedly inflamed and surrounded the second portion of the duodenum. A gap was noted anteriorly. The rest of the pancreas appeared normal.

He had three further admissions in 1976 because of abdominal pain, vomiting, and persistently elevated amylase in the range of 500 units. On one admission, duodenal obstruction dominated the clinical picture. A succussion splash was noted, and 1800 ml of fluid aspirated with nasogastric suction. In 1977 he had no episodes of abdominal pain or vomiting although his serum amylase and lipase were intermittently elevated. In February 1978, the patient was readmitted because of gastric outlet obstruction. He was operated upon and found to have a constricted band of fibrotic tissue surrounding the second portion of the duodenum; a gastrojejunostomy was performed.

Discussion

An annular pancreas consists of a band or ring of pancreatic tissue which surrounds the second portion of the duodenum. Two arms of pancreatic tissue extend from the head of the pancreas, one anteriorly and another posteriorly. Fusion of the two arms usually occurs, but a gap may be present between them, which usually occurs anteriorly. The duct of the annular pancreas usually originates on the left anterior surface of the duodenum, passes posteriorly around it, and opens into the main pancreatic duct or common bile duct near the ampulla.6,10,22 This anomaly is the result of the failure of the ventral anlage of the pancreas to rotate with the duodenum to the right and dorsally to join the dorsal anlage in embryonal life.

Various degrees of duodenal obstruction may result from an annular pancreas, ranging from (a) nearly complete, requiring immediate surgical intervention in the neonate; (b) partial, usually becoming symptomatic in the third to fifth decade; to (c) asymptomatic.1-5 Males are affected more than females. Abdominal pain and vomiting are the major symptoms.4,15

Peptic ulcer disease and pancreatitis are commonly found in association with symptomatic adult annular pancreas. The incidence of peptic ulcer is reported to range from 26% to 48%6,8,11,15-17 and the incidence of pancreatitis ranges from 15 to 50%6,9,11,16.6,11,18 Extrahepatic biliary obstruction has also been reported.6,11,18,19

Our first patient presented with asymptomatic chronic pancreatitis in association with an annular pancreas. She also had a mild degree of biliary tract obstruction and pericholangitis. ERCP clearly demonstrated the annular pancreas before surgery. At surgery she was found to have only the annular pancreas with congenital absence of the remainder of the pancreas. To our knowledge, congenital absence of the main portion of the pancreas with annular pancreas has not been previously described. Her chronic pancreatitis may have been secondary to gallstones or stenosis of the sphincter of Oddi. Having only an annular pancreas clearly made her more susceptible to pancreatic insufficiency and diabetes, once pancreatitis had developed. Our second patient presented with acute and chronic relapsing pancreatitis. ERCP demonstrated pancreatitis limited to the annular pancreas. Alcohol is the most likely cause of the pancreatitis, but the fact that the pan-
Figure 2. ERCP demonstrates a localized narrowing of the gas column of the descending duodenum (small arrows) which is encircled by the pancreatic duct (large arrow) thus conclusively demonstrating the presence of an annular pancreas. This distention of the proximal duodenum is related in part to the hypotonia and gaseous distention which occurs during ERCP. The common bile duct and gall bladder are normal. The stippled calcifications in the pancreas can be seen to the right of the second lumbar vertebra.

Figure 3. An eccentric narrowing of the immediate postbulbar duodenum is evident on the upper gastrointestinal series. The predominant impression occurs in the lateral aspect of the duodenum.
Figure 4. Localized enlargement of the pancreatic head is demonstrated (arrows) which could represent pancreatitis or carcinoma. Despite the administration of contrast material the relationship of the duodenum to the mass was never clearly delineated.

Figure 5. A dilated pancreatic duct (P) is seen encircling the narrowed air column of the postbulbar duodenum confirming the presence of an annular pancreas.
creatitis was limited to the annular pancreas suggests that it is more susceptible to inflammation than the remainder of the pancreas.

Lehman in 1949 first described the radiologic finding on the upper GI series in a patient with this anomaly which provided a presumptive diagnosis preoperatively. Subsequently, criteria for making the diagnosis on upper gastrointestinal series were established. These consist of: (a) an eccentric smooth annular defect of the outer margin of the descending duodenum, with a constant caliber without destruction of the normal mucosal pattern and (b) evidence of duodenal obstruction, e.g., symmetric dilation of the proximal duodenum and reverse peristalsis. Despite these criteria the diagnosis is still difficult to make, especially in cases which are associated with peptic ulcers. In a 1957 review of the literature, only 28% of 50 cases were diagnosed with an upper gastrointestinal series. Of 10 cases reviewed by MacGregor et al. since 1957, only 5 were correctly diagnosed preoperatively. In the 2 cases reported here, the duodenal deformity seen in Figures 1 and 3, although suggestive of annular pancreas, could also represent chronic pancreatitis or carcinoma of the head of the pancreas with surrounding pancreatitis.

Our 2 patients demonstrate that the diagnosis of annular pancreas can be easily established preoperatively with ERCP. Thus, ERCP should become the procedure of choice when annular pancreas is suspected in adults. ERCP would fail to make the diagnosis only if there was obstruction of the duct leading to the annular pancreas, if the duct of the annular pancreas did not empty into the main pancreatic duct or if there was high grade duodenal obstruction, making passage of the duodenoscope difficult. However, in 17 of 20 cases that Anderson and Wapshaw reviewed, the duct of the annular pancreas opened into the main pancreatic duct close to the ampulla. Thus, one will occasionally be unable to make the diagnosis with ERCP, but we believe the accuracy with ERCP should be much greater than that obtained with all previous diagnostic procedures.
References