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Annular pancreas in adults: imaging features in seven patients

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Abstract

Background: Annular pancreas is a rare congenital abnormality that may be associated with variable degrees of duodenal obstruction. This diagnosis is often overlooked in adult patients who present with symptoms suggestive of duodenal obstruction. Imaging evaluation aids in establishing the diagnosis. We evaluated the imaging findings in seven adult patients with known annular pancreas.

Methods: Seven adult patients with abdominal symptoms had evaluation with one or more of the following imaging studies: upper gastrointestinal (UGI) series, computed tomography (CT), and endoscopic retrograde cholangiopancreatography (ERCP). All patients underwent subsequent laparotomy as a part of routine care. Surgical and imaging findings were correlated in each patient.

Results: UGI series is suitable for demonstrating different degrees of duodenal narrowing at the level of pancreatic annulus. Contrast-enhanced abdominal CT is useful in visualizing directly the complete or partial annular pancreatic tissue. ERCP is particularly useful in visualizing the annulus duct coursing around the duodenum.

Conclusions: Imaging plays a pivotal role in the diagnosis of annular pancreas in adult patients avoiding surgery for confirmation with its associated cost and risks.

Key words: Annular — Pancreas — Duodenum — Obstruction.

An annular pancreas is a rare congenital anomaly that normally presents in childhood as the second most common pancreatic aberration, after pancreas divisum [1]. Although the exact etiology is unknown, several theories have been proposed. Tieken proposed that the annulus represents hypertrophy of the pancreatic head tissue [2]. Lecco suggested that adherence of the tip of the right ventral pancreatic tissue to the duodenal wall results in a ring of tissue around the duodenum at the time of dorsal rotation [2]. Baldwin postulated that the annulus results from the abnormal migration and rotation of the ventral anlagen of the pancreatic head, leaving a ring of normal pancreatic tissue around the duodenum [2]. The more accepted theories are those by Lecco [1] or Baldwin [2]. It involves the descending duodenum in 85% of cases and the first and third portions in the remaining 15% [1, 3]. The ring may be complete, or in some individuals only a smaller portion of pancreas (partial ring) will incompletely encircle the duodenum. Histologically, approximately 2–5 cm of normal pancreatic tissue is intermixed with the smooth muscle of the duodenal muscularis extending close to the mucosa [1].

Several congenital anomalies are associated with annular pancreas in up to 75% of patients. These congenital anomalies include trisomy-21 (Down syndrome), tracheoesophageal fistula, esophageal atresia, imperforate anus, and Hirschsprung disease [1]. Duodenal obstruction may also occur in association with annular pancreas. However, the significance of annular pancreas as the cause of duodenal obstruction is unclear because there are often associated abnormalities such as duodenal atresia, web, or stenosis. The degree of obstruction prescribes the age when the clinical manifestations are observed. The greater the obstruction, the younger will be the patient at the time of clinical presentation. Discovery at autopsy is rare [1].

In adults, symptoms are directly related to the degree of duodenal obstruction. In some cases, the obstruction is not significant until inflammation of the annulus narrows the duodenum or the diagnosis is made incidentally when the patient is evaluated for symptoms of pancreatitis or peptic ulcer disease [1]. In adults, the

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presenting symptoms are abdominal pain in 70%, nausea and vomiting in 60%, and hematemesis in 10% of patients [1]. Up to 40% of cases require surgery, at which time duodenoduodenostomy or duodenojejunostomy is performed to bypass the obstructed duodenal segment. Division of the ring is not recommended due to high postoperative incidence of complications that include fistula formation, pancreatitis, pancreatic laceration, or recurrent duodenal stenosis secondary to local fibrosis. Imaging is of paramount importance to establish the correct diagnosis and to minimize surgical intervention [4]. Several imaging modalities have been used for detection of annular pancreas in patients of different ages including plain and contrast radiography [4], sonography [5], computed tomography (CT) [6], magnetic resonance imaging [7], and endoscopic retrograde cholangiopancreatography (ERCP) [2]. Magnetic resonance cholangiopancreatography has also been investigated for evaluating biliary and pancreatic ducts noninvasively [8]. The technique has been shown to be useful in detecting pancreas divisum and pancreaticobiliary maljunction. However, its clinical role in routine practice and specifically in evaluating for annular pancreas remains to be defined. Our focus in this article is the imaging of annular pancreas in adults and its variations. Because the diagnosis of annular pancreas in the adult is often overlooked, we evaluated the radiographic appearance of this entity in seven patients with known annular pancreas as demonstrated at laparotomy.

Subjects and methods

Seven patients (five men, two women) with an age range of 14–72 years presented for imaging with a variety of abdominal symptoms: three with recurrent emesis, one with abdominal pain, one with both abdominal pain and emesis, one with tolerance to only small amounts of food and a long-standing history of duodenal obstruction, and one after blunt abdominal trauma. All patients underwent one or more of the following imaging studies: upper gastrointestinal (UGI) series; contrast-enhanced, 7-mm axial, helically acquired CT scan of the abdomen; and ERCP. Laparaotomy was performed in all patients after imaging as a part of clinical care. Imaging and surgical findings were correlated in each patient.

Results

Plain and UGI radiography

In one patient, plain radiography demonstrated duodenal narrowing associated with variable degrees of gaseous distention in the first portion (Fig. 1A). On UGI series, eccentric narrowing and medial retraction of the duodenal sweep at the level of the annulus and an aberrant pancreatic duct were seen in another patient (Fig. 2).

CT

Duodenal abnormalities associated with annular pancreas such as duodenal web could be detected with CT (Fig. 3). CT could directly demonstrate the pancreatic tissue, which completely (Fig. 1B) or partly (Fig. 4) encircled the duodenum. CT was useful in delineating coexistent abnormalities in the patient with blunt abdominal trauma (Fig. 5). CT was also able to differentiate the complete encircling of the duodenum by pancreatic tissue and an associated circumferential thickening of the duodenal wall (Fig. 6).

ERCP

In one patient who had an ERCP procedure, the classic appearance of the annulus pancreatic duct completely encircling the second portion of the duodenum was seen (Fig. 7).

Discussion

A double-contrast UGI study is generally superior to a single-contrast study in fully evaluating the duodenum [1]. If near-complete obstruction of the duodenum exists, only proximal dilatation may be seen, with a slight trickle of contrast through the narrowed segment. With lesser degrees of obstruction, there are eccentric narrowing and medial retraction of the duodenal sweep at the level of the annulus [1]. The duodenal mucosa is usually normal unless there is an associated process such as pancreatitis or peptic ulcer disease. Other ancillary findings may include gastric distention, pyloric incompetence, and reverse peristalsis [1]. The differential diagnosis includes pancreatitis, pancreatic carcinoma, post bulbar ulcer, and duodenal diverticulitis that may coexist with annular pancreas [1].

On CT, especially with a good arterial phase, the pancreatic tissue will enhance throughout and can be readily differentiated from the duodenum that contains changeable amounts of gas, fluid, or contrast. CT is also useful in delineating coexistent abnormalities such as trauma, pancreatitis, or pancreatic tumor. On intravenous contrast-enhanced CT, the partial or complete encircling of the duodenum by pancreatic tissue and apparent circumferential thickening of the duodenal wall are seen, which may be associated with duodenal stenosis. Negative intraluminal contrast such as water is

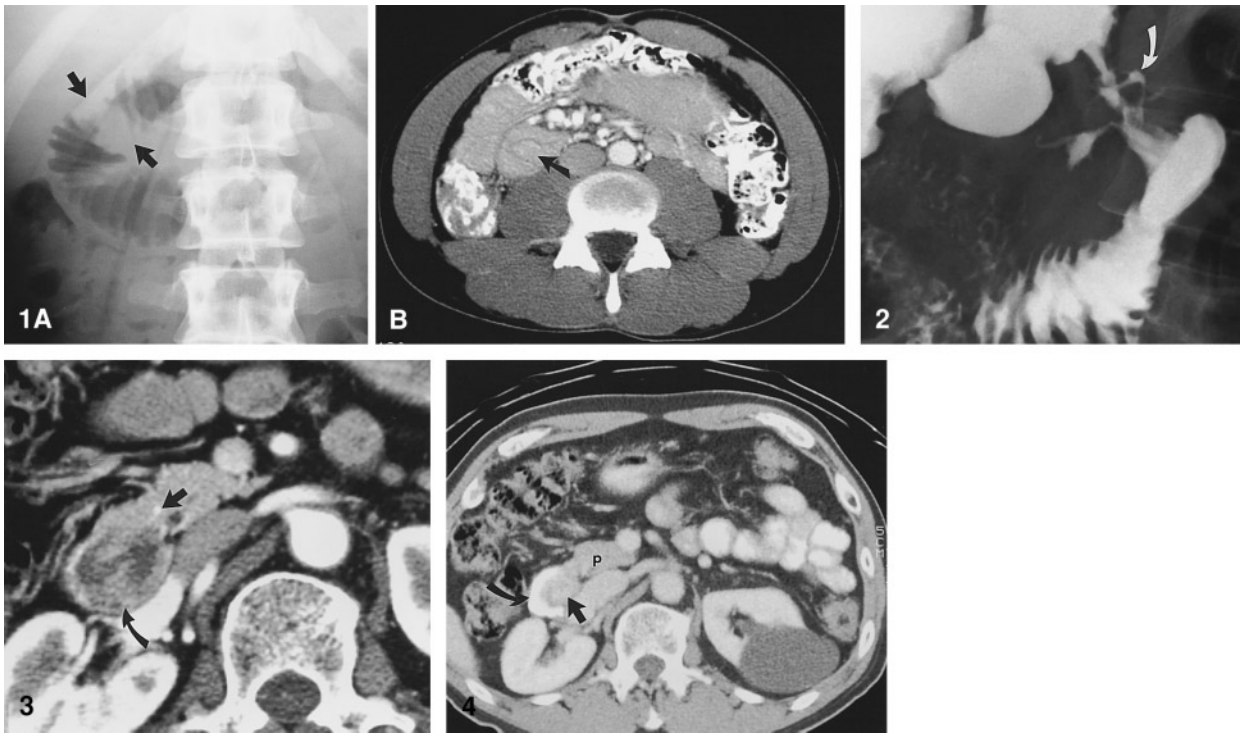

Fig. 1. A Annular pancreas in a 21-year-old man with a long history of abdominal pain and vomiting. The annular pancreas (*arrows*) is outlined by gas within the duodenal sweep. There was massive gastritic distention secondary to a high-grade duodenal obstruction confirmed by an UGI series and subsequent surgery. **B** On the CT scan, the pancreas is noted to encircle the descending duodenum completely (*arrow*).

Fig. 2. Annular pancreas in a 32-year-old woman with repeated episodes of vomiting. On this UGI series, the anomalous duct fills with barium (*arrow*) and encircles the duodenum. There is high-grade stricture of the second portion of the duodenum, resulting in moderate obstruction.

Fig. 3. A 70-year-old man with long-standing obstruction who was only able to tolerate small amounts of food. On CT, the duodenum is massively distended and the edge of the obstructing duodenal web (*curved arrow*) is visualized. In addition, there is a partial annular pancreas that incompletely encircles the duodenum. The anterior superior pancreaticoduodenal artery (*straight arrow*) is visualized within the substance of the anomalous pancreas.

Fig. 4. A 72-year-old man with a life-long history of vomiting. The CT scan demonstrates a partial annular pancreas (*straight arrow*) extending from the main body of the pancreas (*p*) and partly occluding the descending portion of the duodenum (*curved arrow*).

useful for improved differentiation between pancreatic tissue and the duodenum.

ERCP is particularly specific in the diagnosis of annular pancreas. A diagnostic accuracy of 85% has been reported when the duct of the pancreatic annulus opens into the main pancreatic duct [1]. On ERCP, two major patterns are seen [2]. In the less frequent pattern, fusion of the ventral and dorsal pancreas ductal systems has not occurred. In the more common pattern, fusion of the two ductal systems is present. In this latter form, the annulus duct is seen to originate on the left anterior duodenal surface and course posteriorly around the duodenum to open into the main pancreatic duct or the common bile duct near the ampulla of Vater [1, 2]. ERCP is also able to evaluate other diagnoses such as choledochal cyst, aberrant biliary ducts, pancreas divisum, choledochocele, Caroli disease, choledocholithiasis, tumor, or pancreaticobiliary congenital abnormalities [9]. Furthermore, in some cases, an interventional endoscopic procedure, such as sphincterotomy and biliary stent placement, may be performed immediately after diagnosis.

Conclusion

Annular pancreas is an uncommon congenital abnormality that occasionally first presents in adulthood. It may be associated with variable degrees of duodenal stenosis and is often overlooked as the diagnosis for patient's clinical presentation. UGI radiography, CT, and especially ERCP are useful diagnostic imaging tools, obviating the need for surgical confirmation in most cases with its associated risks and cost.

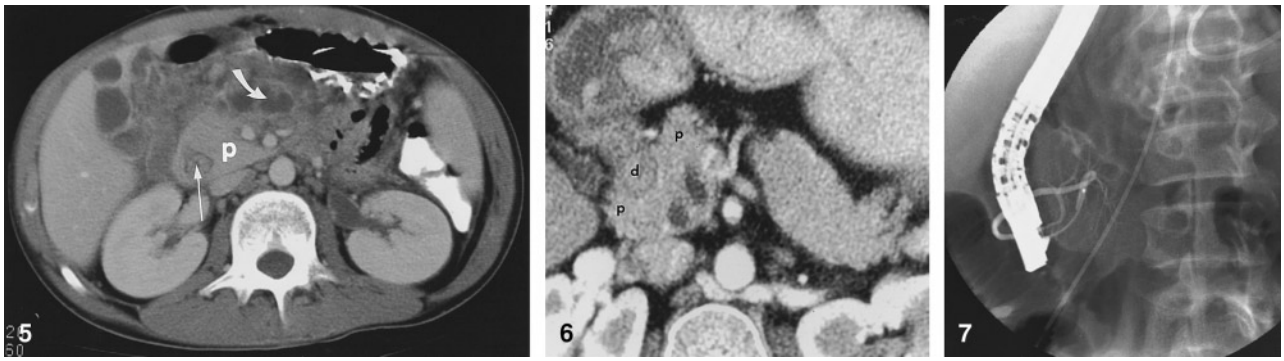

Fig. 5. Blunt abdominal trauma in a 14-year-old boy resulting in a pancreatic fracture. The descending duodenum (*straight arrow*) is surrounded by the pancreas (*p*). There is a laceration of the pancreas (*curved arrow*) at the junction between the neck and body of the pancreas, with accumulation of fluid and dissection of pancreatic enzymes around the superior mesenteric vein. The patient sustained mild abdominal trauma during a game of basketball. It is felt that an anomaly of pancreatic fusion makes a patient susceptible to severe consequences secondary to even minor trauma.

Fig. 6. A 38-year-old woman with recurrent vomiting. The stomach is massively dilated. The duodenum (*d*) is completely surrounded by the pancreas (*p*). The common bile duct and the pancreatic duct are mildly dilated.

Fig. 7. A 30-year-old man with abdominal pain. The ERCP demonstrates the classic appearance of an annular pancreas, with the duct completely encircling the descending duodenum (identified by the tip of the endoscope). Normal ductal arborization is present.

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