

Annular Pancreas, A Rare Cause of Acute Pancreatitis: A Case Report and Literature Review

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ABSTRACT

Annular pancreas is a rare congenital anomaly where a band of pancreatic tissue encircles the second part of the duodenum. Owing to its rarity, neither the true prevalence nor the precise etiology of annular pancreas is known. It is usually diagnosed incidentally on imaging, during an abdominal surgery, or on autopsy. Treatment is individualized according to symptoms, ranging from close follow up to surgical interventions.

In this case report, we present a case of annular pancreas, including the patient's history and clinical assessment, as well as the diagnostic and therapeutic modalities.

INTRODUCTION

Annular pancreas (AP) is an embryologic defect in which pancreatic tissue encases the duodenum, either partially or completely [1]. In 85% of cases, the pancreatic band encircles the second part of the duodenum above the Ampulla of Vater [2]. The diagnosis of AP is made either prenatally or during the neonatal period and, if uncomplicated, it might be discovered incidentally during adulthood or even post-mortem [1]. The complications of AP include intestinal obstruction, acute and chronic pancreatitis, biliary obstruction, peptic ulcer disease, and cholecystolithiasis [1, 2]. There are not well-established guidelines regarding the management of AP, therefore the treatment remains to be individualized [1].

CASE PRESENTATION

On the third of March of 2022, a 55-year-old male patient presented to the emergency department complaining of epigastric pain of 2 days duration. The pain was sharp, intermittent, and radiated to the back. It was associated with nausea, post-prandial vomiting, and a loss of appetite. It was aggravated by food and alleviated by analgesics. There was no change in the patient's bowel habits, and he denied any urinary symptoms.

The patient's first episode of epigastric pain was during the beginning of the previous year (January 2021). Since then, the patient was admitted to the hospital four times for acute pancreatitis, and once for hypokalemia. Moreover, he reported an unintentional loss of 25 kilograms over a period of 10 months.

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His medical history also was significant for an allergic reaction to penicillin and a combination of paracetamol and diclofenac (diclogesic) and peptic ulcer disease. He had only had one surgical procedure in the past for hemorrhoids.

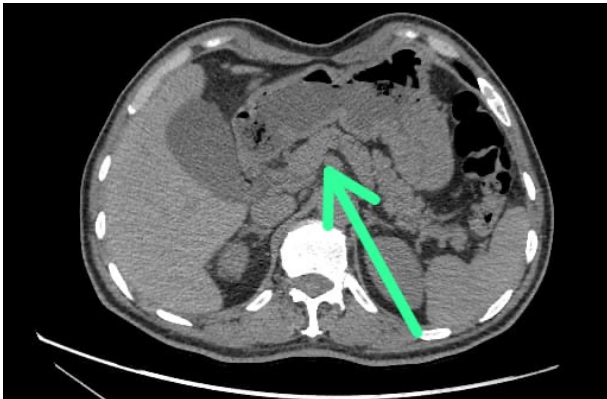
On physical examination, the patient appeared pale. He had clinical jaundice and epigastric tenderness.

His lab results showed a significant increase in amylase levels.

A CT scan of the chest, abdomen, and pelvis was obtained. It revealed a narrowing in the second part of the duodenum because of a near total encasement by pancreatic tissue. The head of the pancreas was enlarged, the pancreatic duct was prominent, and a focal dilatation of the common bile duct of 1cm distally were noticed.

The CT scan report was correlated with the MRCP findings and were suggestive of annular pancreas. [Figure 1]

Figure 1. An Abdominal CT scan. The second section of the duodenum is narrowed due to entirely full encasing by pancreatic tissue



The patient underwent a distal gastric to jejunum anastomosis (40 cm distal to the ligament of Treitz) and Braun jejunojunctionostomy. Intra-operatively, a dilated proximal duodenum and an annular pancreas were noted. The liver, spleen, stomach, small and large bowels were all grossly unremarkable. The patient had stable post operative recovery course and was discharged home on post operative day 2.

DISCUSSION

Acute pancreatitis is the inflammation of the pancreatic glandular parenchyma, leading to the injury of pancreatic acini [3]. It occurs is due to the early activation of trypsinogen to tryp-

sin, which activates enzymes such as elastase and phospholipases, leading to local pancreatic destruction and activation of the inflammatory cascade. The pathophysiology of acute pancreatitis involves the loss of intracellular and extracellular compartments, obstruction of pancreatic secretory transport through the annular duct (Santorini duct), [3] and results in the systemic inflammatory response syndrome (SIRS) [4]. Pancreatitis is associated in 80 to 90% of the cases with alcoholism and biliary tract disease [5]. The remaining 10 to 20% of cases are associated with other tumors, infections, trauma, drugs, lipid abnormalities, postoperative changes, congenital anomalies, or are idiopathic [5-7].

In patients with annular pancreas, the pathophysiology of pancreatitis is still unclear. Pancreatitis is caused by fibrosis, which results a partial occlusion of the pancreatic duct in the pancreatic head. In acute pancreatitis, pancreatic fibrosis mostly only affects the annulus and adjacent pancreatic head, whereas the body and tail are generally spared [1].

The development of the pancreas is a complex process. Anomalies including fusion, migration, or duplication can result from errors occurring during the fusion of the dorsal and ventral pancreatic buds [8]. The pancreas begins to develop in the fourth to eighth week of gestation as two outpouchings of the endodermal lining of the duodenum, the outpouchings are the ventral and dorsal pancreas [1]. By the seventh gestational week, the ventral bud rotates, passes behind the duodenum from right to left, and fuses with the dorsal bud due to duodenal expansion. The dorsal bud gives rise to the tail and the body of pancreas, whereas the ventral bud forms the inferior head of the pancreas and the inferior part of the uncinate process. The major pancreatic duct is formed by the fusion of the two buds' ducts. The failure of the ventral bud to rotate with the duodenum leads to envelopment of the duodenum, which results an annular pancreas [3]. The development of annular pancreas has been explained by many theories: Lecco's theory illustrated that there is adhesion of the right ventral bud to the duodenal wall. Baldwin's theory proposed persistence of the left ventral bud. And a third theory suggests that during rotation, a small part of the left ventral bud adheres to the wall of the duodenum with pancreatic tissue [9,10].

The precise prevalence of annular pancreas is unknown due to its asymptomatic nature. A multi-institutional database in the United States reported that there were 210 individuals (3.4/100,000) had a diagnosis of annular pancreas between April 2015 and April 2020 [11].

AP has been classified into a complete or incomplete, depending on the morphologic distribution of pancreatic tissue. In the complete type AP, the second part of the duodenum is entirely encircled by pancreatic parenchyma or an annular duct, as confirmed by macroscopic examination. In the incomplete type AP, there is partial circumferential encasement of the duodenum by pancreatic tissue confirmed by endoscopic retrograde cholangiopancreatography (ERCP) or surgical evaluation [7].

Annular pancreas is frequently associated with other congenital anomalies. If gone undetected and diagnosed later in adulthood, it can present with chronic or acute biliary colic symptoms and acute pancreatitis. The annular pancreas, either complete or incomplete, should be taken into consideration in the differential diagnosis of acute pancreatitis in the absence of major risk factors for pancreatitis, including cholelithiasis, alcoholism, viral infections, hypertriglyceridemia, or malignancy [9].

The most common symptoms in adult patients with annular pancreas are abdominal pain, post-prandial fullness, and vomiting. The most frequent complications are peptic ulcer, upper gastrointestinal bleeding, pancreatitis, and may progress to duodenal obstruction caused by chronic inflammation and fibrosis of the pancreatic annulus [12].

The initial CT scan typically confirms the diagnosis of pancreatitis caused by an annular pancreas as it shows a ring of inflamed pancreatic tissue encircling the duodenum. Sometimes, the Santorini duct encircling the duodenum or even pancreatic lithiasis may be seen on the CT scan. In case of associated duodenal obstruction, a gastroduodenoscopy reveals concentric narrowing and pre-stenotic duodenal dilatation. In upper gastrointestinal double contrast studies, the classic signs of an obstructive annular pancreas are the presence of an annular filling defect of the duodenum associated with a proximal dilation of duodenal bulb and stomach ("double bubble" sign) [13].

There are no specific guidelines and protocols for the management of pancreatitis caused by annular pancreas [7]. Supportive treatment is effective for treating symptomatic adult individuals who have acute pancreatitis. Patients who present with persistent duodenal and gastric outlet obstruction typically require surgical intervention. Duodenal bypass surgery is the best method for both adults and children when surgical intervention is required. Gastrojejunostomy or duodenoduodenostomy can be used to accomplish relief of obstruction [1].

LITERATURE REVIEW

There are very few case studies reported regarding pancreatitis due to annular pancreas as a result of the rarity of this condition. For example, in a study conducted in San Giovanni Battista Hospital in Italy from 2007 to 2014, only twenty cases were diagnosed with annular pancreas; from which, only two cases showed acute pancreatitis while three cases presented with duodenal obstruction [14]. Moreover, another case study performed in France demonstrated a symptomatic AP with pancreatitis in an old age patient [6].

CONCLUSION

Pancreatitis is an unusual presentation of annular pancreas in adult patients. There are not enough studies pancreatitis secondary to AP in adults and therefore, no specific management guidelines. Treatment protocols must be individualized according to associated complications. Advancements in imaging diagnostic modalities and an index of suspicion may help in reporting more cases that essential for development of such guidelines.

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MAA, MAK and HA wrote the first manuscript. SA, MAS were the physicians of the patient and revised the final manuscript. MAJ lead the team revised, edited and submitted the manuscript.

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DISCLOSURES

The author have declared that no competing interests exist.

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