

Partial Annular Pancreas- A Time Bomb?

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Abstract

Case Report

Annular pancreas is a rare developmental anomaly. Partial annular pancreas is less known and is very rare. A band of pancreatic tissue continues from the pancreatic head and incompletely encircles the pancreas. This typically gives a “crocodile jaw” configuration. In symptomatic cases patients present mostly in 3rd to 6th decade of their life, with abdominal pain, postprandial fullness, vomiting, upper gastrointestinal bleeding, acute or chronic pancreatitis, and in rare instances, biliary obstruction. The person can also be asymptomatic; in that case it will be an incidental finding. We are presenting a case of partial annular pancreas causing nonspecific symptoms. This was an incidental finding, studied and managed in a tertiary care centre.

Keywords: Partial annular pancreas, crocodile jaw configuration, Computed tomography, incidental finding.

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INTRODUCTION

Normally, during embryogenesis between the first 4–8 weeks of embryonic life, the pancreas is formed by the fusion of the dorsal and ventral pancreatic buds. But when there is a failure of fusion of the ventral and dorsal pancreatic buds, the pancreatic tissue can encircle the duodenum. The classification of this embryologically abnormal pancreas into partial/incomplete and complete types depend upon the degree of duodenal encasement.

Annular pancreas is a rare, but a well-known developmental anomaly. In this entity, a portion of pancreas completely (for 360 degrees) encircles the duodenum and results in varying degree of duodenal obstruction. It can result from an adhesion of the duodenal wall to the right ventral bud or from the failure of the left ventral bud to regress. There can be association of other conditions for example Down syndrome, duodenal atresia and Hirschsprung disease. It can affect a wide age range, from neonates to the elderly, when diagnosed. It can present in a wide range of clinical severities, thus making it difficult to diagnosis. Pre-operatively by upper GI series, upper GI endoscopy, or CT scan is possible. In many cases a final diagnosis can be reached only by surgery [1, 2].

Partial annular pancreas is a rarer entity. In this a band of pancreatic tissue, in continuity with the

pancreatic head, incompletely encircles the pancreas. It is thus termed as incomplete/partial annular pancreas. The shape typically assumes a “crocodile jaw” configuration. This is a very rare entity and the exact prevalence of incomplete/partial annular pancreas is not clear. Till date, only a few cases are found in the literature [2, 3]. The main problem with a partial annular pancreas is that, it can be totally asymptomatic and later can present with complications. It is often diagnosed incidentally during imaging work-up for some other complaint. Therefore, knowledge of the imaging appearances of partial or incomplete annular pancreas can significantly reduce patient morbidity and mortality. Early diagnosis can result in an early and proper management.

CASE REPORT

A 32-year-old male patient was referred to department of Radiology from the department of Urology, in All India Institute of Medical Sciences Rishikesh. He had complaints of dull aching pain in the left flank for about a year. He had complaint of early satiety and nausea in early post-prandial period. There was history of mild intermittent reflux. No significant history of vomiting or fever was present. He otherwise had no comorbidities. On examination there was no tenderness in the abdomen. Systemic examination otherwise was unremarkable. Liver function tests and routine blood investigations were normal. On referral to

the Department of Radiology, computed tomographic (CT) urography was done based on his complaint of left flank pain. The scan was done using Siemens Somatom 10 Definition Flash dual source CT 128 slices machine.

IMAGING FINDINGS

On CT Urography, a non-obstructive calculus was seen at the lower pole of left kidney. As an incidental finding, the pancreatic tissue at pancreatic head was seen incompletely encircling the second part of duodenum for ~270 degrees, instead of the normal smooth rounded contour of pancreatic head. This claw-like projection of the pancreatic parenchyma partially encasing the second part of duodenum is called a typical “Crocodile jaw” appearance. The computed tomographic (CT) images are depicted in figures 1 (axial section) and figure 2 (coronal section). No evidence of proximal obstruction or wall thickening was present. There was normal enhancement of pancreatic parenchyma. Pancreatic duct and CBD were normal in diameter without any calculus or dilatation. The pancreatic duct was normally opening into the second part of duodenum without any accessory drainage. Acute pancreatitis was ruled out by preserved peri-pancreatic fat planes. Based upon these CT findings, he was diagnosed to have partial/incomplete annular pancreas. He was referred to the treating physician in department of Urology for further work-up.

AXIAL SECTION ON CT

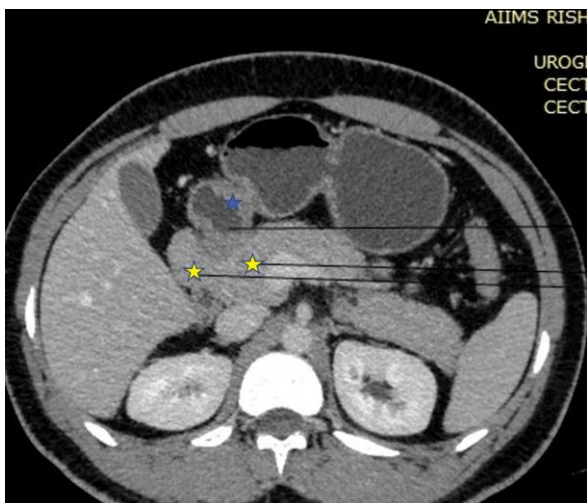


Fig-1: 32 years old male. Axial section of abdomen on contrast-enhanced computed tomography (CECT), showing second part of duodenum (marked by blue star), being incompletely encircled by pancreatic tissue (marked by yellow stars). A typical crocodile jaw configuration of the pancreatic tissue is seen around the second part of duodenum

Coronal section on CT

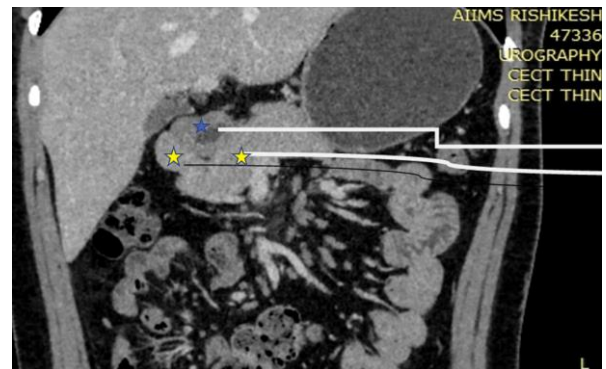


Fig-2: 32 years old male. Coronal section of abdomen on contrast-enhanced computed tomography (CECT), showing second part of duodenum (marked by blue star), being incompletely encircled by pancreatic tissue (marked by yellow stars). A typical crocodile jaw configuration of the pancreatic tissue is seen around the second part of duodenum

DISCUSSION

The embryologic development of the pancreas starts at about the 5th week of gestation. Initially, there is one dorsal bud and two ventral buds. At about the 7th week of gestation, two ventral buds join together. This ventral tissue rotates with the duodenum and forms the head of the pancreas and the uncinate process. The body and tail of the pancreas are formed by the dorsal bud. Thereafter, there is atrophy of the left ventral bud and the right ventral bud normally rotates to the left of the duodenum. This forms the lower part of pancreatic head. The basis of formation of annular pancreas is due to failure of the ventral and dorsal buds to fuse. The duodenum gets entrapped, mostly in the posterior part, within the pancreatic tissue [1].

The anomalies developing annular pancreas can be explained by two main theories given by Lecco and Baldwin. According to Lecco, there is adhesion of the duodenal wall to the right ventral bud. This allows a part of pancreatic tissue to envelop the duodenum. As per the theory given by Baldwin, the failure of the left ventral bud to regress results in annular pancreas. The classification into partial /incomplete and complete types depend upon the degree of duodenal encasement. Complete annular pancreas most commonly presents as small bowel obstruction, during neonatal period or early childhood. Annular pancreas can be classified into the extramural and intramural subtypes as per Johnston. The duodenum is encircled by the ventral pancreatic duct which then joins the main pancreatic duct, resulting in the extramural annular pancreas. MRCP or ERCP images can easily show the duct encircling second part of duodenum.

Annular pancreas is an infrequently reported anomaly. Reportedly in adults, one half to two thirds of cases of annular pancreas can remain asymptomatic. Complete annular pancreas has an estimated prevalence of 5–15 cases per 100,000 adults on autopsy. On ERCP, this is approximately 400 cases per 100,000 people [1]. Rarely, there can be association of other congenital abnormalities such as trachea-oesophageal fistula, Down syndrome, oesophageal atresia, imperforate anus or Hirschsprung's disease [1].

Incomplete annular pancreas is a very rare entity with unknown exact prevalence. Only few cases of Incomplete annular pancreas have been reported. In contrast to annular pancreas, the patients with incomplete or partial annular pancreas can be totally asymptomatic. It may be diagnosed incidentally on imaging. Sometimes, they may remain asymptomatic until adulthood and present symptoms like pancreatitis, duodenal ulceration, and rarely, biliary or intermittent

bowel obstruction [2-4]. Annular pancreas is often an overlooked cause of duodenal obstruction in adult population.

The extension of pancreatic tissue in relation to duodenum can be classified into three types: a) Anterolateral extension, b) Posterolateral extension, and c) Both anterior and posterior extension [2, 3]. The third type (c) gives a crocodile jaw appearance, as depicted the schematic diagram in figure 3. Out of these, anterolateral extension is least specific, while crocodile jaw appearance is considered highly specific of incomplete annular pancreas. This entity is often misdiagnosed and may be undetected. Diagnosis is especially difficult in patients not presenting with features of duodenal obstruction. In totally asymptomatic cases it is an incidental finding, like in our case that had chief complaint of left flank pain [1-4].

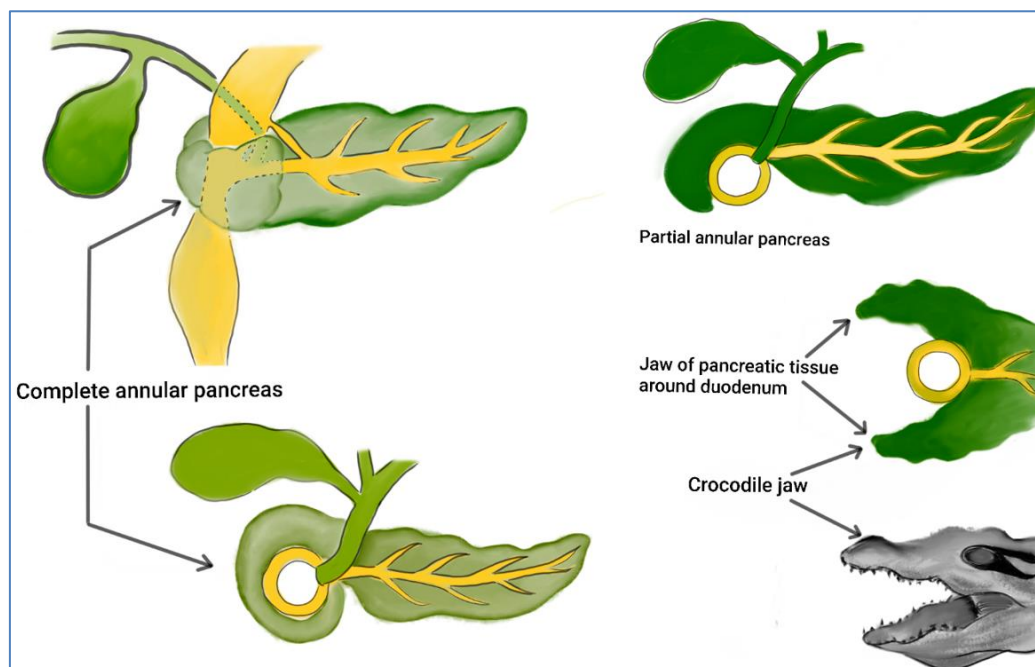


Fig-3: Schematic diagram showing both anterior and posterior extension of pancreatic tissue in relation to duodenum, giving a crocodile jaw appearance

The preoperative diagnosis can be made by Endoscopic Retrograde Cholangiopancreatography (ERCP) or Magnetic Resonance Cholangiopancreatography (MRCP). MRCP is useful of defining any associated ductal anomalies [8]. Multidetector multiplanar computed tomography (CT) can easily diagnose this entity. In a 128 slices helical CT scanner, like used in our case, time required for scanning is minimum. Thus, CT is a fast modality for its detection. With increasing awareness of this condition, more cases are being detected preoperatively [5]. Diagnosis of complete annular pancreas is straightforward with a complete ring of pancreatic tissue encircling the second part of duodenum. The finding of incomplete annular pancreas may be more

subtle and requires a careful examination to come into final diagnosis [6]. In symptomatic cases, treatment is usually by bypassing the duodenal obstruction by duodenoduodenostomy or gastrojejunostomy [7, 8].

In the past ERCP used to be the modality of choice for the diagnosis of annular pancreas. At present, it is being increasingly diagnosed on the basis of cross-sectional imaging such as CT or MRI. MRCP has become the preferred imaging modality. MRI provides excellent evaluation of the pancreatic parenchyma especially with the use of fat suppressed T1-weighted images. It is a brilliant technique to show the pancreatic tissue (annulus) encircling the second part of duodenum; and in ascertaining the degree of

encirclement (complete/partial). The use of heavily T2-weighted images in MRCP enables excellent evaluation of the biliary tract and pancreatic duct and can demonstrate well the duct within the encircling annulus [8].

Management in Our Patient

Our patient complained of dull aching pain in the left flank with satiety and nausea in early post-prandial period and intermittent reflux. There was no significant history of vomiting or fever. Liver function test (LFT) was not deranged. We advised him MRCP and for follow-up. As he did not have any acute or distressing complaints he opted to wait and watch. He was explained about the risk of pancreatitis and pertinent lifestyle changes were advised. After 1.5 years of follow up he still did not have any further distressing complaint and he is leading a healthy lifestyle.

CONCLUSION

Incomplete or partial annular pancreas can be missed radiologically due to asymptomatic patients and subtle findings. But its diagnosis is important as it may be associated with other abnormalities. A clinical suspicion assists in a proper clinical scenario assists in finding more cases of symptomatic and asymptomatic incomplete annular pancreas in adults. Intestinal obstruction and future development of pancreatitis is significant in asymptomatic cases. Therefore, follow up plays a very significant role in its management. The management of this congenital anomaly should be individualized according to the associated complications. The advancements in imaging modalities with increasing knowledge of imaging features of incomplete annular pancreas will help diagnose more cases earlier to development of complications.

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