



CIRCUMPORTAL PANCREAS: CASE REPORT OF A RARE PANCREATIC ANOMOLY.

Radio-Diagnosis

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ABSTRACT

Circumportal pancreas/ Portal annular pancreas is a rare congenital pancreatic anomaly resulting from fusion of the pancreatic parenchyma around the confluence of portal and superior mesenteric veins. It is usually asymptomatic, but if undetected, it could have serious consequences during pancreatic surgery. We describe a variant of this anomaly encountered at our institute during routine examination of an abdominal contrast enhanced computed tomography and ultrasonography.

KEYWORDS

Circumportal pancreas, Portal annular pancreas, pancreatic anomalies,

INTRODUCTION:

There are three types of pancreatic fusion anomalies: annular pancreas, pancreas divisum and Circumportal pancreas/Portal annular pancreas. Circumportal pancreas is the rarest of these and is mostly asymptomatic. Previously, this variant had only been reported post-operatively during surgical resection of pancreas in pancreaticoduodenectomy or pancreaticojejunostomy. To our knowledge this is the first case report which was incidentally found during routine CT and USG examination of a case of acute pancreatitis.

CASE REPORT

We report a 17-year-old male who presented to emergency department with epigastric pain and vomiting since 4 days. An abdominal ultrasonography was performed which showed bulky pancreas with per-pancreatic fat stranding suggestive of Acute pancreatitis and ascitis. Serum lipase was 481 U/L. An abdominal contrast enhanced computed tomography was also performed. CT scan showed enlarged pancreas with heterogenous post-contrast enhancement and patchy areas of necrosis, peri-pancreatic fat stranding and collection, the uncinate process was fused with pancreatic body wrapping around the confluence of portal vein and splenic vein, partial thrombosis of splenic vein and the main pancreatic duct lying anteriorly to the portal vein.

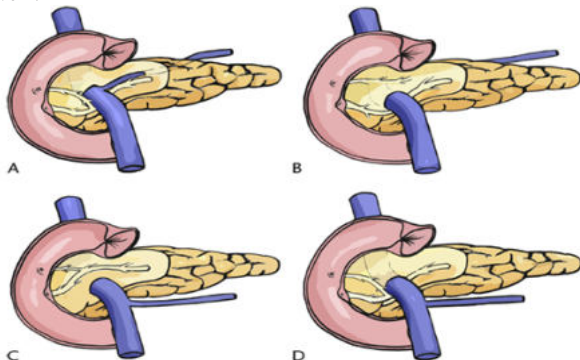


FIGURE 1. Classification of Circumportal pancreas[1]. According to Joseph et al.[2] PAP is classified (types 1–3) depending on the topography of the MPD; type 1 is the fusion of the ventral bud of the pancreas with the body and RMPD (A); type 2 is type 1 associated with pancreas divisum (B); type 3 is the pancreatic encasement with a normal AMPD (C). Following Karasaki et al.[3] each type can be subdivided (A, B, and C) depending on the relation to the portal confluence: suprasplenic (C), infrapleural (B), and mixed type (A). The most common type of PAP is 3A (C) and the second most common type is 1A(D).

FIGURE 2. Circumportal pancreas: Contrast-enhanced CT axial image showing encasement of portal and splenic vein by pancreatic head and uncinate process. (white arrow: portal confluence, white arrow head: splenic vein)

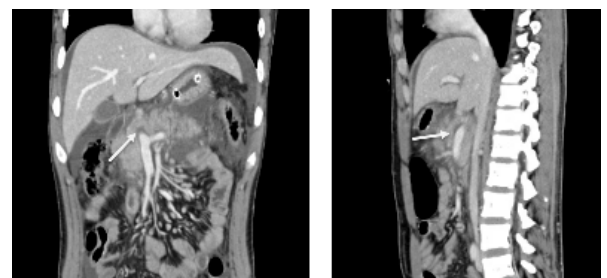
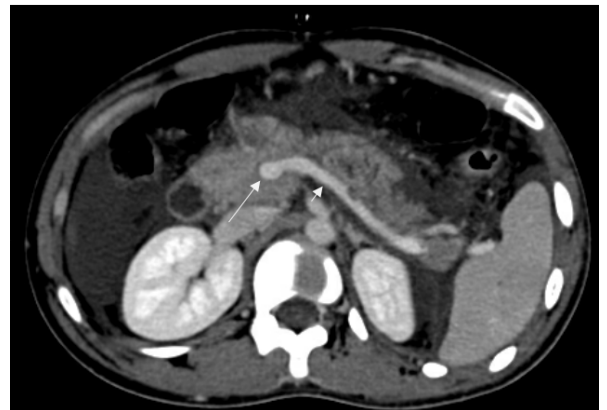


FIGURE 3. Circumportal pancreas: Contrast-enhanced CT coronal and sagittal images showing encasement of portal and splenic vein by pancreatic head and uncinate process (White arrow) The pancreatic neck and tail is above the splenic vein i.e Type a.

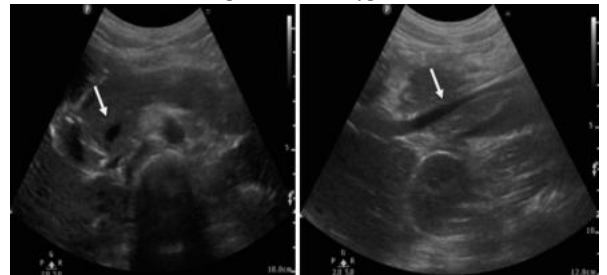


FIGURE 4. Circumportal pancreas: Ultrasonography images in axial and sagittal planes showing encasement of portal and splenic vein by pancreatic head and uncinate process (White Arrow).

DISCUSSION:

During embryogenesis, the pancreas is formed by 2 buds originating from the endoderm in the primitive duodenum. The caudal part of the head and the uncinate process of the pancreas are derived from the

ventral bud, whereas the cranial part of the head, body, and tail of the pancreas is derived from the dorsal bud. After the duodenum rotation, the ventral pancreatic primordium moves dorsally below and behind the dorsal bud. Toward the end of the sixth week, the two primordia fuse and the ducts anastomose, forming the main pancreatic duct (MPD). The uncinate remains its own pancreatic duct, the inferior branch of the pancreatic duct.

Two different hypotheses are discussed regarding the development of circumportal pancreas: either it is a malformation of the portal venous system [4,9] or the pancreas caused by hypertrophy of the ventral [5] or dorsal anlagen and subsequent fusion to the left of the mesenteric or portal vein.

An incidence of 1.14% has been reported by Karasaki et al., based on an institutional review of CT scans [3,5]. The possibility of this anomaly during pancreatic head resection was first described by Sugiura et al. [6]. Complete fusion of the uncinate process with the body of the pancreas was described by Hamanaka et al. during a resection of the pancreas [7].

Circumportal pancreas is a rare congenital anomaly of the pancreas where there is an abnormal fusion of the uncinate process to the main pancreatic body occurring to the left of the portal vein-superior mesenteric vein junction, resulting in complete encasement of the portal vein or superior mesenteric vein [8]. While this occurs normally in pigs, in humans the incidence ranges from 1.1 to 2.5%. [5] It was first reported in 1987 by Sugiura et al. [6] as hypertrophy of the uncinate process of the pancreas encircling the superior mesenteric vein and artery. Various terms have been used to describe it, including portal annular pancreas, [2,3] periportal annular pancreas, complete pancreatic encasement of the portal vein and superior mesenteric vein running through the pancreas. [9,10] Owing to the rarity of the condition, it has been documented mainly as case reports and small case series.

This condition may be diagnosed by a number of ways. Preoperative CT scans can reveal pancreatic tissue completely encasing the portal vein and/ superior mesenteric vein. Arterial phase imaging may be useful for demonstrating any variant arterial anatomy. The course of the pancreatic ducts may be better delineated by Magnetic Resonance Cholangiopancreatogram (MRCP), in which a pancreatic duct ring sign has been proposed to be diagnostic, [11] or by intraoperative ultrasonography. Most often though, this condition is diagnosed by direct visualization during surgery when the pancreatic tissue is seen to be completely wrapped around the portal vein-superior mesenteric vein junction.

In our patient who presented with complaints of epigastric pain and vomiting since 4 days, CECT of abdomen showed the uncinate process was fused with pancreatic body wrapping around the confluence of portal vein and splenic vein. The main pancreatic duct was anterior to the portal vein. The pancreas was enlarged in size with heterogeneous post-contrast enhancement and patchy areas of necrosis, peripancreatic fat stranding and collection. The patient was diagnosed with Acute pancreatitis in a known case of Circumportal pancreas. This pancreatic anomaly is rare but needs to be diagnosed on imaging and informed to the operating surgeons in prior if required in future.

CONCLUSION:

Circumportal pancreas is associated with increased risk for post operative pancreatic fistula. Dissection of retroportal pancreatic tissue and pancreaticojejunal reconstruction is technically challenging during pancreatic surgeries. To reduce the risk of postoperative pancreatic fistula it is necessary to pre-diagnose such anatomic variants. Because of its low degree of familiarity, Circumportal pancreas remains a diagnostic and operative challenge for the radiologist and surgeons, respectively, to avoid preoperative misdiagnosis and postoperative complications.

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