Agenesis of the Dorsal Pancreas (ADP)

MADHURI PANIGRAHI, SANTOSH KUMAR PANDA

ABSTRACT

Agenesis of the dorsal pancreas (ADP) is a rare congenital anomaly. We describe two patients with ADP who presented with acute pain abdomen. Abdominal computed tomography (CT) revealed a normal pancreatic head, but pancreatic body and tail were not visualized. Magnetic resonance imaging (MRI) findings were similar to CT. At magnetic resonance cholangiopancreatography (MRCP), the major pancreatic duct was short and the dorsal pancreatic duct was not visualized. Conclusion of ADP was made. One patient had omental infarction as an additional feature.

Keywords: Agenesis; Pancreatic anomaly; Computed tomography; Magnetic resonance cholangiopancreatography

Introduction

Agenesis of the dorsal pancreas is an extremely rare anomaly which results from defective pancreas formation. Though it is mostly asymptomatic, can present as an incidental finding or with clinical symptoms like abdominal pain, pancreatitis and diabetes mellitus. We report two cases of dorsal agenesis of pancreas which were detected between December 2014 and May 2015 at our institution presenting with pain abdomen.

Case Reports

Case 1: A 40-year-old man presented with recurrent epigastric pain associated with vomiting over a two year period. The pain was continuous and non-radiating in nature. On examination, there was mild epigastric tenderness. Investigations: normal total white count, and amylase and lipase levels. On ultrasound examination the head of pancreas was visible (Fig 1a). The pancreatic body and tail were not identified. CT and MRI scan of abdomen confirmed the absence of body and tail of pancreas (Fig 1b). The pancreatic head was normal in size, density and enhancement pattern. Punctuate calcification at head was seen. No other structural abnormality of abdominal organs was seen. MRCP was further carried out to confirm the diagnosis and identify whether the dorsal duct was present or absent, so as to classify it as partial or complete form of dorsal pancreatic agenesis. The dorsal duct was absent in its entire extent and a short non dilated ventral duct was present, suggestive of complete agenesis of the dorsal pancreas (Figure 1c).

Fig 1a- on USG, only head region of pancreas visible.

Case 2: A 25-year-old male presented with pain in epigastrium and right iliac fossa region. His serum amylase and lipase levels were within normal limits. On investigation his fasting blood sugar level was normal. Ultrasonography examination at our institution revealed enlarged head with absence of body and tail. CT and MRI examination of pancreas showed absence of pancreatic body and tail (Figure 2). No focal calcification in pancreatic head was seen. MRI and MRCP study confirmed absence of pancreatic body, tail as well as the dorsal duct. The ventral duct was present and
The pancreas is formed by ventral and dorsal endodermal buds. The dorsal bud forms the upper part of the head, body and tail of the pancreas and drains through the duct of Santorini. The ventral bud gives rise to the major part of the head and uncinate process which drains through the duct of Wirsung. At 6-7 weeks of gestation the fusion between ventral and dorsal pancreas occurs. During the complex development, congenital abnormalities can occur. Complete agenesis of the pancreas and agenesis of the ventral pancreas are not compatible with life. Agenesis of the dorsal pancreas is described in two forms; partial or complete. In complete dorsal agenesis, the minor papilla, accessory pancreatic duct, body and tail of the pancreas are absent. In partial agenesis, the minor papilla with a remnant of the accessory pancreatic duct and the neck and proximal body of pancreas are present.

Pancreas divisum and autodigestion secondary to chronic pancreatitis must be considered in the differential diagnosis of the dorsal pancreatic agenesis. Failure of the ventral and dorsal pancreatic ducts to fuse is called pancreas divisum. In this entity, the ventral duct drains into major papilla, while the dorsal duct drains into separate minor papilla. Atrophy of the body and the tail of the pancreas secondary to chronic pancreatitis and sparing of the pancreatic head is called pseudo-agenesis. This situation may mimic dorsal pancreatic agenesis. In the differential diagnosis of pseudo-agenesis, histories of previous abdominal pain, pancreatitis, CT scanning and the serum amylase level may be helpful. The complete absence of the dorsal duct and demonstration of short ventral duct is important in the diagnosis of the dorsal bud agenesis. Abdominal CT may not evaluate the pancreatic duct in a detailed fashion. Therefore ERCP or MRCP is necessary for revealing the major and the accessory duct systems. ERCP is an invasive technique. It might challenge the cannulation of the minor papilla in selected cases. There is also radiation risk to the patient. MRCP can help the diagnosis of the dorsal pancreatic agenesis noninvasively with no radiation risk.

The clinical presentation of dorsal pancreatic agenesis can range from complete absence of symptoms to non specific abdominal pain, diabetes mellitus, pancreatitis and sometimes exocrine pancreatic insufficiency. There is no established guideline for treatment of this anomaly. The consensus has been to treat only symptomatic patients with dorsal agenesis of the pancreas.

Pain abdomen has been the commonest symptom seen in approximately 92.9% cases with this anomaly reported in literature. The pain has been assumed to be due to pancreatitis, duodenal obstruction, sphincter of Oddi dysfunction or autonomic neuropathy. In most occasions the pain is localized to epigastric region which is aggravated following meals. Pain unrelated to pancreatitis has been managed by various methods like gastrointestinal decompression, total parenteral alimentation, low fat diet, anti diabetic therapy and analgesics.

Sphincter of Oddi dysfunction or dyskinesia, compensatory hypertrophy and hypersecretion of the remaining ventral pancreas with raised intraductal pressure have been proposed to explain pancreatitis.

The second most common clinical manifestation in association with this anomaly has been diabetes mellitus, which is seen in approximately 43% of cases. Abnormal glucose tolerance has been seen in another 7% of cases with this anomaly.

When diabetes mellitus is associated with dorsal pancreatic agenesis, it is mostly of adult onset type with reported age range between 28 to 39 years. The age of onset of diabetes mellitus in our cases were within the above range. Despite the congenital absence of dorsal pancreas where the beta-cells are maximally located, resulting in impaired insulin secretion, hyperglycemia has been demonstrated only in approximately 50% of reported patients with this anomaly.

Other associated anomalies could be polysplenia, replaced right hepatic artery from superior mesenteric artery, gastroduodenal...
artery originating from right hepatic artery and bicornuate uterus.

In summary, this extremely rare congenital anomaly must be kept in mind when the corpus and tail of the pancreas are not seen at routine examinations or as in our case an incidental finding at the examinations for different pathologies.

**REFERENCES**


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