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Dorsal Pancreatic Agenesis

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Abstract

Dorsal Pancreatic Agenesis (DPA) is a rare congenital anomaly ,agenesis that results from the embryological failure of the dorsal pancreatic bud to form the body and tail of the pancreas. Present case of dorsal pancreatic agenesis have been reported in GMC ,Latur. A 11-year-old girl presented with abdominal pain. Abdominal computed tomography (CT) revealed a normal-appearing pancreatic head, but the body and tail were not visualized. Endoscopic cholangiopancreatogram (ERCP) revealed a short pancreatic duct. Abdominal magnetic resonance imaging (MRI) findings were similar to the CT and ERCP results. The patient was diagnosed with agenesis of the dorsal pancreatic bud by CT, ERCP and MRI.

Key Words : Agenesis; Dorsal Pancreas;, ERCP; CT.

Introduction

Agenesis of pancreas is very rare anomaly. The pancreas owes its development from the endoderm of the duodenal segment of the foregut. It develops in two parts – dorsal and ventral. The ventral bud forms the lower part of the head and the uncinate process of the pancreas, while the upper part of head , the body and the tail are formed from the dorsal bud. Partial or complete agenesis of the dorsal pancreas is a rare congenital anomaly that results from embryologic failure of dorsal pancreatic budding in the developing fetus.[1,3]

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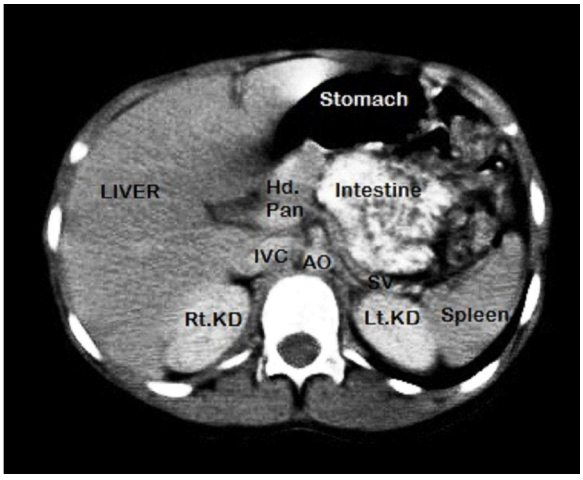
Case report

A 11-year-old girl presented with recurrent episodes of epigastric pain over a four month period. The pain was continuous and non-radiating in nature. Physical examination revealed epigastric tenderness. There was no history of diabetes mellitus. Investigations: elevated serum amylase and lipase levels [1200 U/L(normal values 0-200) and 920 U/L (normal values 0-190), respectively]. Ultrasonography showed only partial visualization of pancreas; the body and tail of the pancreas could not be visualized. CT scan revealed normal head of pancreas; the body and tail of pancreas were absent. Endoscopic retrograde cholangio-pancreatography (ERCP) demonstrated filling of the normal ventral duct only. The minor papilla was not visualized despite careful examination. The common bile duct was normal. The patient was treated conservatively; Absence of the dorsal pancreatic ductal system, accessory duct and minor papilla was documented. Lipid profile, serum calcium and renal function tests were normal. There was no family history . The patient did not have skeletal , dental or cardiac defects.

Discussion

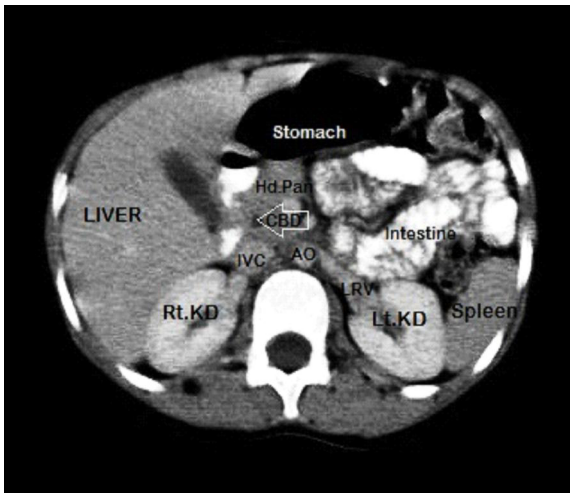
The human pancreas develops from the ventral and dorsal buds of the foregut endoderm.[1] The ventral bud forms the uncinate process and the posteroinferior part of the head. The Wirsung duct drains along with the bile duct through the major papilla. The dorsal bud forms the remaining ventrosuperior part of the head, the isthmus, the body and the tail of the pancreas and drains through the Santorini duct into the minor papilla. Agenesis of the dorsal pancreas is derived embryologically from the absence or regression of the dorsal bud.[1-4] This anomaly may be partial or complete. In partial agenesis of the dorsal pancreas, the minor papilla, duct of Santorini or the pancreatic body are present. In complete agenesis of the dorsal pancreas, the neck, the body and the tail of the pancreas, duct of Santorini and minor papilla are absent. Few cases of agenesis of the dorsal pancreas have been reported in the English literature.[2,3] Complete agenesis of the pancreas is incompatible with life.[5] Agenesis of the dorsal pancreatic bud results in complete absence of the dorsal ductal system. ERCP is necessary to differentiate DPA from "partial agenesis".[6] A diagnostic triad is required, documenting absence of the dorsal ductal system, the accessory duct (Santorini) and the minor papilla. The absence of the body and tail of the pancreas is best demonstrated on CT scan, MRI or MRCP. With increasing availability of MRCP, awareness of DPA is required. DPA condition is very rare; a total of 15 cases have been reported since 1913 till date.[2,7] Abdominal pain and diabetes mellitus are commonly reported. The association of DPA and pancreatitis is less well defined.[8] The possible mechanism contributing to pancreatitis are proposed , Sphincter of Oddi dysfunction may play a role in the pathophysiology of dorsal pancreatic hypoplasia and pancreaticobiliary diseases associated with it.[9] However, the role of genes in the pathogenesis of acute and chronic pancreatitis is increasingly recognized.[10] In southern India, genetic predisposition results in a high incidence of pancreatitis.[11,12] Agenesis of dorsal pancreas is a very rare congenital anomaly that may be associated with diabetes mellitus and abdominal pain. However, hereditary mechanisms may play a role in the development of this anomaly but remains to be further clarified. Wildling et al and Schnedl et al reported familial occurrence of agenesis of dorsal pancreas in the mother and her sons.[13,14] In these reports, the authors suggested that the genetic mode of transmission for this anomaly is most likely autosomal dominant or X-linked dominant. If

Fig 1: CT abdomen showing head of pancreas; body and tail are absent



(Where Hd.Pan is Head of pancreas,IVC is Inferior vena cava, AO is Aorta & SV is Spleenic Vein)

Fig 2: CT abdomen showing head of pancreas



(Where Hd.Pan is head of pancreas,IVC is Inferior vena cava, AO is Aorta,CBD is common bile duct & LRV is left renal vein.)

DPA is suspected, the combined use of CT and ERCP or MRCP is needed. However, ERCP is invasive procedure and operator-dependent for successful identification of opacity of the main and accessory pancreatic duct. By contrast, MR cholangiopancreatogram (MRCP) clearly demonstrates pancreatic duct morphology.[15-17] In cases where cannulating the pancreatic duct fails, MRCP may be helpful. Therefore, the combined use of CT and ERCP or MRCP is useful for confirmation of the diagnosis of agenesis of dorsal pancreatic bud.

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