PARTIAL AGENESIS OF THE DORSAL PANCREAS: CASE REPORT WITH IMAGING FINDINGS

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ABSTRACT

Partial or complete agenesis of the dorsal pancreas is a rare congenital anomaly that results from the embryological failure of the dorsal pancreatic bud to form the part of head, body and tail of pancreas. This condition is exceedingly rare; less than 100 cases have been reported in the literature since 1911.[5] We report an additional case; a 19-year-old woman presented with chronic epigastric pain. Abdominal computed tomography (CT) revealed a normal-appearing pancreatic head, but the neck, body and tail were not visualized. The patient was diagnosed with partial agenesis of the dorsal pancreas on CT followed by MRCP with strong clinical evidence of persistently raised serum amylase levels.

Key words: Pancreas agenesis, amylase, MRCP.

Introduction

Pancreatic agenesis is a congenital malformation in which either the entire dorsal pancreas or a part of it fails to develop (complete or partial agenesis, respectively). Agenesis of the dorsal pancreas is very rare and described in literature as case reports only. Complete pancreatic agenesis in which whole pancreas including dorsal and ventral parts are not developed is incompatible with life.[1] However patients of dorsal pancreatic agenesis are usually asymptomatic and may present with certain clinical conditions.[1] Partial agenesis of dorsal pancreas may be appre-ciated as a short, rounded pancreatic head adjacent to the duodenum with absence of the pancreatic neck, body, and tail. In complete agenesis of dorsal pancreas superior part of head is also not developed along with absence of neck, body and tail. The diagnosis of dorsal pancreatic agenesis is inconclusive without demonstration of the absence of the dorsal pancreatic duct. USG, computed tomography (CT), and magnetic resonance (MR) imaging are only suggestive; however, endoscopic ultrasound (EUS), endoscopic retrograde cholangiopancreatography (ERCP), and magnetic resonance cholangio pancreatography (MRCP) give detailed pancreatic ductal anatomy for diagnostic confirmation.

Case Report

We report a 19-year-old female patient who was requested to our department for CT scan abdomen with clinical complaints of having repeated epigastric pain.

Prior to presentation,the patient was diagnosed with cholelithiasis and chronic pancreatitis on ultrasound. Rest of the USG findings were reported to be normal, but images were not available. She underwent cholecystectomy, however, her symptoms persisted. Laboratory investigation revealed raised serum amylase level 2000 U/L (normal values 0-200 U/L) even after surgery. Physical examination revealed abdominal tenderness. Ultrasonography was done for variable times during her illness outside our hospital

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and revealed no specific findings relating to pancreatitis. But patient was suffering from periodic episodes of epigastric pain related to meal intake and persistently raised amylase levels. A contrast-enhanced abdominal (CT) examination revealed only partial visualization of the pancreas. The pancreatic head and uncinate process were normal, but the neck, body, and tail of pancreas were absent. There were no sign of peripancreatic inflammation,no fluid collection and neither the visualized pancreas appeared swollen to suggest any feature of pancreatitis in current study. The stomach and loops of jejunum could be seen in the distal pancreatic bed (dependent stomach/dependent intestine sign (Fig. 1 & 2), suggesting partial agenesis of dorsal pancreas (ADP)

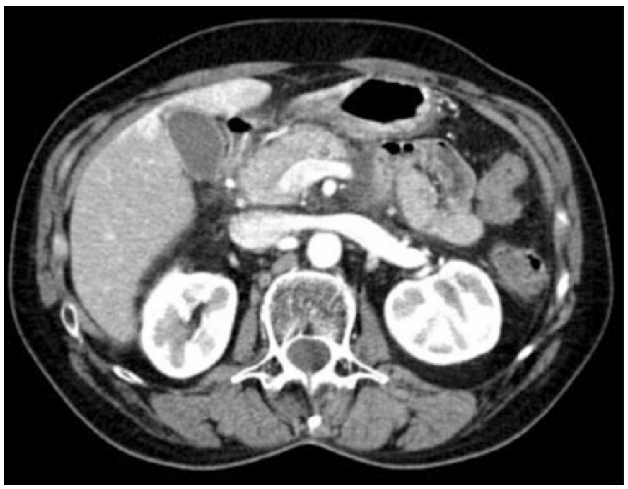

Figure 1: CT abdomen shows absence of pancreatic body and tail

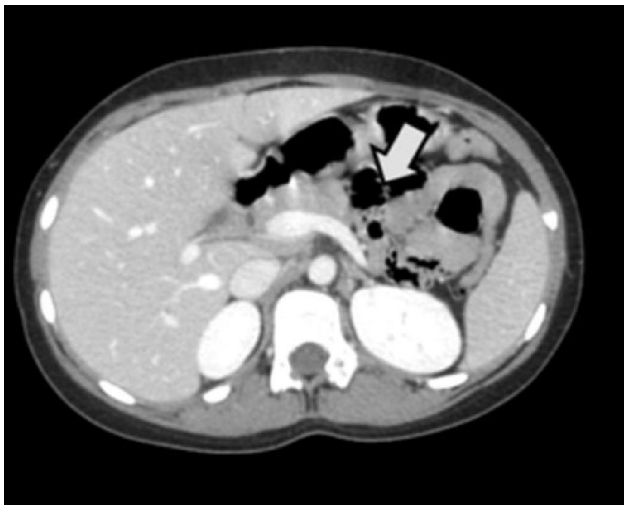

Figure 2: CT abdomen shows distal pancreatic bed is filled with small bowel which is abutting the splenic vein (dependent intestine sign)

as a possible diagnosis. To confirm the diagnosis, magnetic resonance cholangiopancreatography (MRCP) was performed. On MRCP, the dorsal pancreaticduct (duct of Santorini) could not be visualized. The common bile duct and ventral duct of Wirsung were normal and clearly seen (Fig. 3). These findings were compatible with partial dorsal pancreatic agenesis, so that there was no need for endoscopic retrograde cholangiopancreatography (ERCP).

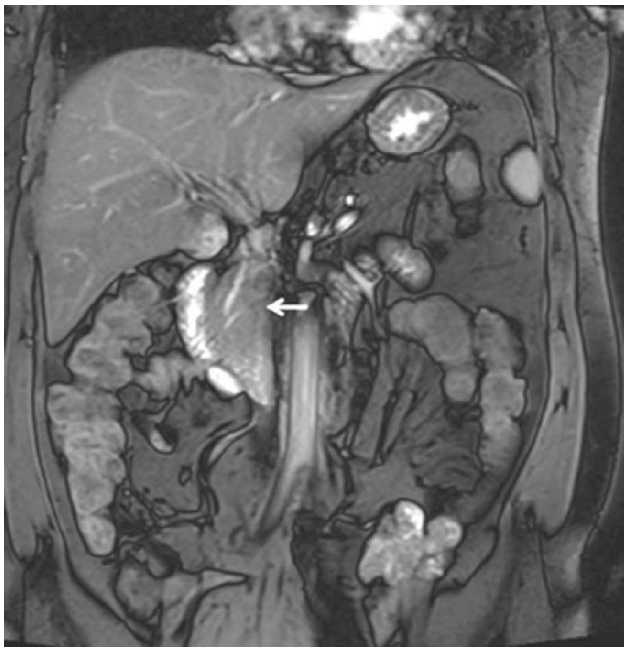

Figure 3: MRI showing partial visualization of the pancreas. Well-developed, normal head and uncinate process of pancreas are visible, but the neck, body, tail and dorsal pancreatic duct cannot be seen

Discussion

The pancreas develops from dorsal and ventral buds originating from the endodermal lining of the duodenum. During the seventh gestational week, the ventral bud turns posteriorly and to the left, connecting with the dorsal bud toform the mature gland.[2] The neck, body, tail, and superior part ofhead of the pancreas originate from the dorsal bud. This part is drained through duct of Santorini. At the 12th week of embryogenesis,discrete islets of Langerhans form primarily within the tail of the pancreas and the dorsal pancreas. The ventral bud becomes the inferior portion of the head and the uncinate process, which is drained through duct of Wirsung.[3,4] Abnormal embryogenesis can lead to developmental failure of the dorsal pan-

creas, resulting in agenesis of dorsal pancreas.This condition is exceedingly rare; less than 100 cases have been reported in the literature since 1911.[5] Agenesis of ventral pancreas and complete agenesis of the pancreas are incompatible with life.[5] The exact mechanism and aetiology of ADP is not known.

Most ADP patients are asymptomatic, but 92.9% of the symptomatic cases present with epigastric pain. About half of affected individuals develop diabetes mellitus, resulting from reduced islet cell mass secondary to the absence ofendocrine structures, which are normally located in the body and tail of the pancreas. Pancreatitis results from loss of function of sphincter of Oddi, hypertrophy of remnant ventral gland resulting in enzyme hypersecretion and higher intrapancreatic duct pressures.[6-7] There is a subclass which is defined as “anatomic” chronic pancreatitis, namely, a pancreatitis associated with obstructive pancreatitis, pancreas divisum, post-traumatic pancreatic duct scars and periampullary duodenal wall cysts.[8] Chronic pancreatitis due to dorsal pancreatic agenesis can also be defined as “anatomic” pancreatitis which probably causes an impairment of pancreatic secretion and higher intraductal pressures in the dense ventral part. Its diagnosis requires a high index of suspicion. Imaging appearances are not consistent with pancreatic inflammation but serum amylase remains high due to“anatomic” chronic pancreatitis. Our patient’s epigastric pain related to meals and persistently raised serum amylase level can be explained by ADP. Other abnormalities such as heterotaxy, polysplenia syndrome, ectopic spleen, bowel malrotation, coarctation of the aorta, tetralogy of Fallot, atrioventricular valvular abnormalities, and total anomalous pulmonary venous connection have also been reported to be associated with ADP.[6]

Other differential diagnosis of agenesis of the dorsal pancreas include: pseudoagenesis (atrophy of pancreas secondary to chronic pancreatitis); carcinoma of the head of pancreas (proximal atrophy of the gland); pancreas divisum (absence of fusion or incomplete fusion of the ventral and dorsal pancreatic ducts, pancreatic pseudolipodystrophy; pancreatic masses; and distal pancreatic lipomatosis (abundant fat tissue anterior to the splenic vein.[9,10] It is essential to differentiate these conditions from ADP through a careful medical history and appropriate imaging studies: ultrasonography, computed axial tomography (CT), magnetic resonance pancreatogram (MRI, including MRCP) or endoscopic retrograde cholangio pancreatography (ERCP), and a recent addition-endoscopic ultrasound (EUS) in order to exclude the above mentioned differential diagnosis. Previously, the diagnosis of ADP was only made at autopsy. ERCP is considered to be the gold standard for detailed description and evaluation of the biliary and pancreatic tree because of its superior spatial resolution however, the examination is invasive, operator-dependent, requires radiation exposure and morbidity risk (pancreatitis can result from catheterization of the minor duodenal papilla).

USG is the first modality, but sometimes pancreas is not adequately visualized due to obesity or excessive gases. CT depicts parenchymal abnormality very well. It shows the deficient part of the pancreas and dependent stomach or dependent intestine signs. But proper description of the pancreatic ductal anatomy is mandatory for diagnostic confirmation. There fore, evaluation with MRCP or ERCP is required.

The same findings can be seen in patients who have undergone a distal pancreatectomy, but in these patients the splenic vein is absent. In the case of distal pancreatic lipomatosis, abundant fat tissue is observed anterior to the splenic vein. Dependent stomach and/or dependent intestine signs on MDCT imaging confirm the diagnosis of ADP.[9] Magnetic resonance imaging, including MRCP, is the choice of investigation for further confirmation, as it is non-invasive and accurately tells the pancreatic duct morphology and parenchyma in the same examination. In a study reported by Kahl et al., endoscopic ultrasound (EUS) was described as a relatively new minimally invasive imaging technique which provides direct visualization of the entire pancreatic parenchyma and the pancreatic ductal system.[12] EUS also provides the opportunity for fine needle aspiration cytology (FNAC)and may be as good as ERCP.[10,12]

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