A Rare Case of Agenesis of Dorsal Pancreas

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ABSTRACT:

Agenesis of dorsal bud of the pancreas is an extremely rare congenital anomaly which results in absence of neck, body and tail of the adult pancreas. It may be associated with number of clinical features like diabetes mellitus, abdominal pain and chronic pancreatitis. Because of its rarity, we are reporting a case of agenesis of dorsal pancreas associated with early onset diabetes. Ultrasonography and Computed tomography showed absence of neck, body and tail of pancreas anterior to splenic vein and portal confluence; however head and uncinate process were normally present. Patient was thus diagnosed as agenesis of dorsal bud of pancreas.

Keywords: Dorsal pancreas, Agenesis, Congenital anomaly of pancreas.

INTRODUCTION:

Pancreas develops from dorsal and ventral pancreatic buds arising from caudal part of foregut. The dorsal bud elongates to form upper portion of head, body and tail of the pancreas; while ventral pancreatic bud gives rise to lower portion of the head and uncinate process1. 'Agenesis of dorsal pancreas' is an extremely rare anomaly5-6. Only 19 such cases have been reported so far in the literature until 2010. This case-report describes a patient with agenesis of dorsal pancreas associated with early-onset diabetes mellitus. It is reported here because of its rarity.

CASE SUMMARY:

A 34 year-old female patient presented with epigastric pain on and off for two months. When the history was probed, it was found that she had early-onset of diabetes mellitus for which she was on insulin therapy for the past five years. There was no previous history of abdominal surgery or past history of similar episodes of epigastric pain.

When ultrasonography was done by using Philips HD 11 machine using B-mode and colour Doppler, it was surprising to note that no pancreatic parenchyma could be observed anterior to splenic vein and portal confluence, instead of which only small bowel loops were found. This suggested absence of neck, body and tail of pancreas (Fig 1). However, head and uncinate process of pancreas could be observed. Superior mesenteric vessels were observed anterior to the uncinate process (Fig 2). There was no evidence of ectopic pancreatic tissue. Rest of the abdominal organs were found to be normal.

Patient was then subjected to Computed Tomography (CT scan) using Asteion single slice spiral scanner...
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Fig. 2 USG image in transverse plane just caudal to the previous image showing head of pancreas; uncinate process behind superior mesenteric artery (SMA) and superior mesenteric vein (SMV). Small bowel loops occupying the empty space which is supposed to be filled with body of pancreas.

Fig. 3 Contrast Enhanced Computed Tomography (CECT) axial section shows the head of pancreas in the normal location, but there is absence of neck, body and tail of pancreas. The space is occupied only by the contrast filled stomach. Portal vein confluence (PV-C) and Aorta (Ao) are shown.

Fig. 4 Contrast Enhanced Computed Tomography (CECT) showing axial section just caudal to the previous section. Arrow-heads show absence of neck, body and tail of pancreas, anterior to the course of splenic vein (Spl V). The space is occupied only by the contrast filled stomach. However, head and uncinate process are seen normally.

(Toshiba Medical System). Intravenous and oral contrast agents were given and axial images were taken. In contrast enhanced CT sections, head and uncinate process were observed normally (Fig 3); but neck, body and tail of pancreas were not observed. The space anterior to the course of splenic vein was just occupied by the contrast filled stomach (Fig 4). Thus the patient was diagnosed as a case of 'Agenesis of dorsal pancreas'.

DISCUSSION:

Pancreas develops from endoderm of duodenal segment of foregut. At fourth week of gestation, dorsal pancreatic bud develops cephalic to the hepatic bud. Later, ventral pancreatic bud develops distal to dorsal bud. Due to rotation of duodenum, the ventral pancreatic bud is carried dorsally and fuses with the dorsal pancreatic bud at seventh week of gestation. The dorsal bud elongates to form the upper portion of head, body and tail, while ventral pancreatic bud gives rise to the lower portion of the head and uncinate process. The main pancreatic duct of Wirsung is formed by whole of ventral pancreatic duct and distal portion of dorsal pancreatic duct and drains into major duodenal papilla. The accessory duct of Santorini is formed by proximal portion of dorsal pancreatic duct and drains into minor duodenal papilla.

Among the congenital anomalies of pancreas, agenesis of dorsal pancreas is an extremely rare entity. Agenesis of dorsal pancreas may be complete or partial. In complete agenesis of dorsal pancreas, neck, body, tail, duct of Santorini and minor duodenal papilla are absent. However, in partial agenesis of dorsal pancreas, body, duct of Santorini and minor duodenal papillae are present.

Wilding et al reported a case with familial occurrence of agenesis of dorsal pancreas in both the mother and her son. This suggested the possibility of genetic mode of transmission through autosomal dominant or X-linked dominant pattern.
Agenesis of dorsal pancreas may be associated with diabetes mellitus, abdominal pain and chronic pancreatitis. Most of the patients had early-onset of diabetes mellitus, as reported in the present case, because major amount of Islet of Langerhans are lodged in tail and body of pancreas\(^5\). Pathogenesis of pancreatitis in these patients may be due to the sphincter of Oddi dysfunction\(^9\) and due to the hypertrophy of remnant ventral pancreatic bud leading to increase in the intra-pancreatic duct pressure\(^10\). The probable reason for abdominal pain may be due to insufficient drainage by the ill-defined duct system\(^6\) and due to under-development of duodenal papillary muscle\(^5\) or due to duodenal obstruction and autonomic neuropathy\(^5\). Other clinical features associated with this anomaly were malrotation of gut\(^1\), horse shoe kidney\(^1\), polyspleenia and coarctation of aorta\(^11\).

Conditions which may mimic agenesis of dorsal pancreas are pancreas divisum, congenital short pancreas, pancreatic pseudolipodystrophy, obstructing pancreatic tumors\(^2\), fatty infiltration of pancreas, pseudogenes or auto-digestion of pancreas due to chronic pancreatitis\(^5\). The fatty replacement of pancreas can be differentiated from agenesis. In fatty infiltration, entire gland is replaced by the by fat, but the pancreatic duct is spared and is normally present\(^2\). In Pseudoagenesis of dorsal pancreas, only the parenchyma is auto-digested and duct system is spared. However in true agenesis, both parenchyma and duct system of dorsal pancreas are absent\(^1\).

Before the use of modern imaging, the diagnosis was made only by autopsy or at surgery. Nowadays, Ultrasonography (USG), Computed Tomography (CT), Endoscopic Retrograde Cholangio-Pancreatography (ERCP), Magnetic Resonance Cholangio-pancreatography (MRCP) and Endoscopic Ultrasonography (EUS) help us to diagnose agenesis of dorsal pancreas. EUS is much more sensitive than other modalities\(^2\). The absence of body and tail are best demonstrated by CT and MRI. ERCP and MRCP help to rule out auto-digestion of pancreas and fatty infiltration by identifying the duct system\(^5\). ERCP is invasive and cannulation of minor papilla is a challenge in some cases. It also has radiation risks. However, MRCP is non-invasive and there is no risk of radiation\(^6\).

To summarize, 'Agenesis of dorsal pancreas' is a very rare congenital anomaly of pancreas. It may be associated with various clinical features. Due to advances in modern imaging techniques, it can be accurately diagnosed.

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