

Dorsal pancreas agenesis - a rare case report

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SUMMARY

Dorsal pancreatic agenesis is a rare pancreatic anomaly. We report a 47-year-old female patient who attended the medicine outdoor with complaint of intermittent upper abdominal pain for 6 months of duration. She was not a known diabetic or hypertensive. Mild splenomegaly was detected on abdominal examination. Ultrasonography of abdomen showed cholelithiasis, splenomegaly with dilated portal veins and multiple porto-systemic collateral. The pancreas was not visualized due to bowel gas shadow. Contrast-enhanced computed tomography of the abdomen revealed absent pancreatic body and tail with stomach/small bowels occupying the pancreatic bed anterior to splenic vein along with features of portal hypertension, cholelithiasis, malrotated left kidney and left extrarenal pelvis. Magnetic resonance cholangiopancreatography confirmed nonvisualization of the pancreatic duct except a small remnant of the ventral duct of the pancreas and an absent dorsal part of pancreas. Our case report is a rare combination of dorsal pancreatic bud agenesis with malrotated kidney and extrarenal pelvis.

Key words: Agnesis – Dorsal bud – Pancreas – Anomaly

INTRODUCTION

Agnesis of dorsal pancreas (ADP) is a rare congenital anomaly in comparison with other congenital pancreatic anomalies such as pancreatic divi-

sum, annular pancreas, ectopic pancreas and common bile duct syndrome. It is represented in the adult anatomy by the absence of the body and tail of the pancreas. In the last 20 years, with the improvement of the imaging diagnosis techniques, ADP has been reported in many cases associated to different pathologies and anomalies. However, it appears only superficially mentioned in the classical textbooks of embryology, but not in anatomy handbooks. In literature reviews of the last five years in PUBMED, only 18 cases have been reported. We have not found a case of ADP with malrotation of kidney with extrarenal pelvis. It occurs due to partial or non-development of dorsal pancreatic bud (Boopathy et al., 2013). Most commonly, it presents with nonspecific pain abdomen, symptoms of pancreatitis or diabetes mellitus. We report a complete agnesis of dorsal pancreas with malrotated left kidney, left extrarenal pelvis in a 47-year-old woman with pain abdomen as a rare anatomical anomaly.

CASE REPORT

A 47-year-old female patient reported to medicine outdoor with complaint of intermittent dull aching abdominal pain for 6 months of duration. The pain was non-radiating in nature and not associated with fever or vomiting. No relevant clinical history was present for pain abdomen. On clinical examination, her vitals were within normal limit. Chest and cardiovascular examinations are unremarkable. Mild splenomegaly was detected on per abdominal examination. Ultrasound evaluation of the abdomen was advised, which revealed moderate splenomegaly, cholelithiasis, dilated portal vein with multiple porto-systemic collaterals at lower gastro-esophageal junction (suggestive of portal hypertension). The body and tail of pancreas were not visualized due to excessive bowel gas shadow.

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Complete hemogram of the patient showed normal blood sugar level and normal renal function. Serum amylase and lipase level were normal. Liver function test showed mild elevated serum transaminase enzyme levels and normal serum alkaline phosphatase level. Contrast-enhanced computed tomography (CT) examination of the abdomen revealed a short pancreas with absence of the body and tail of pancreas, with dependent bowels in the pancreatic bed anterior to the splenic vein. Pancreatic head and uncinete process appeared bulky in size (4.4 cm in longest dimension). Moderate splenomegaly, dilated splenic vein, splenic hilar collaterals, dilated portal vein and a large gall bladder calculus were noted (Fig. 1b). The left kidney was malrotated, with evidence of extrarenal pelvis. Magnetic resonance imaging (MRI) of the abdomen revealed absent body and tail of the pancreas with dependent bowels in the pancreatic bed and bulky pancreatic head and uncinete process. Magnetic resonance cholangiopancreatography (MRCP) revealed absent pancreatic duct except a very small pancreatic duct near the ampulla. No evidence of pancreatitis was seen (Fig. 1a-e). The radiological diagnosis of CT and MRI study of the abdomen was agenesis of dorsal pancreas with cholelithiasis, left malrotated kidney with extrarenal pelvis and evidence of portal hypertension. Upper gastrointestinal endoscopy revealed grade II esophageal varices with absent minor papilla. Prophylactic variceal banding was done. She was advised for liver function test, liver biopsy and laparoscopic cholecystectomy in later date.

DISCUSSION

The pancreas develops from ventral and dorsal endodermal outpouching of the duodenum, and develops into ventral and dorsal pancreas respectively. The dorsal part grows rapidly in comparison with the ventral part, and forms an elongated nodular structure within dorsal mesentery by the 6th week of intrauterine life. The smaller ventral part is carried away from the duodenum due to its attachment with bile duct. The ventral and the dorsal parts of the pancreas are opposite to each other by the 7th week. The dorsal pancreas forms the tail, the body and part of the head of the pancreas, and drains to the duodenum at minor papilla through the accessory duct. The ventral part forms the uncinete process and the remainder of the pancreatic head, and drains at major papilla through the main pancreatic duct (Arey et al., 1974).

Aberrations in the complex development of the pancreas can occur, though rarely. The etiology of dorsal pancreatic agenesis is unknown. Primary dysgenesis or ischemic events in the developing pancreas are possible explanations (Macari et al., 1998). Agenesis of the dorsal pancreas (ADP) can be partial or complete, depending on partial or non

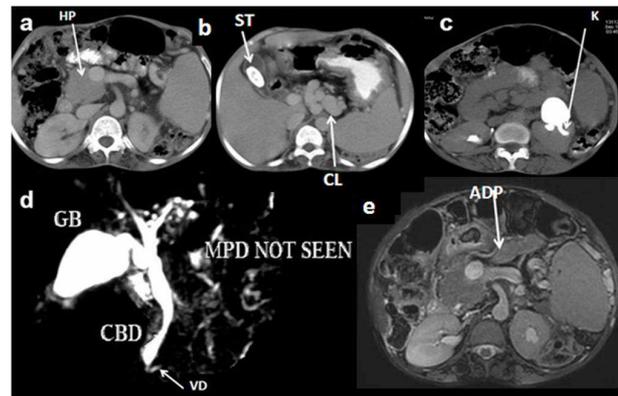


Fig. 1. (a) CT scan of abdomen shows presence of pancreatic head (HP) with absent pancreatic body and tail; (b) splenomegaly, porto-systemic collaterals (CL) and cholelithiasis (ST); (c) malrotation of left kidney with extrarenal pelvis (K). MRI of abdomen (d) shows absent main pancreatic duct in pancreas body and tail with remnant small ventral pancreatic duct (VD) in MRCP, and (e) absence of pancreatic body and tail with dependent small bowels in pancreatic bed anterior to splenic vein (ADP).

-development of the dorsal pancreatic endodermal bud. In complete dorsal pancreatic agenesis, the body and tail of the pancreas, the duct of Santorini and the minor papilla are absent, while in partial agenesis the minor papilla, the remnant of the accessory duct, the neck and a small portion of the body of the pancreas may be seen (Ulusan et al., 2005). Complete agenesis of pancreas or ventral pancreas is incompatible with life (Mohapatra et al., 2012). Though ADP is usually sporadic, it can occur as an autosomal dominant or X-linked dominant inheritance or with polysplenia/heterotaxy syndrome (Macari et al., 1998). Hepatocyte nuclear factor 1B (*HNF1B*) gene mutation may be responsible for agenesis of the pancreatic body and tail (Haldorsen et al., 2008).

Malrotation of the kidney occurs due to abnormal rotation of developing metanephrons relative to collecting systems, and are of four types. In non-rotation, the collecting systems are seen arising anteriorly. In incomplete rotation, the collecting systems are arising from the ventromedial aspect of the kidney. In rare reverse and excess rotation, the position of renal pelvis depends on the degree of rotation. The embryological basis of malrotation is still speculative. It is often associated with or caused by aberrant renal vessels (Ingole et al., 2005). In our case, ADP was associated with non-rotation of the left kidney.

ADP is often associated with other congenital anomalies such as ectopic spleen, polysplenia, interruption of the inferior vena cava, azygos or hemiazygos continuation, left-sided vena cava, symmetrical liver, anomalous liver fissure, anomalous liver lobe, median location of the gallbladder, inverted stomach, gallbladder and pancreas, intestinal malrotation, congenital heart disease or a combination of multiple visceral anomalies

(Hidehiro et al., 2003). Horse-shoe kidney may be associated with ADP (Michael et al., 2010). In our case, left renal malrotation and left extrarenal pelvis were seen. Literature review showed no case report of ADP, malrotated kidney with portal hypertension. The association of ADP with portal hypertension could be incidental, and further study is required for establishment of a causal relation between the two if at all exist.

CT study of the abdomen and MRCP of the abdomen are preferred as noninvasive diagnostic modality for evaluation. The diagnosis of dorsal pancreatic agenesis is inconclusive unless absence of duct is demonstrated by endoscopic retrograde pancreatography (ERCP) or MRCP (Pasaoglu et al., 2008). Absence of the body and tail of the pancreas with stomach and/or bowels occupying the pancreatic bed anterior to the splenic vein is described as dependant stomach and dependant intestine sign in CT study of abdomen (Mohapatra et al., 2012). Ultrasound of abdomen may not show pancreas body and tail in case of gas-filled intestine and stomach and in obese patients. MRCP is the choice method of investigation for detection of ADP, and is preferred over ERCP, as it is noninvasive in nature and has no risk of radiation (Uygur-Bayramicli et al., 2007). Schnedl et al. (2009) describes the presence of compensatory pancreatic head hypertrophy in some of the cases of ADP as well as normal to atrophic pancreatic head in the rest of cases. In our case, the pancreatic head was bulky in size.

Congenital short pancreas, pseudo-agenesis and pancreatic lipomatosis are the common differential for ADP. In pancreatic lipomatosis and pseudo-agenesis, the pancreatic duct is usually seen in MRCP or ERCP. In pancreatic lipomatosis, dependant stomach or dependant intestine sign will not be seen (Mohapatra et al., 2012).

There was a certain limitation to our study, as the diagnosis of dorsal pancreatic bud agenesis was based on radiological imaging only, and there was no ERCP correlation or no anatomical/ histological confirmation of the rare anomaly.

So ADP is a rare anatomical anomaly and often associated with other anomalies. It is increasingly detected by use of advanced imaging modality such as MRCP, multidetector CT and ERCP. The patient with ADP is treated according to symptomatology.

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