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Case Report

Agenesis of dorsal pancreas causing extra hepatic portal vein obstruction in a patient of symptomatic cholelithiasis: a case report

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ABSTRACT

Agenesis of dorsal pancreas (ADP) is an extremely rare entity. We report a case of 50-year-old lady with complete agenesis of the dorsal pancreas presenting with extra-hepatic portal vein obstruction (EHPVO) with symptomatic gall stone disease. Contrast enhanced computed tomography (CECT) revealed a normal pancreatic head, but pancreatic body and tail were not visualized, pancreatic head was compressing upon the portal vein with marked luminal narrowing and cavernoma formation. Magnetic resonance imaging (MRI) and magnetic resonance cholangio pancreatography (MRCP) findings confirmed the CT findings of ADP.

Keywords: ADP, Agenesis of dorsal pancreas, EHPVO

INTRODUCTION

Developmental anomalies of the pancreas have been reported but dorsal pancreatic agenesis is an extremely rare entity. In 1911, the first description of agenesis of the dorsal pancreas (ADP) was published as an autopsy finding. ADP is mostly asymptomatic but abdominal pain, pancreatitis and diabetes mellitus may be associated.[1]

Wolfgang J. Schnedl et al, revealed just 53 reported cases of agenesis of the dorsal pancreas with association of different diseases and anomalies but none has reported its association with EHPVO.[2] Authors report a case of ADP causing EHPVO in a case of symptomatic cholelithiasis.

CASE REPORT

50 years old female, a case of symptomatic cholelithiasis with no other co-morbidities, was referred to us from a peripheral hospital after failed laparoscopic cholecystectomy. The procedure was abandoned because of cavernoma formation around the porta-hepatis. There was no history of pancreatitis, jaundice, upper gastrointestinal bleed or malaena.

Her general physical and systemic examination was essentially normal.

Her routine investigations were normal except for HbA1c in pre-diabetic range. Ultrasound abdomen coupled with Doppler study showed cholelithiasis and non-visualized portal vein with multiple tortuous veins at porta hepatis.

On CECT of abdomen, there was complete agenesis of dorsal pancreas with non-visualized body and tail portion (Figure 1A and Figure 1B).

The tissue present in the head region was compressing the portal vein with marked luminal narrowing with cavernoma formation. Upper gastro-intestinal endoscopy was normal with no features of portal hypertension.

Patient was taken up for open cholecystectomy under general anesthesia. Intra operatively - liver and spleen were normal, no ascites was present and there were multiple dilated, tortuous veins of 4-5 mm diameter at porta hepatis and calot's triangle (Figure 1C and Figure 1D).

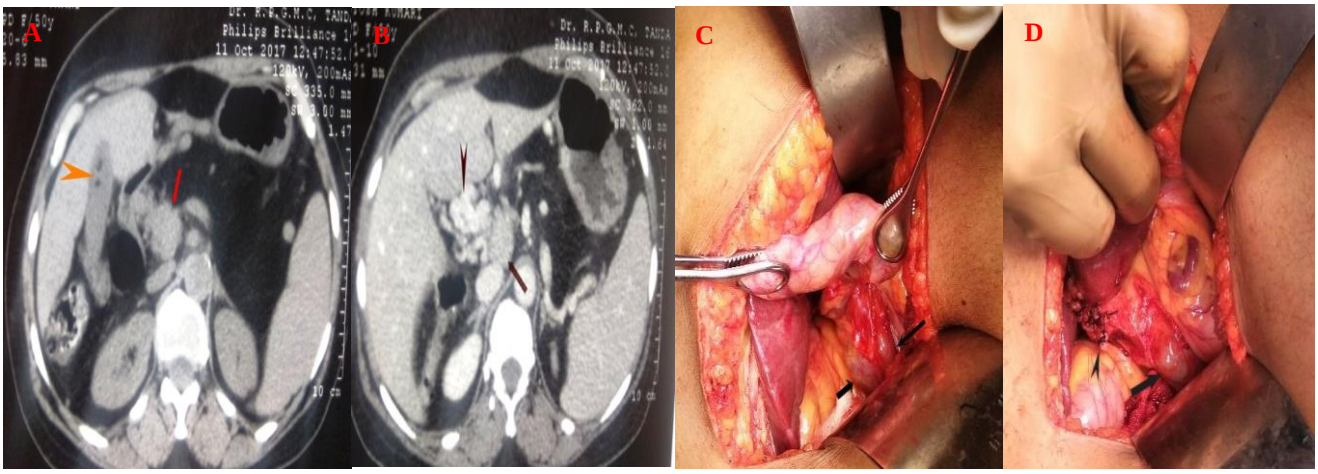

Figure 1: A) CECT abdomen and Intra - operative pictures: arrow head highlighting gallstone and arrow pointing to the pancreatic head. B) arrow head pointing cavernoma formation at porta hepatis, and arrow highlighting the absence of body and tail of pancreas. C and D) Intra-operative- there were multiple dilated, tortuous veins of 4-5 mm diameter, traversing over Calot's triangle and porta hepatis (Arrows – cavernoma, Arrow head – cystic duct and artery stump).

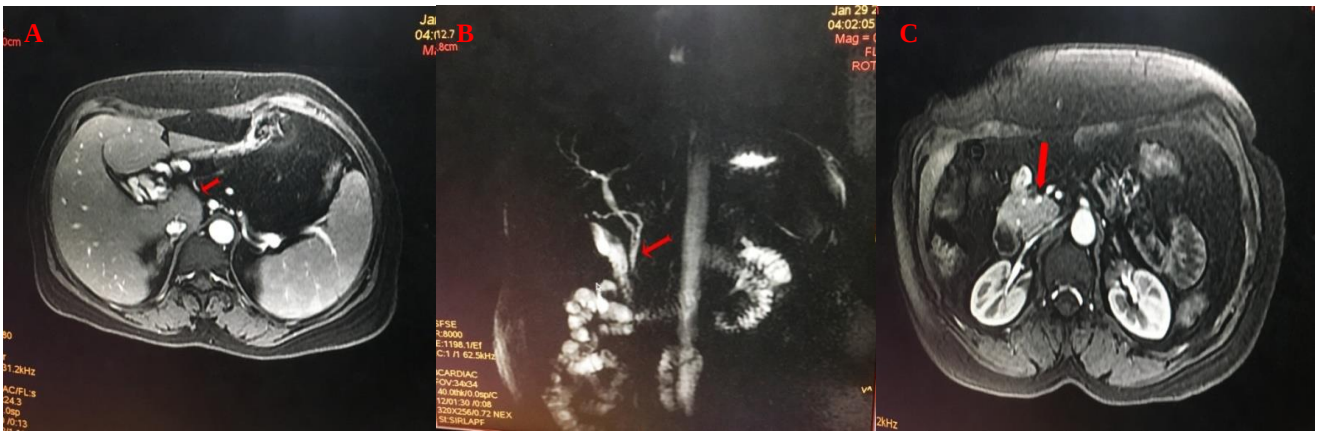

Figure 2: A) CEMRI and MRCP images showing absence of body; B) tail of pancreas and low insertion of cystic duct with absence of dorsal duct. C) Only head and uncinate part of pancreas is visible.

No other Porto-systemic collaterals were present. Cholecystectomy was done and cavernoma was not disturbed. Contrast enhanced MRI and MRCP, which was done in post-operative period to confirm the diagnosis of ADP, showed the pancreatic head ventral to the splenic vein.

The dorsal pancreatic duct, pancreatic body and tail were absent. Multiple collateral vessels were present at porta hepatis, suggestive of EHPVO (Figure 2). Postoperative recovery was uneventful, and patient is on regular follow up.

DISCUSSION

The pancreas develops 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 Santorini duct.

The ventral bud gives rise to the major part of the head and uncinate process and drains through Wirsung duct.[3] Agenesis of the ventral pancreas and complete agenesis of the pancreas are lethal conditions.[4] Dorsal pancreatic agenesis is an extremely rare congenital anomaly.

Although the size of the pancreas can be determined with CT, a diagnosis of the ADP is inconclusive without demonstration of the absence of the dorsal pancreatic duct, either with endoscopic retrograde cholangiopancreatography or MRCP.[5] In some patients, an enlarged or prominent or compensatory hypertrophy of the pancreatic head has been described, whereas other descriptions include normal sized pancreatic head, as well as mildly atrophic and small head of the pancreas.[2] In the absence of forthcoming history of acute pancreatitis, the compensatory hypertrophy of pancreatic head could be the reason for EHPVO in this case. Around 100 cases of ADP have been reported in the literature so far but none in association with EHPVO. We report the case of ADP presenting with EHPVO where the pancreatic head itself is compressing the portal vein, leading to marked luminal narrowing with cavernoma formation.

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