It is short-but so what!

Sir,

A 34-year-old male presented with gradually worsening right upper-quadrant pain. There was no history of any significant past illness or abdominal surgery. His laboratory test results revealed hemoglobin 12.9 g/dl; total leukocyte count 13 200 cells/mm^3 (neutrophils-63%); normal liver and kidney function tests. Ultrasound of the abdomen revealed features suggesting acute-on-chronic cholecystitis following which the patient was subjected to contrast-enhanced CT. Apart from calculus cholecystitis, an important incidental finding was detected. CT sections through the pancreas revealed normal size head, neck and the uncinate process of the pancreas with absent pancreatic body and tail. The distal pancreatic bed was seen filled by stomach and intestine [Figure 1]. The patient was subsequently evaluated and was found to have mildly elevated serum glucose (fasting blood glucose – 105 mg/ dl).

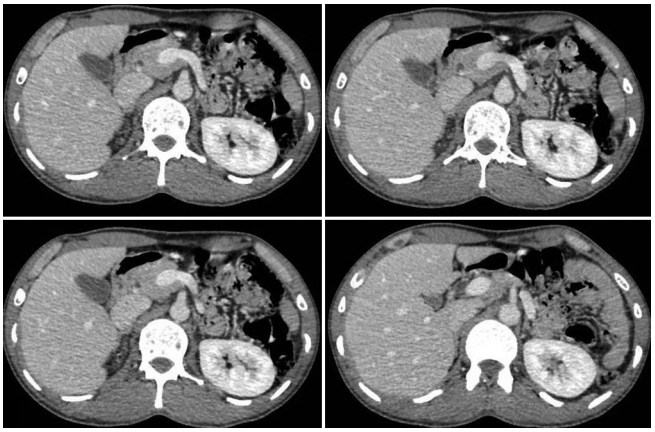

Figure 1: CT sections through the abdomen reveal normal size head, neck and the uncinate process of the pancreas with absent pancreatic body and tail

Agenesis of the dorsal pancreas is a rare congenital anomaly that represents embryological failure of the dorsal pancreatic bud to form the body and tail of the pancreas.[1-3] Till date, less than 100 cases have been described in the world literature.[2] The embryogenesis of pancreas is complex as the gland develops from the endoderm-lined dorsal and ventral buds of the duodenum. The ventral bud rotates posteriorly during the 7th week of gestation to fuse with the dorsal bud and form the fully grown gland. The ventral bud forms the major portion of the head (lower portion) and uncinate process, while the dorsal bud develops into the upper part of the head, the body, and the tail of the pancreas. The developmental failure or regression of the dorsal pancreatic bud is considered responsible for the dorsal agenesis. [1-3] Complete dorsal agenesis is extremely rare and is characterized by complete absence of the neck, body, and the tail along with missing accessory duct of Santorini and minor papilla. In contrast, in partial agenesis, the pancreatic body is of variable size, a remnant of the accessory duct exists and the minor papilla is present. Generally, the patients remain asymptomatic but some of them manifest abdominal pain, pancreatitis, or diabetes mellitus. The cause of pancreatitis is contentious; however, dysfunction of the sphincter of Oddi has been implicated.[1] As many as 50% of the affected individuals have hyperglycemia which has been attributed to the bulk of the insulin-producing beta cells being located in the dorsal part of the gland.[1] The diagnosis is usually suggested on cross-sectional imaging and it manifests as a short truncated pancreas with absent pancreatic parenchyma ventral to the splenic vein.[2,3] It is crucial to distinguish agenesis of the pancreas from atrophy and lipomatous replacement of the pancreas secondary to chronic pancreatitis. Dependent stomach and/or dependent intestine signs on cross- sectional imaging allow differentiation of the two.[3] The dependent stomach or dependent intestine sign refers to the distal pancreatic bed getting filled by stomach or intestines which abut the splenic vein [Figure 2a], while in case of distal lipomatosis, abundant fat tissue is observed anterior to the splenic vein and hence the bowel fails to abut the splenic vein [Figure 2b].

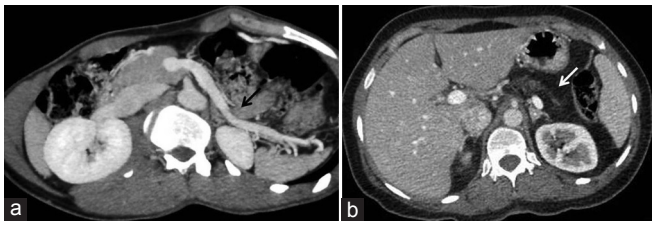

Figure 2: (a) Reformatted axial-oblique CT image of the same patient reveals that the stomach and bowel loops are occupying the pancreatic bed anterior to the splenic vein (black arrow) described as dependent stomach and dependent intestine signs. (b) Axial CT section in a different patient with chronic pancreatitis exhibits lipomatous replacement of pancreas (white arrow) with absent dependent stomach and dependent intestine signs

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