

Case Report



CT Diagnosis of Dorsal Pancreas Agenesis

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ABSTRACT

Agnesis of the dorsal pancreas is a rare congenital anomaly characterized by the absence of body and tail of pancreas. We report a case which presented with epigastric pain 52-year old woman with complete agnesis of the dorsal pancreas. Multidetector CT (MDCT) revealed a normal pancreatic head, but pancreatic body and tail were not visualized. The presence of small (jejunal loops) intestine and stomach in the distal pancreas bed, adjacent to the splenic vein. The final diagnosis was dorsal pancreatic agnesis. Dependent stomach and dependent intestine signs on MDCT imaging can be diagnostic obviating further radiologic examinations. In contrast to the reports of radiologic literatures, both dependent stomach and dependent intestine sign are noted in our patient.

Key words: *Agnesis, pancreas, multidetector computed tomography*

ÖZET

Dorsal Pankreas Agenezisinin BT Tanısı

Dorsal pankreas agenezisi pankreasın korpus ve kuyruğunun yokluğu ile karakterize nadir görülen pankreasın konjenital anomalisidir. Bu çalışmada epigastrik ağrı ile başvuran komplet dorsal pankreas agenezisi saptanan 52 yaşında kadın olgu sunuldu. Çok kesitli bilgisayarlı tomografi (ÇKBT) ile pankreasın başı normal şekilde izlenirken, pankreasın korpus ve kuyruk kesimi gözlenmedi. Splenik venin anteriorundaki distal pankreas yatağını incebarsak (jejunal segmentler) ve midenin doldurduğu izlendi, dorsal pankreas agenezisi tanısı konuldu. Dependan barsak ve dependan mide işareti olarak tanımlanan bu bulgular ile ÇKBT de dorsal pankreas agenezisi tanısı konulabilir ve ileri radyolojik incelemeler önlenebilir. Bizim olgumuzda literatürden farklı olarak hem dependan mide hem de dependan barsak işareti bulunmaktadır.

Anahtar Sözcükler: *Agnezi, pankreas, çok kesitli bilgisayarlı tomografi*

During the complex embryologic development of the pancreas, several morphologic anomalies can develop. One of these anomalies is complete agnesis of the dorsal pancreas which is a rare entity (1). Agnesis of the dorsal pancreas results in characteristic Multidetector CT (MDCT) findings. Two useful signs (distal pancreas agnesis and dependent stomach sign, distal pancreas agnesis and dependent intestine sign) have been described for differentiation of the distal pancreatic agnesis from fat replacement of the pancreas (2). In case of distal lipomatosis abundant fat tissue is observed anterior to splenic vein. In the absence of distal pancreas, distal pancreatic bed can be filled by stomach or intestine, which abut splenic vein (2). In our patient both dependent stomach sign and dependent intestine sign were identified on MDCT.

CASE REPORT

A 52-year-old woman patient presented with epigastric pain of a two week duration. Physical examination was unremarkable. Patient denied any previous surgery, major trauma or car accident. No history of diabetes or prior episodes of pancreatitis were reported. The biochemical evaluation of the patient revealed mild elevation of alkaline phosphatase (134 IU/L; reference range: 25-100 IU/L). Pancreatic amylase and

lipase levels in serum were within normal limits.

A contrast-enhanced abdominal CT examination was performed using a four-channel multidetector computed tomography (MDCT) scanner. The scanning parameters were contiguous 2.5 mm collimation with a pitch value of 1.25, with 5-mm-thick slices reconstructed secondarily at 1-mm intervals. The images were analyzed on a workstation using post-processing (three-dimensional reconstructions). At this examination pancreas body and tail were not seen. The presence of stomach and small bowel (jejunal) loops in the distal pancreas bed, adjacent to the splenic vein ('dependent stomach sign' and 'dependent intestine sign' respectively) were observed. Pancreatic head was normal in size and shape (Figure 1).

DISCUSSION

The pancreas grows dorsal and ventral buds originating from the endodermal lining of the duodenum. During the seventh gestational week, the ventral buds turn posteriorly and to the left, connecting with the dorsal bud. Each of the pancreatic buds grows into a pair of branching arborized ductal systems. The neck, body, tail, and cephalic aspects of the head of the pancreas originate from the dorsal bud. The ventral bud becomes the inferior portion of the head and the uncinate process (3, 4).

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Agenesis of the entire pancreas is incompatible with life (4). Dorsal pancreatic agenesis is a rare congenital anomaly. In the literature, 54 cases of partial agenesis of dorsal pancreas were reported (5). Now, as new imaging technologies have been developed and improved, the number of patients reported to show agenesis of the dorsal pancreas has increased rapidly over the last years. Patients with agenesis of dorsal pancreas often present with non-specific abdominal pain, which may or may not be caused by pancreatitis. In approximately 50% of reported patients with this congenital malformation, hyperglycemia was demonstrated (5).

Fat replacement (also termed lipomatosis, adipose atrophy, or fat infiltration) of the pancreas must be considered in the differential diagnosis of the dorsal pancreatic agenesis. In general, the differentiation of dorsal pancreatic agenesis and fat replacement of the distal pancreas depends on the presence of the dorsal pancreatic duct (1-4). In patients with dorsal pancreatic agenesis, the ductal structures and endocrine structures are absent. In general, these structures are thought to be preserved in patients with distal pancreas lipomatosis (2). However, Park et al. (6) recently reported a case having distal fat replacement with absent ductal and acinar cells. Therefore ERCP or MRCP is not necessary for revealing the major and the accessory duct systems.

Distal pancreas lipomatosis occurs as a result of distal pancreas atrophy. Abundant fat tissue anterior to splenic vein in patients with distal pancreatic lipomatosis. In distal pancreas agenesis fat tissue anterior to the splenic vein is absent or not abundant and therefore this potential space can be filled with either stomach or intestine and possibly with combination of both (2). Dependent stomach and/or dependent intestine signs on MDCT imaging can allow differentiation of distal pancreas agenesis from distal lipomatosis (2).

Now, as new imaging technologies have been developed and improved, the number of patients reported to show agenesis of the dorsal pancreas has increased rapidly over the last years. Dependent stomach and/or dependent intestine signs on MDCT imaging can be diagnostic obviating further radiologic examinations.

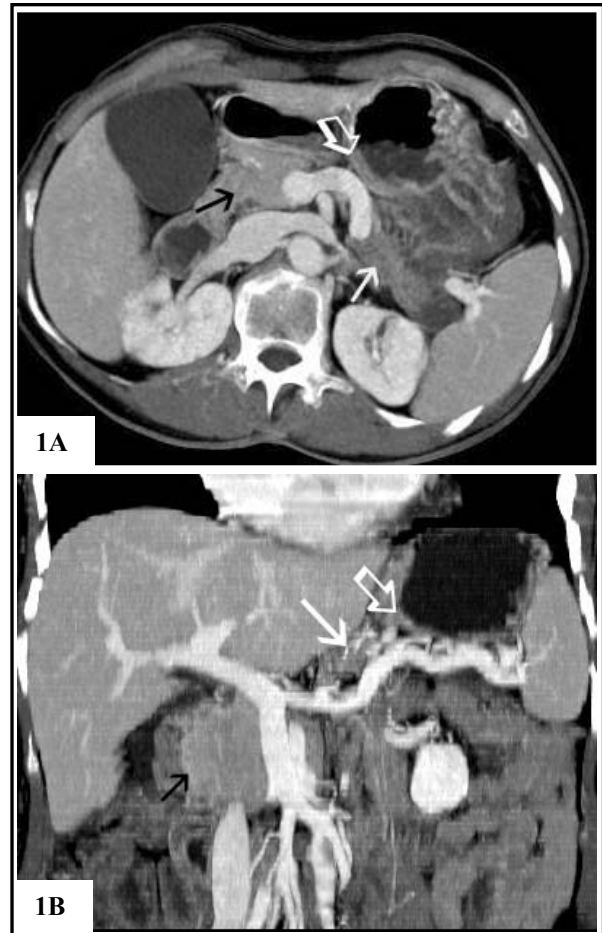


Figure 1. Axial MIP image (a) and Coronal MPR image (b) show pancreatic head (black arrow) and the absence of distal pancreas anterior to splenic vein. Note this potential space is occupied by stomach (dependent stomach sign) and jejunal segments (dependent intestine sign).

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