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Case Report RADIOLOGY

Complete agenesis of dorsal pancreas with pancreatic cyst: A case report

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ABSTRACT

Agenesis of the dorsal pancreas (ADP) is extremely rare disease with no specific symptoms and there is no clear pathogenesis. Approximately half of the affected individuals develop diabetes resulting from reduced islet cell mass secondary to lack of endocrine structures. In this case, we aimed to present a 17-year-old female patient with ADP accompanied by a pancreatic cyst.

Keywords: Agenesis of dorsal pancreas; magnetic resonance imaging; pancreatic cyst.

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Complete or ventral pancreatic agenesis is a condition that does not incompatible with life [1]. Agenesis of the dorsal pancreas (ADP) is also very rare and described in literature as case reports only. Herein, we present a 17-year-old female patient with ADP accompanied by a pancreatic cyst.

CASE REPORT

A 17-year-old female patient with resistant hypomagnesemia and insulin-dependent diabetes mellitus presented with intermittent epigastric pain for several months. The pain was not associated with fever or vomiting. Her laboratory tests revealed: fasting plasma glucose: 411 mg/dl, glycated hemoglobin/HbA1c: 7.8%, magnesium level: 1.02 mg/dl, and cholesterol: 222 mg/dl. Amylase and lipase levels were normal.

The patient was referred to the radiology department for abdominal ultrasound (US) examination. US revealed a cystic lesion on the head of pancreas (Fig. 1), whereas body and tail of pancreas could not be visualized due to bowel gas shadow. A contrast-enhanced abdominal computed tomography (CT) was performed for better evaluation of pancreas and cystic lesion. CT examination demonstrated the absence of neck, body, and tail of the pancreas (Fig. 2). Only the head and uncinate segment of the pancreas was visualized and the hypodense unilocular cystic lesion was revealed at the head of pancreas (Fig. 3). Furthermore, an accessory spleen was spotted. Abdominal magnetic resonance imaging (MRI) was ordered to evaluate the nature of the cyst. Contrast (gadolinium diethylenetriaminepentaacetic acid) enhanced abdominal MRI with routine sequences revealed no contrast enhancement on the cyst and its wall. The cyst was hypointense in T1-weighted imaging and hyperintense in T2-weighted imaging (Fig. 4). These findings were compatible with dorsal pancreatic agenesis with pancreatic cyst.

DISCUSSION

Dorsal pancreatic agenesis is a rare congenital anomaly associated with abnormal embryogenesis [2]. The pancreas develops from ventral and dorsal endodermal outpouching of the duodenum. At the 6–7th week of gestation, the ventral and the dorsal parts of the pancreas fuses and then both of these buds form the main pancreatic duct. During the 7th week of gestation, the ven-

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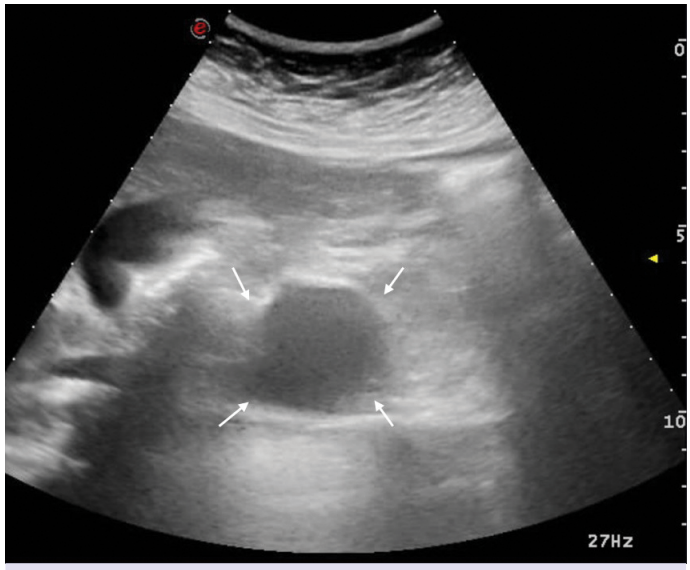

Figure 1. Transabdominal ultrasonography showed hypoechoic cystic lesion in the region of head of pancreas (white arrows).

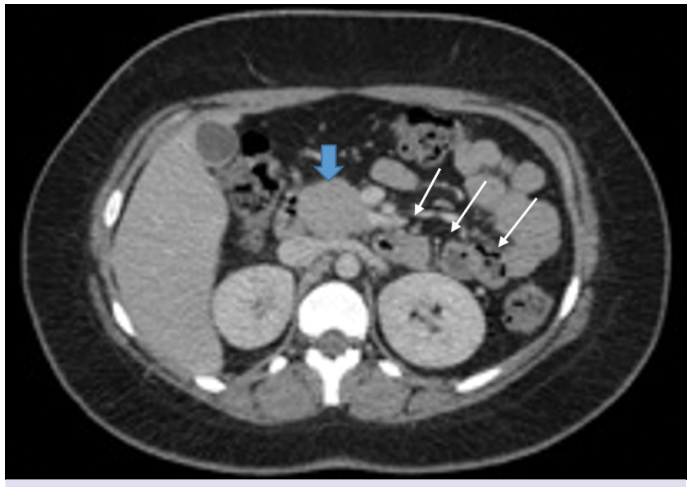

Figure 2. On the axial section, contrast-enhanced computed tomography scan images, head of pancreas could see (blue arrow) but the neck, body, and tail of the pancreas are not visualized and potential space is filled with intestinal loops (white arrows).

tral bud turns posteriorly to connect with the dorsal bud behind the duodenum to form the mature pancreatic gland [3, 4]. Developmental failures caused by abnormal embryogenesis may lead to partial or complete ADP [5]. In complete dorsal pancreatic agenesis, the body and the tail of the pancreas, the duct of Santorini, and the minor papilla are totally absent, whereas in partial agenesis, the minor papilla, the remnant of the accessory duct, and a small portion of the body of the pancreas can be seen [6].

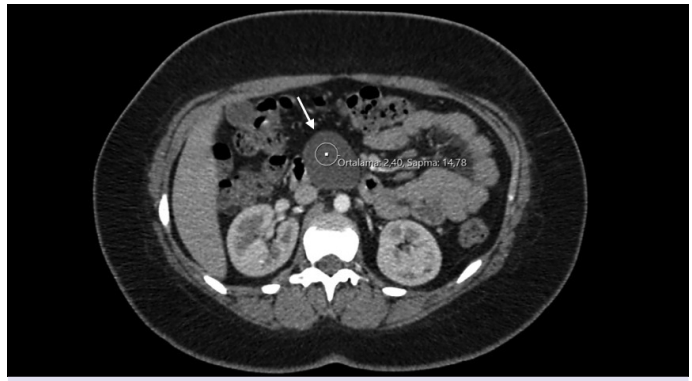

Figure 3. On the axial section, computed tomography scan, cystic lesion could see (arrow) in the area at head of pancreas. Density of the cystic lesion is 2.4 Hounsfield Unit (HU).

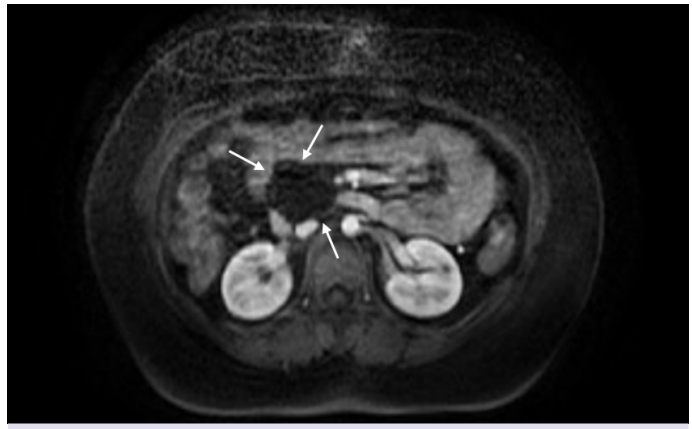

Figure 4. On the axial section, magnetic resonance imaging dynamic contrast-enhanced scan, cystic lesion showed no contrast enhancement in its wall (white arrows).

The first case of dorsal pancreatic agenesis was reported as an autopsy finding in 1911 [7] and there have been around 100 cases published in the literature until today.

Most ADP patients are usually asymptomatic. In symptomatic patients, abdominal pain is the most common symptom. Half of the ADP patients may present with hyperglycemia due to the involvement and underlying diabetes mellitus [8, 9]. Acute and chronic pancreatitis may also be seen in ADP patients due to Oddi sphincter dysfunction, enzyme hypersecretion, and higher pancreatic duct pressures [9, 10]. Chronic pancreatitis can lead to pseudocyst formations [11]. Our patient did not have any symptoms which can be associated with acute or chronic pancreatitis.

Despite its extremely rare occurrence, it is crucial to distinguish solid pseudopapillary neoplasms (SPNs) combined with ADP from cyst and pseudocyst forma-

tions. Because SPN may appear cystic due to its necrotic areas [12–14]. In our case, there were no solid component or calcification in the pancreatic cyst that may cause suspicion of SPN.

Other developmental abnormalities may accompany ADP such as polysplenia syndrome, heterotaxy, ectopic spleen, bowel malrotation, coarctation of the aorta, tetralogy of Fallot, and atrioventricular valvular abnormalities [15–17].

Pancreatic carcinoma with proximal atrophy, autodigestion from chronic pancreatitis, pancreatic divisum, pancreatic masses, distal pancreatic lipomatosis, and pseudoagenesis can mimic ADP [18, 19]. Chronic pancreatitis may cause replacement of the pancreas by fat and it may lead to atrophy of the pancreatic body and tail which is called "pseudoagenesis."

There are several useful modalities for the diagnosis of ADP. Initially, US is the most commonly used diagnostic modality for abdominal pain and other abdominal symptoms. On US, head of pancreas appears as a small hypoechoic structure in young population. At the junction of the head and neck of the pancreas, a hyperechoic line separates the hypoechoic pancreatic head from the more echogenic retroperitoneal fat. Nevertheless, US is often not adequate to visualize pancreas. Contrast-enhanced abdominal CT and MRI of the abdomen are better methods for ADP diagnosis. On CT study of abdomen, in the absence of pancreatic corpus and tail, small intestine or stomach can be seen within the distal pancreas area (dependent stomach or dependent intestine signs) [20]. MRI including Magnetic resonance cholangiopancreatography (MRCP) is a non-invasive method that can reveal the pancreatic ductal anatomy and confirm the absence of the dorsal duct system.

Treatment is usually symptomatic if there is not any suspicion for malignancy in pancreas.

Conclusion

ADP, although rare, sometimes could constitute a diagnostic challenge in routine practice. In our case, ADP was diagnosed evaluation of the patient for abdominal pain.

Informed Consent: Written, informed consent was obtained from the patient's family for the publication of thís case report and the accompanying images.

Conflict of Interest: No conflict of interest was declared by the authors.

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REFERENCES

1. Voldsgaard P, Kryger-Baggesen N, Lisse I. Agenesis of pancreas. Acta Paediatr 1994;83:791–3. [CrossRef]
2. Güçlü M, Serin E, Ulucan S, Kul K, Ozer B, Gümürdülü Y, et al. Agenesis of the dorsal pancreas in a patient with recurrent acute pancreatitis: case report and review. Gastrointest Endosc 2004;60:472–5. [CrossRef]
3. Adda G, Hannoun L, Loygue J. Development of the human pancreas: variations and pathology. A tentative classification. Anat Clin 1984;5:275–83.
4. Van Hoe L, Claikens B. The pancreas: normal radiological anatomy and variants. In: Baert AL, Delorme G, Van Hoe L, editors. Radiology of the Pancreas. Springer; 1999. p. 19–68. [CrossRef]
5. Schnedl WJ, Reisinger EC, Schreiber F, Pieber TR, Lipp RW, Krejs GJ. Complete and partial agenesis of the dorsal pancreas within one family. Gastrointest Endosc 1995;42:485–7. [CrossRef]
6. Fukuoka K, Ajiki T, Yamamoto M, Fujiwara H, Onoyama H, Fujita T, et al. Complete agenesis of the dorsal pancreas. J Hepatobiliary Pancreat Surg 1999;6:94–7. [CrossRef]
7. Heiberg KA. Ein Fall von fehlender Cauda pancreatis (bei einem Diabetiker). Zbl Path Anat 1911;22:676.
8. Lång K, Lasson A, Müller MF, Thorlacius H, Toth E, Olsson R. Dorsal agenesis of the pancreas - a rare cause of abdominal pain and insulin-dependent diabetes. Acta Radiol 2012;53:2–4. [CrossRef]
9. Schnedl WJ, Piswanger-Soelkner C, Wallner SJ, Reittner P, Krause R, Lipp RW, et al. Agenesis of the dorsal pancreas and associated diseases. Dig Dis Sci 2009;54:481–7. [CrossRef]
10. Uygur-Bayramiçli O, Dabak R, Kiliçoglu G, Dolapçioglu C, Oztas D. Dorsal pancreatic agenesis. JOP 2007;8:450–2.
11. Suh PS, Lee JH, Yu JS, Hee Kim J, Kim B, Kim HJ, et al. Three cases of pancreatic pseudocysts associated with dorsal pancreatic agenesis. Radiol Case Rep 2018;14:79–82. [CrossRef]
12. Jha BM, Shah P, Agarwal A. Solid pseudopapillary neoplasm of the pancreas mimicking a pseudocyst of pancreas. Med J Armed Forces India 2015;71:S5–7. [CrossRef]
13. Nakamura Y, Egami K, Maeda S, Hosone M, Onda M. Solid and papillary tumor of the pancreas complicating agenesis of the dorsal pancreas. J Hepatobiliary Pancreat Surg 2001;8:485–9. [CrossRef]
14. Ulusan S, Bal N, Kizilkilic O, Bolat F, Yildirim S, Yildirim T, Niron EA. Case report: solid-pseudopapillary tumour of the pancreas associated with dorsal agenesis. Br J Radiol 2005;78:441–3. [CrossRef]
15. Hatayama C, Wells TR. Syndrome of externally bilobed lungs with normal bronchial branch pattern, congenital heart disease, multiple spleens, intestinal malrotation and short pancreas: an apparently hitherto undefined malformation complex. Pediatr Pathol 1984;2:127–33.
16. Herman TE, Siegel MJ. Polysplenia syndrome with congenital short pancreas. AJR Am J Roentgenol 1991;156:799–800. [CrossRef]
17. Sakpal SV, Sexcius L, Babel N, Chamberlain RS. Agenesis of the dorsal pancreas and its association with pancreatic tumors. Pancreas 2009;38:367–73. [CrossRef]
18. Macari M, Giovanniello G, Blair L, Krinsky G. Diagnosis of agenesis of the dorsal pancreas with MR pancreatography. AJR Am J Roentgenol 1998;170:144–6. [CrossRef]
19. Rastogi R, Kumar R, Bhargava S, Rastogi V. Isolated pancreatic hypoplasia: a rare but significant radiological finding. Saudi J Gastroenterol. 2009 Oct-Dec;15(4):289-90. [CrossRef]
20. Mohapatra M, Mishra S, Dalai PC, Acharya SD, Nahak B, Ibrarullah M, et al. Imaging findings in agenesis of the dorsal pancreas. Report of three cases. JOP 2012;13:108–14.