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Case Report

Three cases of pancreatic pseudocysts associated with dorsal pancreatic agenesis

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ABSTRACT

Agenesis of the dorsal pancreas (ADP) is an extremely rare congenital anomaly. Human pancreas is formed by ventral and dorsal endodermal buds of the foregut endoderm. The dorsal bud forms the upper part of the head, neck, body, and tail of the pancreas and the ventral bud generates most of the head and uncinat process. ADP is derived from the embryologic failure of the dorsal pancreatic bud to form the pancreatic body and tail. ADP can be related to some diseases and conditions such as pancreatitis, hypoglycemia, and rarely pancreatic tumors. The association between cystic lesions with ADP has previously been reported. Three cases of cystic lesions of the pancreas with ADP were diagnosed clinically based on the imaging features and without any past history of pancreatitis. However, the pathologic diagnosis of resected lesions confirmed pseudocysts without pathologic evidence of tumor. We report 3 cases of pancreatic pseudocysts associated with ADP

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Introduction

Pancreatic divisum is the most common anomaly of the pancreas, and in autopsy series its prevalence was as high as 5%–10% [1]. Other common pancreatic anomalies include annular pancreas and agenesis of either the ventral or the dorsal bud. The exact frequency of agenesis of the dorsal pancreas (ADP) is not known, and has been reported only

rarely [1]. ADP is described in 2 forms: partial and complete [2]. Pancreatic anomalies are occasionally reported and the association of ADP has been reported. ADP can be related to some diseases and conditions such as pancreatitis, hypoglycemia, diabetic autonomic neuropathy, duodenal ileus, sphincter Oddi dysfunction, and rarely pancreatic tumors such as pancreatic adenocarcinoma [3].

The association between cystic lesions with ADP without past history of pancreatitis has been reported. To the best of

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my knowledge, only 2 cases of solid pseudopapillary neoplasm (SPN) and 1 case of mucinous cystic neoplasm of the pancreas in association with ADP have been reported [1,4,5].

Three cases of cystic lesions of the pancreas with ADP were diagnosed clinically based on the imaging features and without any history of pancreatitis. However, the pathologic diagnosis of resected lesions confirmed pseudocysts without pathologic evidence of tumor. We report 3 cases of pancreatic pseudocysts associated with ADP.

Case reports

Three unrelated female patients (ages 51, 62, and 75 years) with a clinical diagnosis of cystic pancreatic tumors with ADP were evaluated with computed tomography (CT) and other imaging studies. All patients underwent surgical resection and the pathologic diagnosis was pancreatic pseudocyst, because sections from the cyst wall showed extensive hemorrhage and necrosis without epithelial lining.

Case 1

A 51-year-old woman was admitted for the treatment of mass lesions of pancreatic head. She was diagnosed with noninsulin dependent diabetes mellitus 2 months earlier. She had no history of pancreatitis, and did not consume alcohol. Enhanced CT depicted cystic mass lesion in the pancreatic head with peripheral calcification and complete ADP (Fig. 1A). Endoscopic retrograde cholangiopancreatography (ERCP) showed the ventral Wirsung duct, without the dorsal pancreatic duct (not shown). Microscopically, the surgical specimen revealed a unilocular cyst with granulation tissue, recent hemorrhage and fibrosis without epithelial lining (Fig. 1B).

Case 2

A 75-year-old woman presented with left flank pain. Her medical history was unremarkable except for pulmonary tuberculosis 40 years ago. She had no history of pancreatitis and did not consume alcohol. Enhanced CT scan shows short pancreatic body without tail and round, cystic lesion with peripheral calcification (Fig. 2).

Case 3

A 62-year-old woman presented with epigastric discomfort. She had noninsulin-dependent diabetes mellitus diagnosed 1 month earlier. Her medical history was otherwise unremarkable with no past history of pancreatitis. A contrast-enhanced CT scan shows a calcified cystic mass in the pancreatic head and complete ADP (Fig. 3A). MR cholangiopancreatography (MRCP) maximum intensity projection image shows common bile duct and short ventral duct of Wirsung. No dorsal pancreatic duct is visualized (Fig. 3B).

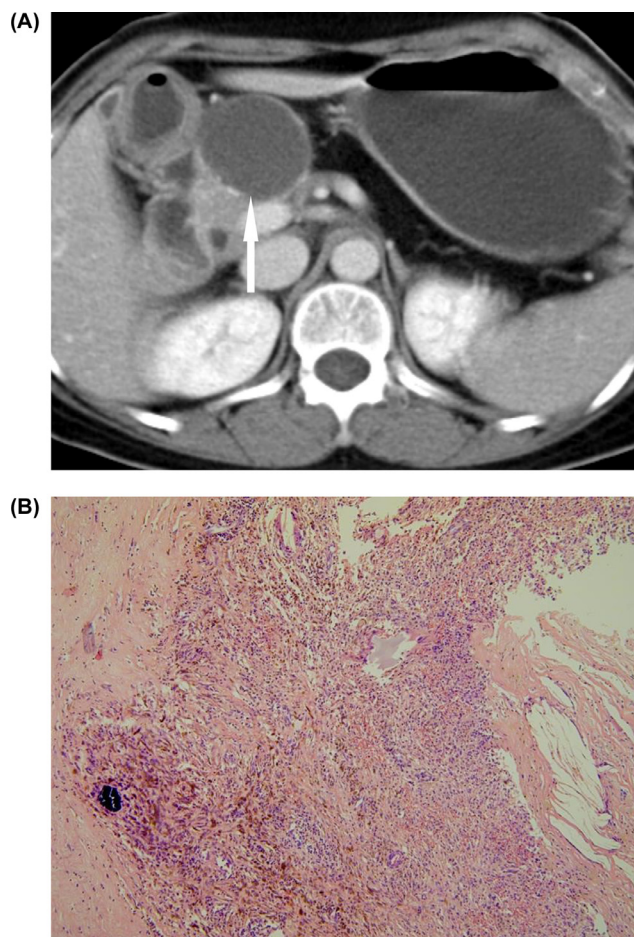


Fig. 1 – A 51-year-old woman with complete dorsal pancreatic agenesis with pseudocyst. (A) Contrast-enhanced CT shows round, cystic lesion (arrow) in the pancreatic head with peripheral calcification. The pancreatic body and tail are absent. (B) Photograph of the surgical specimen showing that the cystic lesion is surrounded by inflamed fibrous tissue, cholesterol granuloma, and dystrophic calcification. The luminal aspect has no epithelial lining.

Discussion

ADP is an extremely rare congenital anomaly, which results from embryologic failure of the dorsal pancreatic bud to form the pancreatic body and tail. It may be asymptomatic and detected incidentally on cross-sectional imaging [1,2].

Human pancreas is formed by ventral and dorsal endodermal buds of the foregut endoderm. The dorsal bud forms the upper part of the head, neck, body, and tail of the pancreas and drains through the duct of Santorini. The ventral bud generates most of the head and uncinate process, which drains through the duct of Wirsung [2]. ADP is derived embryologically from the absence or regression of the dorsal bud. ADP is described in 2 forms: partial or complete. In complete ADP, the minor papilla, the duct of Santorini, pancreatic neck, body, and tail are absent. By contrast, the minor papilla, the



Fig. 2 – A 75-year-old woman with partial agenesis of dorsal pancreas with pancreatic pseudocyst. (A) Contrast-enhanced CT shows short pancreatic body and cystic lesion with calcification. (B) Coronal MPR CT image also shows round, calcified mass (arrow) in the pancreas with no visible pancreatic parenchyma distal to the cystic mass.

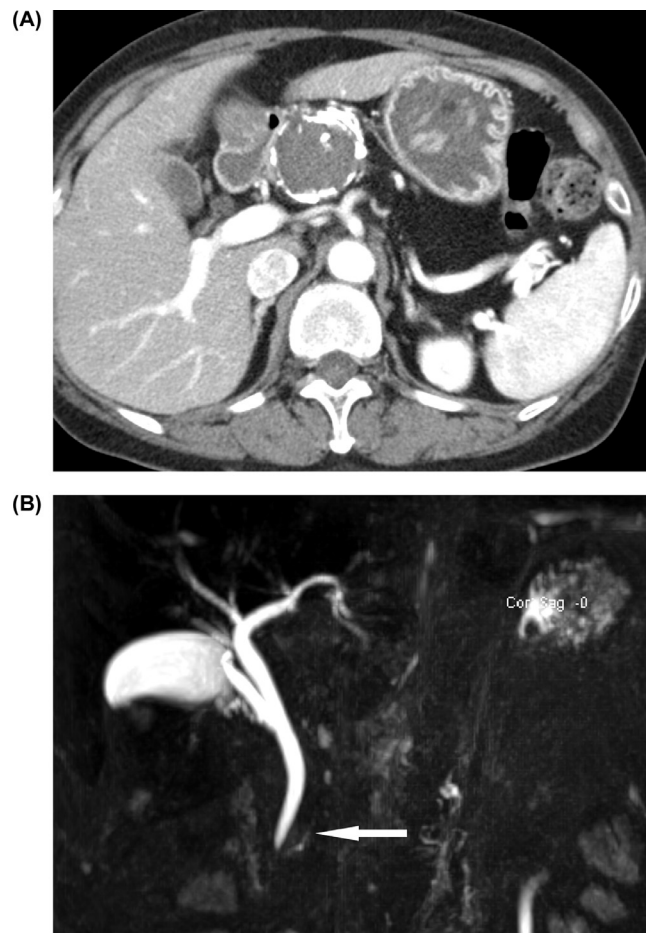


Fig. 3 – A 62-year-old woman with complete dorsal pancreatic agenesis with pseudocyst. (A) Contrast-enhanced CT shows round, cystic lesion in the head of pancreas with peripheral calcification and complete agenesis of dorsal pancreas. (B) MRCP maximum intensity projection image shows common bile duct and short ventral duct of Wirsung (arrow). No dorsal pancreatic duct is visualized.

duct of Santorini and variable amount of body of the pancreas are present in partial ADP [2,4].

Pancreatic fatty replacement and pseudoagenesis must be considered in the differential diagnosis of ADP [2]. Atrophy of the pancreatic body and tail secondary to acute or chronic pancreatitis, with sparing of the uncinete process may mimic ADP and has been designated as pseudoagenesis of the pancreas. This condition can be differentiated by demonstrating the dorsal duct, which is either absent or is very short in dorsal agenesis and usually present in lipomatosis and pseudoagenesis [2,4].

ERCP is necessary to confirm ADP by defining the anatomy of the pancreatic ducts. However, ERCP is an invasive procedure and operator-dependent for successful identification of opacity of the primary and accessory pancreatic ducts [2]. However, MRCP is a noninvasive examination, which adequately delineates pancreatic duct morphology [2]. MRCP may be extremely helpful in cases of cannulation failure of

the pancreatic duct. Therefore, the combined use of CT and ERCP or MRCP is useful to establish the diagnosis of ADP [2].

In our cases 1 and 3 with complete ADP, the dorsal pancreatic duct was not identified by ERCP or MRCP. Absence of the distal pancreas may contribute to the development of diabetes, as most insulin-secreting cells are located in the pancreatic body and tail [4]. However, our case 2 manifested partial ADP.

SPN associated with ADP is extremely rare, and can be misdiagnosed as a pseudocyst because of its cystic appearance and marked areas of necrosis [1,4,6,7]. Extensive hemorrhage and necrosis of SPN can mimic a pseudocyst of pancreas on gross examination [6,8]. Kumashiro et al. [8] reported a case manifesting typical clinical and imaging features of SPN of pancreas, whereas pathologic diagnosis of pseudocyst without tumor cells in the cyst wall.

Initially, we thought the calcified cystic mass in the pancreas as pancreatic cystic neoplasm such as SPN rather than

pseudocyst, because the patients had no past history of pancreatitis and occurrence of calcification is very rare in pancreatic pseudocyst [9].

It is believed that formation of pancreatic pseudocyst in our cases may have resulted from repeated subclinical pancreatitis, as ADP can be associated with clinical or subclinical pancreatitis, and pancreatic pseudocyst may calcify in cases with chronic pancreatitis [9].

In conclusion, we report 3 cases of pancreatic pseudocysts associated with ADP. Although ADP is a rare malformation, it can be associated with various medical diseases and tumorous conditions.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:[10.1016/j.radcr.2018.09.023](https://doi.org/10.1016/j.radcr.2018.09.023).

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