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Case Report

Pancreas bifidum: an extremely rare cause of acute pancreatitis☆

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ABSTRACT

A wide spectrum of anomalies of the pancreas, the pancreatic ductal system and the biliary tree are commonly encountered at radiologic evaluation. Pancreas bifidum, also known as bifid pancreas or fish-tail pancreas, is an extremely rare congenital branching anomaly of the main pancreatic duct characterized by its duplication. These 2 separate ducts are laid from the pancreatic tail to neck and they generally join at the pancreas body-tail draining via the major papilla; the pancreatic parenchyma is also bifurcated with separated dorsal and caudal buds. The clinical impact of this condition is not well established: although some authors sustained that probably does not cause or contribute to abdominal pain or overt pancreatic diseases, others argued that could be considered as a possible cause of acute pancreatitis.

We herewith describe the case of a 51-year-old woman presenting to our hospital with epigastric pain, nausea, and vomiting. Biochemical tests were suspicious for acute pancreatitis. Ultrasound examination was negative. MRI, including MR cholangiopancreatography revealed bifid pancreas characterized by duplication of the main pancreatic duct with 2 separate ducts that join at the pancreas head and draining via the minor papilla. On T2-weighted images the ventral bud of the pancreas was enlarged and characterized by slightly hyperintensity without peripancreatic fluid collections. The MRI findings were consistent with acute pancreatitis limited to the ventral bud of a bifid pancreas. Patient was treated with intravenous fluid resuscitation, pain control and institution of early enteral nutrition and discharged on the seventh day after admission.

Introduction

A wide spectrum of anomalies of the pancreas, the pancreatic ductal system and the biliary tree are commonly encountered at radiologic evaluation [1]. Pancreatic anatomic varia-

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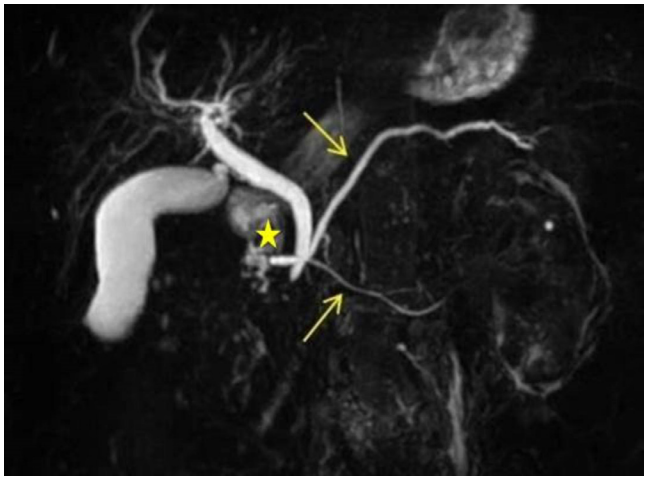

Fig. 1 – Coronal maximum intensity projection (MIP) reformat showed duplication of the main pancreatic duct (MPD) with two separate ducts that join at the pancreas head (arrows), and draining via the minor papilla (star). The biliary tract was not dilated and no signs of cholecysto-choledocholithiasis or biliary sludge were identified.

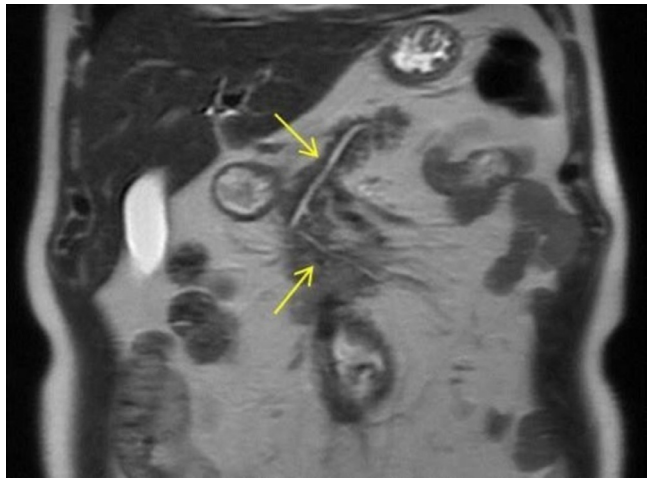

Fig. 2 – Coronal T2-weighted image revealed bifurcation of the pancreatic parenchyma (arrows) with bifid neck and a common head.

tions can be divided in 3 main types. The first group, migration anomalies, includes so-called ectopic pancreas and annular pancreas. The second group, fusion anomalies, represents failure of the dorsal bud to merge successfully with the duct of Santorini and the ventral bud with the duct of Wirsung, resulting in pancreas divisum, the most common pancreatic variation. The third and less common type of anomaly affects the number and form or only the configuration of the main pancreatic duct such as pancreas bifidum (PB) [2,3]. In particular, PB also known as bifid pancreas or fish-tail pancreas, is an extremely rare congenital branching anomaly of the main pancreatic duct (MPD) characterized by its duplication [4]. These 2 separate ducts are laid from the pancreatic tail to neck and they generally join at the pancreas body-tail draining via the major papilla; pancreatic parenchyma is bifurcated with separated dorsal and caudal buds. Usually, the pancreatic tail does not tend to reach the splenic hilum [4]. The exact frequency of the disease has not been reported since it is usually an incidental finding requiring no treatment. We found less than 25 case reports in literature describing “pancreas bifidum”, “bifid pancreas” or a “bifid tail of the pancreas” [3–26]. Knowledge of pancreatic embryology and of normal anatomic variants is essential to identify these entities and help differentiate them from pathologic conditions.

We herewith describe the case of a 51-year-old woman presenting to our hospital with epigastric pain, nausea and vomiting. Biochemical tests were suspicious for acute pancreatitis. Ultrasound examination was negative. MRI, including MR cholangiopancreatography (MRCP) revealed PB characterized by duplication of the main pancreatic duct with two separate ducts that join at the pancreas head and draining via the minor papilla.

Case report

A 51-year-old woman, with a history of recurrent abdominal pain presented to the Emergency Department of our hospital with upper abdominal pain radiating to the back that began in the epigastric region, nausea, and vomiting, suspicious for acute pancreatitis. The patient had no known risk factors such as history of alcoholism, smoking, autoimmune disorders or regular use of medications. There was no history of surgical interventions nor endoscopic retrograde cholangiopancreatography (ERCP).

On physical exam, the abdomen was slightly tender to palpations, particularly over the epigastric region, with positive rebound with decreased bowel sounds. The rest of physical exam was unremarkable.

Laboratory tests showed white blood cell count of 13,500/mm^3, amylase of 228 (normal 30-151) U/L, lipase of 3150 (normal 11-82) U/L. and elevated C-reactive protein (62 (normal less than 5). Liver tests revealed normal aspartate aminotransferase (AS) of 32 (normal 14-36) U/L, alanine aminotransferase (ALT) of 23 (normal 9-52), alkaline phosphatase of 72 (normal 53-141) U/L, and total bilirubin of 0,4 g/dL. Patient had a previous allergic reaction to iodinated radiological contrast media.

Ultrasound examination was regular and in particular no signs of cholecysto-choledocholithiasis or biliary sludge were identified; the biliary ducts were not dilated. Subsequently we performed an MRI, including MR cholangiopancreatography (MRCP), to better assess the hepato-pancreato-biliary tract. MRI revealed duplication of the main pancreatic duct with 2 separate ducts that join at the pancreas head and draining via the minor papilla (Fig. 1). Pancreatic parenchyma was also bifurcated with bifid neck and a common head (Fig. 2). The findings were compatible with a rare case of PB. In particular, on axial T2-weighted images with fat-suppression, the body-tail of the ventral limb of the pancreas was enlarged and

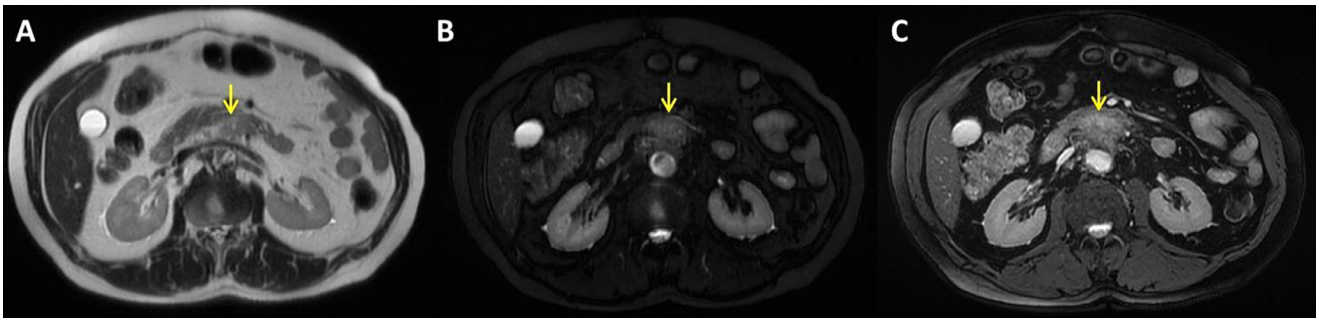

Fig. 3 – Axial T2-weighted sequence (A) Axial T2-weighted sequence with fat-suppression (B) and FIESTA (Fast Imaging Employing Steady-State Acquisition) image (C) demonstrated an enlargement of the body-tail of the ventral bud of the pancreas characterized by slightly hyperintensity (arrow). No peripancreatic fluid collections were detected. The MRI findings were consistent with localized acute pancreatitis.

characterized by sligthly hyperintensity without peripancreatic fluid collections. No abnormal areas of parenchymal signal were observed in the pancreatic head or dorsal bud. The biliary tract was not dilated and pancreatic or periampullary lesions were excluded (Fig. 3). Autoimmune causes of pancreatitis were ruled out by blood tests.

The MRI findings were consistent with the clinical suspicious of acute pancreatitis and the most interesting aspect was that the inflammation was limited to the ventral bud of the pancreas. Since the most common causes were excluded, PB was considered to be responsible for the acute pancreatitis.

Patient was treated with intravenous fluid resuscitation, pain control and institution of early enteral nutrition with a rapid improvement in clinical conditions and she was discharged on the seventh day after admission without complications. No recurrence of symptoms was observed in the following 15 months of follow-up.

Discussion

By the 4th week of embryologic development, the pancreatic duct develops from separate ventral and dorsal buds originating from the endodermal lining of the duodenum. The gallbladder, extrahepatic bile ducts, central intrahepatic bile ducts, and ventral pancreas with its ductal network are derived from the ventral bud or outpouching; the dorsal bud is the precursor to dorsal pancreas and its ductal system. At approximately the 7th gestational week, the ventral pancreas rotates clockwise posterior to the duodenum and comes into contact with the dorsal pancreas in the seventh gestational week to develop into the future pancreatic neck. The dorsal and ventral pancreatic buds grow into a pair of branching, arborizing ductal systems, each with its own central or main duct. The two anlagen fuse with each other and, together with the duodenum, fuse with the abdominal wall. After fusion, a new duct connects the distal portion of the dorsal pancreatic duct with the shorter duct of the ventral pancreas to form the main pancreatic duct, also known as the duct of Wirsung, which empties into the major papilla. The remnant of the dorsal duct forms the duct of Santorini, which drains into minor papilla [1,2]. In 30% of individuals, however, the duct of Santorini loses its communication with the minor papilla and persists only as a branch of the MPD.

The process of pancreatic fusion is complicated, and a wide spectrum of anatomic variants may occur during the course of pancreatic development [1,2].

During pancreatic development the ventral anlagen initially has 2 lobes with 2 primitive ducts. One lobe eventually develops to become the main pancreatic duct, the other lobe and duct regress and is thought to represent what becomes the uncinated process of the fully developed pancreas. It has been suggested that PB is caused by failure of one of these lobes to regress producing duplication of the MPD creating a dorsal and ventral tail [3].

The exact frequency of the disease has not been reported since it is usually an incidental finding requiring no treatment. In a report describing 650 patients with duplication variants of the pancreatic ductal system investigated with ERCP, only 6 patients had pancreatic bifurcation [5,27]. We found less than 25 case reports in literature describing “pancreas bifidum”, “bifid pancreas” or a “bifid tail of the pancreas”. In most of the cases the duplication involved only the tail of the pancreas, there was a single case describing a bifid neck with a common head, such as in our patient, and three cases describing bifid pancreatic duct without mention of the anatomic bifid tail [3,4,6–10].

The differential diagnosis of PB included pancreas divisum, which is the most common congenital anomaly of the pancreatic ductal system (occurring in 4%-14% of the population), and annular pancreas [1,2].

Pancreas divisum results when the ventral and dorsal pancreatic ducts fail to fuse. The ventral duct (Wirsung) drains only the ventral pancreatic anlage, whereas the majority of the gland empties into the minor papilla through the dorsal duct (Santorini) [2]. Focal dilatation of the terminal portion of the dorsal pancreatic duct, a condition known as Santorinicele, is described in association with pancreas divisum and relative obstruction at the minor papilla [2]. In most cases, there is no communication between the ventral and dorsal duct, independent drainage sites and dominant dorsal pancreatic duct with ventral duct typically short and very narrow. However in some individuals, there is a filamentous commu-

nication remaining and in others the ventral duct is totally absent [1]. Pancreas divisum is usually asymptomatic, but is more frequently seen in patients with chronic abdominal pain and idiopathic pancreatitis than in the general population.

The main anatomic feature of pancreas divisum, continuity of the dorsal pancreatic duct with the duct of Santorini draining into the minor papilla, is readily identified at both ERCP and MRCP.

On MRCP images, the normal ventral system may be so small in caliber that the ducts are not visible; however, it is the predominant pancreatic duct drainage into the minor papilla at a level superior to the level of the bile duct emptying into the major papilla that indicates the presence of divisum. The administration of secretin during MRCP increases the visibility of the main pancreatic duct and its side branches and also increases the sensitivity and specificity for detection of anatomic variants such as pancreas divisum. With the advent of multidetector CT scanners and high-spatial-resolution thin-section imaging, pancreas divisum may be routinely seen with use of CT as well [2].

As previously described, in PB MRI revealed duplication of the main pancreatic duct with 2 separate ducts that join at the pancreas head and draining via the minor papilla. Pancreatic parenchyma was also bifurcated with bifid neck and a common head.

Annular pancreas is a rare congenital anomaly (with a prevalence of 1 in 20,000 persons) in which incomplete rotation of the ventral anlage leads to a segment of the pancreas encircling the second part of duodenum, either completely or incompletely. There are 2 types of annular pancreas: extramural and intramural. In the extramural type, the ventral pancreatic duct encircles the duodenum to join the main pancreatic duct; in the intramural type, the pancreatic tissue is intermingled with muscle fibers in the duodenum wall, and small ducts drain directly into the duodenum [1]. Annular pancreas usually presents in neonates with vomiting due to severe duodenal obstruction. In adults, approximately 50% of the patients may be asymptomatic for life, with the anomaly discovered incidentally. When symptomatic, the presentation is usually in the third to sixth decade with abdominal pain, postprandial fullness and vomiting resulting from gastric outlet obstruction, upper gastrointestinal bleeding from peptic ulceration, acute or chronic pancreatitis, or in rare instances, jaundice from biliary obstruction [5]. The anomaly can be diagnosed on the basis of CT and MRI. Findings on CT scans include enlargement of the pancreatic head, which encircles the second portion of the duodenum; MR imaging demonstrates pancreatic tissue and occasionally the small annular duct encircling the descending duodenum [2].

To the best of our knowledge this is the first case of PB with 2 separate pancreatic ducts that join at the pancreatic head and draining via the minor papilla.

There are no currently established procedures to follow in the case of this anomaly. The clinical impact of this condition is not well established – in the vast majority of cases PB represents an incidental finding, with asymptomatic presentation and benign course. In these cases the anatomical variation does not cause or contribute to abdominal pain or overt pancreatic disease and requires no specific therapy once diagnosed correctly. However, some authors supported the hypothesis that PB could cause or act as a predisposing factor of acute and/or chronic pancreatitis and in particular the inflammation is usually confined to only one of the 2 tails [3,6–8]. The ventral bud is more frequently involved by the inflammation probably because of the relatively small caliber of the ventral duct that that could lead to a higher risk of outflow obstruction [7].

Knowledge of pancreatic embryology and normal anatomic variants is essentials to identify these entities and help differentiate them from pathologic conditions, thus preventing potential unnecessary imaging investigation or more invasive procedures such as biopsy or surgery. Recognition of developmental anomalies of the pancreas and pancreatic duct is important also because these anomalies may be a surgically correctable cause of recurrent pancreatitis or the cause of gastric outlet obstruction.

In conclusion, we present one of the first cases of a patient with acute pancreatitis limited to one side of a PB that suggests the consideration that this rare congenital malformation may increase the predisposition to localized acute pancreatitis. PB should be considered an additional cause of localized acute pancreatitis when no other anatomical ductal abnormality is highlighted on MRCP.

Patient consent statement

Written informed consent was obtained from the patient for publication of this case report, including accompanying images. The data that support the findings of this study are available from the corresponding author, [LV], upon reasonable request.

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