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## Case Report

# Idiopathic recurrent acute pancreatitis in the context of pancreas divisum: A case report<sup>☆</sup>

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## ARTICLE INFO

## Article history:

Received 2 June 2020

Revised 21 July 2020

Accepted 24 July 2020

## Keywords:

Recurrent acute pancreatitis

Idiopathic

Pancreas divisum

Multidetector CT (MDCT)

Minimal-intensity-projection

(MinIP) reconstruction

## ABSTRACT

Recurrent acute pancreatitis is characterized by frequent episodes of inflammation in the pancreas. The most common causes are alcohol abuse and gallstones but approximately 30% of cases are defined as idiopathic because initial evaluation fails to detect the etiology. In these patients, extensive laboratory and imaging evaluations usually lead to the uncovering of an occult biliary duct stone or sphincter of Oddi dysfunction as the main reason for frequent pancreatitis. We report a 42-year-old female with a longstanding history of acute recurrent pancreatitis in the setting of pancreas divisum due to its rarity. We focus on appropriate investigations by application of specialized laboratory and radiological modalities because determining the etiology is a crucial step in patient management.

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## Introduction

Acute pancreatitis is a sudden inflammation of the pancreas associated with nausea, vomiting, appetite loss, and severe epigastric pain radiating to the back that can cause systemic inflammatory response syndrome. In recurrent cases, patients experience more than 2 attacks of pancreatitis [1]. The main characteristic that differentiates recurrent acute pancreatitis from chronic pancreatitis is the normal morphology of the

pancreas but the initial presentation of chronic pancreatitis may not become evident until after a few years. Although most cases of acute pancreatitis result from gallstones or alcohol consumption, in almost one-third of cases, the cause of pancreatitis is undiagnosed by initial study and, as a result, “idiopathic” recurrent acute pancreatitis is used to describe this disease of unknown etiology. In such patients, specified laboratory tests and imaging modalities like endoscopic retrograde cholangiopancreatography (ERCP), magnetic resonance choangiopancreatography (MRCP), and endoscopic ultrasound

**Abbreviations:** MRI, magnetic resonance imaging; CT, computed tomography; MDCT, multidetector CT; MinIP, minimal intensity projection; ERCP, endoscopic retrograde cholangiopancreatography; MRCP, magnetic resonance cholangiopancreatography; EUS, endoscopic ultrasound; EGD, esophagogastroduodenoscopy.

<sup>☆</sup> Acknowledgment: The authors thank senior science writer Edmund Weisberg, MS, MBE, who helped us to review and edit the article.

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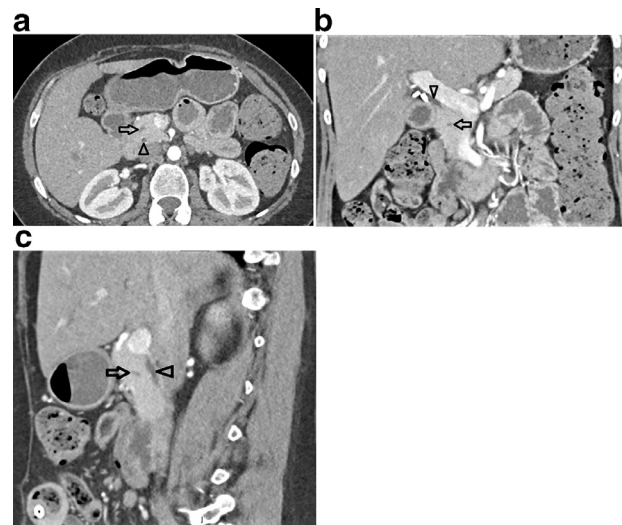
<https://doi.org/10.1016/j.radcr.2020.07.065>

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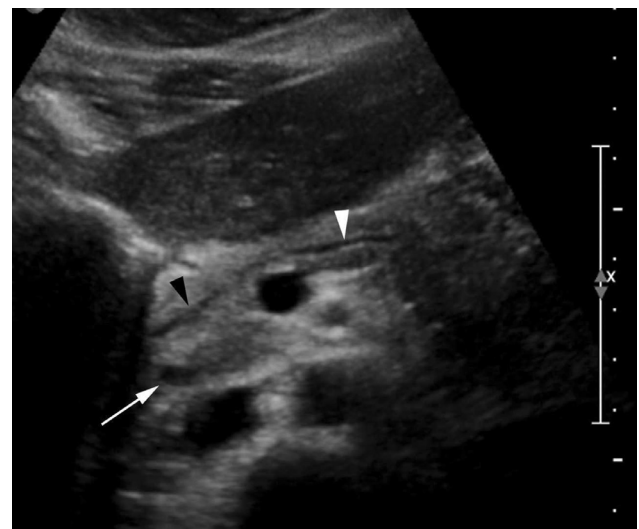
(EUS) can help to identify less common causes like occult biliary stone disease, sphincter of Oddi dysfunction, and pancreatic duct anatomical abnormalities [2,3]. In some cases, even after extensive evaluation, the main etiology may not be discovered. The first step of therapy consists of supportive care with fluid replacement, pain control, nothing by mouth, and nutritional support. In the next step, proper treatment should be chosen based on the etiology of acute pancreatitis. Determining the cause(s) of acute pancreatitis is important in directing proper treatment and improvement of long-term prognosis. Pancreatectomy with islet autotransplantation is considered a last resort in a patient with recurrent acute pancreatitis when other treatments have failed [4].

### Case report

We report a 42-year-old female with a significant past medical history for idiopathic recurrent acute pancreatitis with pancreas divisum, chronic abdominal pain, hypothyroidism, polycystic ovary syndrome, cholecystectomy, appendectomy, cervical spine surgery, and vitamin D deficiency. Her initial episode of pancreatitis started more than twenty years ago during pregnancy when she suffered from 4 episodes of acute pancreatitis and went on to have a cholecystectomy during the pregnancy. She had reported episodes of acute pancreatitis and underwent diagnostic ERCP at an outside hospital, and the appearance of the pancreatogram was consistent with pancreas divisum. She underwent minor papilla sphincterotomy due to pancreas divisum a year later and did well for a good period afterward. However, she began having multiple episodes of acute pancreatitis ten years ago with elevated pancreatic enzymes documented, requiring multiple hospitalizations despite normal ERCP and EUS findings except pancreas divisum. The pancreatic parenchyma was normal in signal and appearance without mass or pancreatic duct dilatation on these tests. Her symptoms have increased in severity over the past year to the point where she required total parenteral nutrition. She also underwent an EUS with a celiac plexus block, which lessened pain severity and the need for pain medications. She needed to stop parenteral nutrition due to line sepsis and had a laparoscopic jejunostomy tube placed through which she was receiving enteral nutrition. Other than a stenotic minor papillary orifice on ERCP, she did not display significant radiographic features of acute or chronic pancreatitis on imaging studies including multidetector CT (MDCT) or transabdominal ultrasound (Figs. 1 and 2). MDCT with minimal intensity projection (MinIP) reconstruction image clearly demonstrated pancreas divisum anatomy (Fig. 3). Several years ago, she was evaluated in a multidisciplinary pancreatitis clinic and was deemed eligible for total pancreatectomy, splenectomy, and autologous islet cell transplant. Her postoperation course was complicated by a nonhealing incision and an abdominal wall abscess that required multiple admissions for drainage, wound packing, and IV antibiotic therapy. The patient's history also was remarkable for a pyogenic liver abscess that developed on hematoma after a trauma, which was managed by percutaneous abscess



**Fig. 1 – Contrast-enhanced arterial phase computed tomography scan showing pancreas divisum and normal pancreatic parenchyma without peripancreatic inflammatory changes or fluid collection: (A) axial (B) coronal (C) sagittal reformations. The accessory pancreatic duct (duct of Santorini) (arrow) extends to the minor papilla anterior to the common bile duct (arrowhead). The duct of Wirsung is not visible on computed tomography.**



**Fig. 2 – Gray-scale transverse image of transabdominal ultrasonography illustrates pancreas divisum with normal caliber common bile duct (arrow) with normal caliber main pancreatic duct (white arrowhead) communicating with accessory pancreatic duct (duct of Santorini) (black arrowhead) entering the minor papilla anterior to the common bile duct. The pancreatic parenchyma has normal echogenicity without pseudocyst.**



**Fig. 3 – Oblique coronal minimal intensity projection (MinIP) reconstruction image of contrast-enhanced arterial phase computed tomography shows the accessory pancreatic duct (duct of Santorini) (black arrowhead) entering the minor papilla. The air within the main pancreatic duct (white arrowhead) is secondary to minor papilla sphincterotomy. The common bile duct (white arrow) drains to the major papilla inferior to the minor papilla.**

drainage. At that time, she reported vomiting, postprandial abdominal pain, uncontrolled diabetes mellitus, and multiple hospitalizations for drug-induced serotonin syndrome resulting in fever and leukocytosis. She recovered from postoperative complications, but a few years later, she was hospitalized for one week due to active bleeding of a gastroduodenal anastomosis ulcer found on esophagogastroduodenoscopy. After treatment for the ulcer, she did well and, one year later, her follow-up esophagogastroduodenoscopy was unremarkable.

## Discussion

Recurrent acute pancreatitis is defined as more than 2 episodes of a sudden inflammation of the pancreas. Clinical symptoms usually start with epigastric pain that sometimes spreads to the back and is associated with nausea, vomiting, and fever. Most cases of acute pancreatitis result from biliary stone disease and longstanding alcohol consumption. Idiopathic acute pancreatitis in which the etiology of the clinical condition cannot be determined with an initial evaluation requires a focused approach and extensive workup to elucidate the cause [1]. Tests of choice for detecting rare causes are ERCP, EUS, and MRCP. Since ductal abnormalities, microlithiasis, and sphincter of Oddi dysfunction are responsible for the majority of the idiopathic cases, cholecystectomy and, eventually, pancreatic sphincterotomy is therapeutic in most cases [2]. Intractable cases like our presented case can be treated by celiac plexus block and total pancreatectomy with islet cell autotransplantation.

Pancreas divisum represents the most common congenital anomaly in pancreatic ductal anatomy in which two distinct

dorsal (duct of Santorini) and ventral ducts (Wirsung duct) fail to fuse. This condition may occur in approximately 10% of the population [5]. In such cases, the dorsal duct plays the main role in drainage of pancreatic secretions through minor papilla and a small ventral duct opens to the major papilla [6]. Most individuals with pancreas divisum remain asymptomatic but in about 5% of cases, it can be the hidden reason for idiopathic recurrent acute pancreatitis. It has been suggested that dorsal duct opening to relatively smaller minor papilla increases intraductal pressure which could transiently impair drainage of pancreatic secretion and causes inflammation [7].

For the diagnosis of pancreas divisum, detailed imaging studies such as ERCP, MRCP, and MDCT is required to detect this anomaly. The key imaging features of the pancreas divisum are separate pancreatic ducts of Santorini and Wirsung which drain to the minor and major ampulla, respectively. The dorsal duct is located anterior to ventral and common bile ducts in the pancreatic head. ERCP is the most accurate test for diagnosing pancreatic divisum but is an invasive procedure with potential complications such as pancreatitis, perforation, and bleeding. These days, with the availability of non-invasive imaging like EUS and MRCP, ERCP mostly has a therapeutic role. MRCP produces high-quality images of pancreatobiliary tree, and is the current gold standard method of evaluation [8]. Another imaging modality that is broadly used for visualization of the pancreas is EUS but because of intervening air, it has lower sensitivity and specificity compared to other imaging modalities. In our case, MinIP reconstruction image of MDCT well demonstrated pancreas divisum anatomy with relationship to the common bile duct (Fig. 3). Unlike maximum intensity projection (MIP) imaging, MinIP reconstructions project the lowest attenuation voxels in a dataset, and useful for visualization of fluid-filled structures such as the biliary or pancreatic ducts, particularly when these structures are dilated or obstructed [9,10]. This technique is also useful for demonstration of congenital anomalies of the pancreatic duct including pancreas divisum [10] and annular pancreas [11].

No treatment is necessary in asymptomatic cases of pancreas divisum, but if the patient is symptomatic, the best approach is decided based on the intensity, frequency of symptoms, and presence of complications. Treatment in the case of acute, chronic, or recurrent pancreatitis secondary to pancreas divisum is endoscopic and surgical treatment. Endoscopic therapy includes endoscopic interventions include minor papillotomy, trans minor papilla dorsal duct stenting, and balloon dilation of the minor papilla. A surgical minor papilla procedure such as surgical sphincteroplasty is reserved for patients who fail endotherapy [5].

Venesmaa et al [12] described a case of a 17-year-old boy with recurrent episodes of pancreatitis with pancreas divisum in 2013. An abnormal pancreatic duct was seen in ultrasonography. Irregularity and focal dilation of the terminal part of the main pancreatic duct was found on MRCP. The diagnosis was reached by ERCP, which showed a small stone at the minor papilla of the accessory duct. The stone was removed and papillotomy was performed successfully. The patient was asymptomatic in the 6-month follow-up visit. Anyfantakis et al [13] reported a 54-year-old Caucasian male who was

admitted to the hospital with intermittent epigastric pain accompanied by nausea. Pancreas divisum was diagnosed based on illustrating two separate pancreatic ducts in MRI. The diagnosis was confirmed with MRCP that revealed findings suggestive of pancreas divisum. Recently, Kuzel et al [14] published a case of a 20-year-old female with a history of recurrent acute pancreatitis admitted to the hospital complaining of severe and sharp abdominal pain and nausea. A 10-mm calcification in the head of the pancreas was discovered in CT scan. MRCP findings showed inflammation in the pancreas and evidence of pancreas divisum with a small remnant connection between the dorsal duct as the main source of drainage and the ventral duct. The patient was encouraged to change her lifestyle, namely stop smoking and drinking alcohol and following a low-fat diet to avoid any further episodes of pancreatitis. In the next step, patients with recurrent acute pancreatitis due to pancreas divisum corrective surgical options can be considered.

Our report describes in detail a case of real idiopathic recurrent acute pancreatitis in the context of pancreas divisum, where even a thorough study could not pinpoint the exact etiology. This case report highlights the diagnostic utility of MDCT with MinIP reconstruction image and ultrasonography in the evaluation of recurrent acute pancreatitis and also different approaches in the management of the disease.

#### REFERENCES

- [1] Testoni PA. Acute recurrent pancreatitis: etiopathogenesis, diagnosis and treatment. *World J Gastroenterol* 2014;20(45):16891–901.
- [2] Jagannath S, Garg PK. Recurrent acute pancreatitis: current concepts in the diagnosis and management. *Curr Treat Options Gastroenterol* 2018;16(4):449–65.
- [3] Somani P, Navaneethan U. Role of ERCP in patients with idiopathic recurrent acute pancreatitis. *Curr Treat Options Gastroenterol* 2016;14(3):327–39.
- [4] Sánchez Rodríguez E, García García de Paredes A, Albillos A. Current management of acute idiopathic pancreatitis and acute recurrent pancreatitis. *Rev Clin Esp* 2019;219(5):266–74.
- [5] Gutta A, Fogel E, Sherman S. Identification and management of pancreas divisum. *Expert Rev Gastroenterol Hepatol* 2019;13(11):1089–105.
- [6] Covantev S. Pancreas divisum: a reemerging risk factor for pancreatic diseases. *Rom J Intern Med* 2018;56(4):233–42.
- [7] Farooqi R, Burke C, Chahal P, El-Khider F, Zahid U. Acute pancreatitis in pancreas divisum secondary to an impacted stone in the minor papilla. *Cureus* 2019;11(8):e5481.
- [8] Wang D-B, Yu J, Fulcher AS, Turner MA. Pancreatitis in patients with pancreas divisum: imaging features at MRI and MRCP. *World J Gastroenterol* 2013;19(30):4907–16.
- [9] Raman SP, Fishman EK. Abnormalities of the distal common bile duct and ampulla: diagnostic approach and differential diagnosis using multiplanar reformations and 3D imaging. *AJR* 2014;203:17–28.
- [10] Salles A, Nino-Murcia M, Jeffrey RB. CT of pancreas: minimal intensity projections. *Abdom Imaging* 2008;33:207–203.
- [11] Kim HC, Park SI, Park SJ, Shin HC, Lee HK, Kim YT, et al. Diagnosis of annular pancreas using minimum intensity projection of multidetector row CT: case report. *J Korean Radiol Soc* 2004;51:641–4.
- [12] Venesmaa SV, Heikkinen M, Kainulainen S, Manninen H. Imaging and treatment features of idiopathic pancreatitis and pancreas divisum in a young man: a case report. *Gastroenterology Res* 2013;6(3):112–16.
- [13] Anyfantakis D, Partalis N, Polimili G, Kastanakis S. Acute non-traumatic pancreatitis in a patient with pancreas divisum: a case report. *J Med Life* 2013;6(3):332–5.
- [14] Kuzel AR, Lodhi MU, Rahim M. Pancreatic divisum: an unusual cause of chronic pancreatitis in a young patient. *Cureus* 2017;9(11):e1856.