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Case Report

Pancreas divisum causing recurrent pancreatitis in a young patient: A case report $^{\bigstar, \grave{\times} \diamond}$

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ARTICLE INFO

Article history: Received 24 March 2023 Revised 10 July 2023 Accepted 14 July 2023

Keywords:

Pancreas divisum MRI-magnetic resonance imaging Recurrent acute pancreatitis Magnetic resonance cholangiopancreatography Endoscopic retrograde cholangiopancreatography Case report

ABSTRACT

Pancreas divisum (PD) is the most common congenital variant of the pancreatic ductal system, in which only a few patients develop symptomatic disease. Overall, PD is an underrecognized cause of many cases of recurrent acute pancreatitis. The PD must be systematically suspected in case of multiple episodes of acute idiopathic pancreatitis when exhaustive etiological investigations are negative. We present a 37-year-old woman whom presented several previous pancreatic pains. She came to the emergency department for epigastric pain, accompanied by post-prandial dietary vomiting after a copious meal. Lipasemia was greater than 3 times normal (498 UI/L). An abdominal computed tomography (CT) and magnetic resonance cholangiopancreatography (MRCP) showed a PD. The patient improved after the initial management measures. An endoscopic cholangiopancreatography was planned after the resolution of the acute episode. Recurrent pancreatitis is defined as 2 or more episodes of distinct acute pancreatitis with more than 3 months between episodes. Patients with this condition are usually asymptomatic while 5% of patients develop acute pancreatitis or chronic pancreatitis. We can underline the interest of deepening the radiological and endoscopic investigations to make the diagnosis of PD and to propose an endoscopic or surgical treatment, in order to avoid recurrences.

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Introduction

Pancreatitis is a severe inflammatory disease that is critical in some patients. Alcohol usage and gallstone disease are 2 of the many recognized causes of pancreatitis. However, patients with acute recurring pancreatitis (ARP) present conditions such as pancreas divisum and genetic factors. About 4% of patients with pancreatic divisum (nonfused dorsal and ventral ducts) had a higher incidence of recurrent pancreatitis [1]. The majority of pancreatic divisum (PD) patients are asymptomatic, however in a few rare instances, PD may be the cause

☆☆ Competing Interests: None.

^{*} Acknowledgments: This research received no specific grant from any funding agency in the public, commercial or not-for-profit sectors.

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https://doi.org/10.1016/j.radcr.2023.07.028

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of repeated pancreatitis crises. The incidence varies, although it ranges from 5% to 14% of the general population [2].

In the case of repeated symptomatology, we must consider malignancies engaged in the dorsal canal in addition to the pancreas divisum [3,4]. We describe a rare instance of pancreatic divisum diagnosed after an etiological assessment of recurrent acute pancreatitis.

Case presentation

A 37-year-old woman had no history of smoking and consumed small amounts of plants. She had no history of alcohol intake and no history of metabolic disease (including diabetes or dyslipidemia). She had no autoimmune conditions, no biliary lithiasis, and no previous surgical history. There were no additional health issues or drug usage that would have increased the patient's risk of developing acute pancreatitis. She claimed to have experienced 2 earlier incidents of epigastric discomfort, but they were not fully investigated knowing her condition quickly got better following the severe event.

She presented to the emergency department with significant abdominal pain in the epigastric area, radiating to the back. The pain was also associated with nonbilious vomiting.

The patient was in good general condition, with a performance status at 1, apyretic, hemodynamically stable with a heart rate of 72 bpm, a normal blood pressure. Body mass index (BMI) was at 25 with a mild mucocutaneous pallor and no jaundice. She had no signs of dehydration or severe malnutrition. Abdominal examination found a nondistended abdomen with epigastric tenderness. The rectal examination was normal. The rest of the clinical examination was normal.

Laboratories examination showed a lipase 3 times higher than normal at 498 UI/L (normal range < 160 UI/L), with a Creactive protein at 53 mg/L (normal range < 5 mg/L) and creatinine at 3.7 mg/L (normal range 6.7-11.7 mg/L). The white blood cell count was 4100 cells/mm³ (normal range 4000-11,000 elements/mm³), and hemoglobin at 13.1 g/dL (normal range 13.0-18.0 g/dL). The liver tests were normal: aspartate aminotransferase of 16 IU/L (normal range < 36 IU/L), alanine aminotransferase of 12 UI/L (normal range < 36 IU/L), alkaline phosphatase of 50 UI/L (normal range 44-147 UI/L), γ -glutamyltransferase of 49 UI/L (normal range 5-40 UI/L), total bilirubin at 5.9 mg/dL (normal range < 12 mg/L) and direct bilirubin 2.3 mg/dL (normal range < 2 mg/dL). Lipid analyses were normal knowing that total cholesterol concentrations measured at 0.98 g/L (normal range < 2 g/L), the serum triglyceride at 0.56 g/L (normal range 0.35-1.40 g/L), the concentration of serum calcium at 88 mg/L (normal range 86-100 mg/L). Hepatitis B and C virus serology and serum IgG4 levels were normal. The abdominal ultrasonography (US) did not show any gallstones.

A thoracic-abdominopelvic computed tomography (CT) scan showed the ventral pancreatic duct and the dorsal pancreatic duct in a parallel direction indicating an anatomical variant (complete pancreas divisum). The pancreatitis was classified as stage A according to the Balthazar staging (Fig. 1).

A Magnetic resonance cholangiopancreatography (MRCP) was performed which showed the main channel that joins with the bile duct at the level of the major papilla



Fig. 1 – Axial contrast-enhanced CT scan in the portal phase highlighting the ventral pancreatic duct (green arrow) and the dorsal pancreatic duct (red arrow) in a parallel direction indicating an anatomical variant (complete pancreas divisum). The enhancement of the pancreatic parenchyma is normal and there are no foci of glandular necrosis in favor of pancreatitis stage A according to the Baltazar staging.

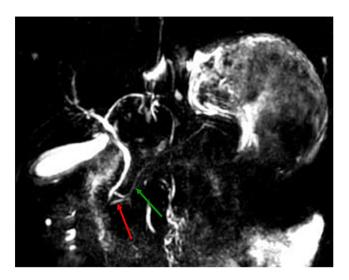


Fig. 2 – 3D biliary MRI of the anatomical variant of a pancreas divisum: – Green arrow: The ventral duct (main pancreatic duct) joins the bile duct at the level of the main papilla – Red arrow: The dorsal canal (accessory canal of Santorini) ends in the minor papilla.

and the accessory channel that terminated alone at the level of the minor papilla (Figs. 2 and 3).

The patient was managed with rehydration, administration of proton pump inhibitors, and analgesic treatment. The patient evolved well after the resumption of eating and loss of pain. Following the resolution of the acute episode, an endoscopic retrograde cholangiopancreatography (ERCP) was planned to perform endoscopic drainage via minor papillotomy but was refused by the patient. The patient had no other episodes of acute pancreatitis during the 12 months. The risk

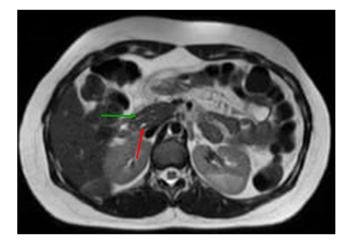


Fig. 3 – Axial section of MRCP in T2-single-shot fast spin-echo sequence showing the ventral pancreatic duct (green arrow) and the dorsal pancreatic duct (red arrow) in a parallel direction demonstrating a complete pancreas divisum.

of recurrence of the symptoms was explained to the patient in the absence of endoscopic treatment

Discussion

Recurrent pancreatitis is defined as 2 or more episodes of distinct acute pancreatitis separated by more than 3 months between episodes according to the current definition of RAP [5]. PD may be associated with RAP due to insufficient drainage of pancreatic secretions through the dorsal pancreatic duct and papilla minor. PD is a congenital embryonic disease caused by a lack of fusion between the ventral and dorsal pancreatic ducts in the early stages of embryogenesis. It is the result of a developmental abnormality in which the dorsal canal (minor or accessory canal) and the ventral (main channel) of the pancreas merge incompletely or not at all [6]. It is found in less than 10% of individuals. There are 3 types of PD identified: classic or complete divisum, dominant type or dorsal duct PD, and incomplete PD [3,7]. In our case, the type was complete divisum.

Patients with this condition are usually asymptomatic while 5 % of patients develop acute pancreatitis or chronic pancreatitis [8].

MRCP is used to thoroughly assess the bile ducts and pancreatic ducts. It is a fast, noninvasive method of evaluating the pancreatic ductal system [9]. The use of ERCP is controversial, although ERCP has been shown to have high diagnostic accuracy in the excellent description of the bile duct and pancreatic system [7,8]. Secretin-enhanced MRCP (S-MRCP) has been developed specifically for the evaluation of the pancreas and pancreatic ducts. Further, it improves the diagnostic yield of PD. In our case, MRCP confirmed the diagnosis.

It is important to have the reflex to think of pancreatic cancer associated with PD in the event of recurrent symptomatology and deterioration of the general state [7]. The long-term efficacy of endoscopic treatment in PD is controversial [10]. Endoscopic treatment is performed via a minor papillotomy and, if necessary, by dilation of the canal with the implantation of a plastic stent in the pancreatic canal [11]. The insertion of a stent into the dorsal canal and minor papillary sphincterotomy decrease the rate of recurrent acute pancreatitis. Surgical treatment is required in cases of failure of endoscopic drainage and in complicated cases of chronic pancreatitis and local complications [12].

Treatments via the minor papilla are effective where deep cannulation via the major papilla is difficult [12]. The cannulation through the minor papilla requires superior endoscopic skills [13]. Endoscopists must be well-trained to become familiar with the anatomical variants of the pancreas and its canal system. The limitation of the use of this difficult therapeutic approach is considered an issue in the management of PD.

Conclusion

PD can be marked by recurrent pancreatitis which is particularly significant. Our report describes in detail a case of real idiopathic recurrent acute pancreatitis in the context of PD. To prevent the pancreas' chronic degeneration and repeated hospital stays, it is crucial to detect the congenital defect of the pancreas sooner. Through this case, we tried to sensitize gastroenterologists to think about the diagnosis of PD in a young patient with recurrent pancreatitis. The diagnosis requires careful clinical evaluation and an inquiring history of past recurrent pancreatitis attacks. For improved care of this uncommon entity, endoscopic and radiographic examinations must be expanded.

Patient consent

Consent for publication has been obtained from the patient

Provenance and peer review

Not commissioned, externally peer-reviewed.

Author contribution

All authors contributed.

Research registration

Not applicable.

Guarantor

Not applicable.

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