Bifurcation of the main pancreatic duct in the body of the pancreas. Two case reports and literature study of a rare anatomical variant of the pancreatic duct

L. Krott[1], D. De Wulf[2], S. De Coninck[3], H. Degroote[1]

(1) University hospital Ghent, Department of Gastroenterology, Ghent, Belgium ; (2) AZ Delta Roeselare, Department of Gastroenterology, Roeselare, Belgium ; (3) AZ Sint-Andries Tielt, Department of Gastroenterology, Tielt, Belgium..

Abstract

Introduction **: Bifurcation of the pancreatic duct is a very rare anomaly and clinical significance is not known. Literature on this topic is scarce. We present two similar case reports with bifurcation of the main pancreatic duct from the body to the tail of the pancreas. Both cases were symptomatic, one had acute pancreatitis and the other recurrent pancreatitis. In both cases the most ventral duct was aberrant as a consequence of pancreatitis.**

Discussion **: We performed a literature study and found 22 relevant articles containing 26 case reports, of these cases, 12 were considered asymptomatic and were found incidentally, the other 14 cases were symptomatic with signs of acute, chronic or recurrent pancreatitis. To our knowledge this is the first article with a summary of previous published data on the subject.**

Conclusion **: Bifurcation of the pancreatic duct seems to be a possible cause of pancreatitis, but a large group remains asymptomatic. Since diagnosis is often difficult, the incidence is probably underestimated. More attention to this anomaly is recommended. Further reports are needed to draw conclusion.** (Acta gastroenterol. belg., **2020**, 83, **639-642**).

Key words : pancreatic duct anomaly, bifurcated pancreatic duct, pancreatitis, clinical relevance.

Introduction

A wide variety of anatomic anomalies are seen in the pancreas. Bifurcation of the pancreatic duct is a very rare pancreatic anomaly. This variation is only described in case reports and its relevance remains questionable. In this article we describe two symptomatic cases with a similar anatomical diagnosis of bifurcation of the main pancreatic duct, one presenting with acute pancreatitis and the other with recurrent pancreatitis. In both cases there was no other explanation. We analysed literature and previous published case reports of this topic.

Case report 1

A 76 year old man was followed for recurrent idiopathic pancreatitis since the last 14 years. He is known with claustrophobia and was chronically treated for arterial hypertension. His first pancreatitis was a necrotising pancreatitis of the neck area at the age of 62. Following episodes were non-necrotising and mostly mild. There was no alcohol, nicotine or medication abuse. His laboratory results showed no hypercalcemia, hypertriglyceridemia or arguments for auto-immune pancreatitis. There was no cholecystolithiasis on imaging and CT scan of the abdomen demonstrated no suspect

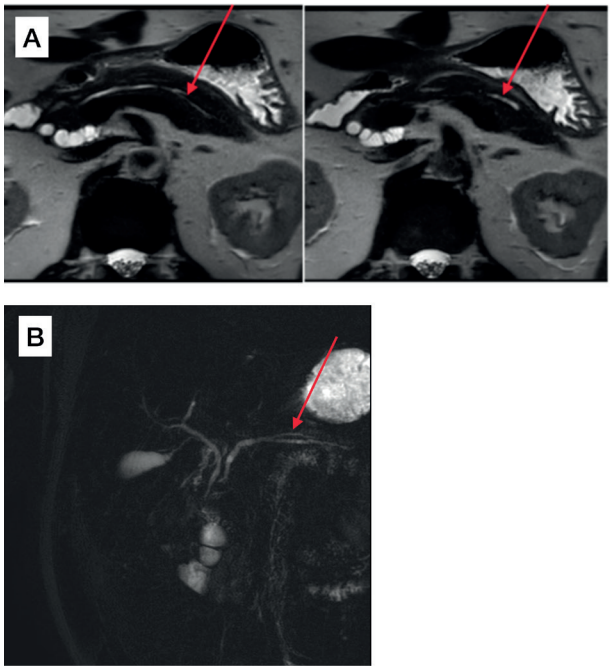

Figure 1. — A : MRI images case 1. MRI of the pancreas with MRCP images of the patient form the first case report shows a bifurcation of the main pancreatic duct, starting in the body of the pancreas and further visible until the tail. One ductal system (the most ventral duct, indicated with the red arrow), shows signs of previously occurred pancreatitis with a broad and irregular course, the other ductal system is normal. B : MRI image case 2. MRI of the pancreas with MRCP images of the patient form the first case report shows a bifurcation of the pancreatic duct (indicated with the red arrow) starting in the body of the pancreas and further visible until the tail. One ductal system (the most ventral duct) shows signs of previously occurred pancreatitis with a broad and irregular course, the other ductal system is normal. The Wirsung duct is also slightly dilated.

pancreatic lesions. Genetic analysis was negative for hereditary pancreatitis (no mutations were found in the *CFTR1* and *PRSS1* gene). He underwent cholecystectomy for his recurrent pancreatitis of unknown aetiology at

Correspondence to : Louise Krott, University hospital Ghent, Department of Gastroenterology. C. Heymanslaan 10, 9000 Ghent, Belgium.
E-mail : Louise.krott@ugent.be

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the age of 63, pathologic examination of the gallbladder showed microlithiasis. Endoscopic ultrasound at that time documented no pancreatic anomalies. Despite the cholecystectomy, recurrent mild episodes of pancreatitis occurred. He refused Magnetic Resonance Imaging because of his claustrophobia. At the age of 76 he was convinced to perform MRI of his pancreas with MRCP imaging, this showed a bifurcation of the main pancreatic duct, starting in the body of the pancreas and further visible until the tail. One ductal system (the most ventral duct) showed signs of previously occurred pancreatitis with a broad and irregular course, the other ductal system was completely normal.(Figure 1A)

Case report 2

A 56 year old man was hospitalized for an acute pancreatitis of unknown aetiology. His medical history showed right partial nefrectomy for renal cell carcinoma. Aetiology for his pancreatitis was unknown, there was no alcohol, nicotine or medication abuse. His laboratory results showed no hypercalcemia, hypertriglyceridemia or arguments for auto-immune pancreatitis. Biopsy of the papilla region was negative for IgG4 related disease. There was no cholecystolithiasis on imaging. However CT scan of the abdomen showed suspicion of a small mass at the pancreas head region and mild dilated pancreatic duct. Because of CT scan findings, further imaging was performed. Additional endoscopic ultrasound did not visualise a pancreatic mass, the pancreatic duct was indeed slightly dilated from the head of the pancreas and his further course. MRI at that time showed a mild pancreatitis of the head region and confirmed the mild dilated pancreatic duct, no further anomaly was described. Surveillance MRI 4 months later revealed a bifurcation of the pancreatic duct starting in the body and continuing to the tail of the pancreas. One ductal system (the most ventral duct) showed signs of previously occurred pancreatitis with a broad and irregular course, the other ductal system was completely normal. In this case the common pancreatic duct was also slightly dilated.(Figure 1B)

Discussion

Background

Acute pancreatitis is an important gastrointestinal cause of hospitalization, the annual incidence ranges from 13 to 45 per 100 000 (1). Alcohol abuse and gallstones remain the most important cause of pancreatitis. Approximately 15 to 25 percent of patients with recurrent pancreatitis are idiopathic, meaning there is no known cause found after extensive work out (2). Occasionally pancreatitis is caused by an anatomic variant of the pancreas (3). Embryologically the pancreas develops by fusion of the dorsal and ventral pancreatic parenchymal buds. Both buds first develop independently, each with a dominant

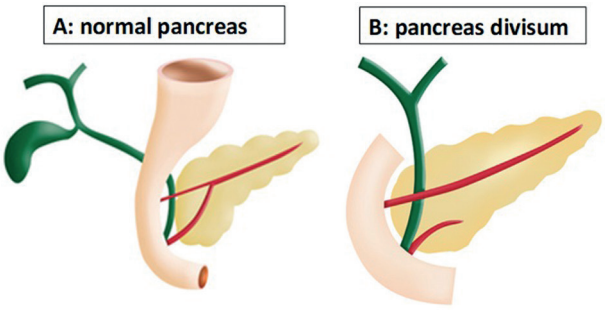

Figure 2. — A : normal pancreas, shows the anatomy of a normal pancreas. The pancreas is predominantly drained through the ventral duct, which joins with the common bile duct at the level of major papilla and the dorsal duct drains at level of minor papilla (29). B : pancreas divisum, shows the anatomy of a complete pancreas divisum where the ventral duct drains through the major papilla and the dorsal duct through the minor papilla (29).

duct draining in the duodenal lumen. The fusion of both pancreas buds takes place in the seventh gestational week. The dorsal pancreas accounts for most pancreatic tissue, including the anterior head, body and tail. The ventral pancreas forms the posterior head and uncinate process (4,3).

There are several anatomical anomalies of the pancreas, the most common is pancreas divisum which occurs in approximately 10 percent of autopsy specimens. In pancreas divisum there is failure of fusion of the dorsal and ventral pancreatic ducts. This results in a ventral (Wirsung) duct emptying at the major papilla and a dorsal (Santorini) duct emptying at the minor papilla. It is generally considered a benign condition, however case reports of symptomatic patients are published (3,5). (Figure 2).

Other anomalies include migration variants such as pancreas annulare and ectopic pancreas, and duplication variants, further divided in number and form variants (6,7). Bifurcation of the pancreatic duct, belonging to the duplication number variants, is a very rare anomaly of the pancreas. Little is known about the prevalence, Uomo et al. analysed the indication of pancreaticography in 485 cases, of these, 13 (2.7%) showed bifurcation of the main pancreatic duct (6). Unlike pancreas divisum, the separated ducts are generally laid from the pancreatic tail to neck, join at the pancreas head and drain via the major papilla, however less extended variants are possible.
Our cases both show bifurcation of the main pancreatic duct in the body to the tail of the pancreas and are both symptomatic. Both cases had one aberrant duct as a consequence of pancreatitis and one normal duct, notably in both cases the abnormal duct was the most ventral duct of the bifurcated pancreatic duct.

Literature on this topic is limited and the clinical relevance is not well established, likewise embryological development is unknown.

Literature study

Literature on the clinical impact of bifurcation of the pancreatic duct is limited and only consists of case reports.

Some authors use different terms for bifurcation of the pancreatic duct, such as duplex pancreas, pancreas bifidum, bifid pancreatic duct and double pancreatic ducts (5,8-10). Likewise 'duplicated pancreatic duct' is often used to describe a bifurcated pancreatic duct, this however could cause misunderstanding (11-13). As mentioned above, bifurcation of the pancreatic duct, belongs to the group of duplication variants, namely the number variants (6,7).

We found 22 relevant articles containing 26 case reports, mostly full text was found, for 8 articles, information is based only on availability of the abstract. Of these 26 case reports, 12 cases were considered asymptomatic and were found incidentally (5,9,11,14-19). 14 cases were symptomatic with signs of acute, chronic or recurrent pancreatitis (8,10,12,13,20-28).

Diagnosing this anatomical variant is often difficult. In our cases, endoscopic ultrasound failed to show the anomaly in both cases and, in the second case, the first MRI was also negative. High quality ERCP and MRCP give the most detailed information of the pancreatic ductal system. In 7 cases, diagnosis was made because of presumed malignancy (of which 2 cases were symptomatic (10, 25) and 5 were asymptomatic (5,11,14,15,18). In 5 cases, diagnosis was made during surgery. In 1 case surgery was performed for complications of chronic pancreatitis (24). In the other cases surgery was performed because of presumed malignancy, of which 2 cases were asymptomatic (5,18) and 2 were symptomatic ; Vasiliadis et al. reports a case with a history of recurrent pancreatitis and Ball et al. describes a case of a patient with presumed alcohol induced pancreatitis (10,25). The authors of these case reports highlight the complexity of the surgery and the importance of the diagnosis. Preferably the diagnosis is made prior to surgery to avoid complications (5,10,18,25).

Some cases not only show bifurcated pancreatic duct, but also show parenchymal bifurcation, which is even rarer than bifurcated pancreatic duct alone. In some of the cases the presence of parenchymal bifurcation is not mentioned. Again different terms are used ; pancreas bifidum, bifid pancreas, the duplicated pancreas and "fish tail pancreas" when the pancreas tail is completely bifurcated (17,25-28).

The location of the bifurcation of the pancreatic duct can vary. In some cases the bifurcation is limited to the head, the body or the tail and in some a total bifurcation is seen. In a few of the found articles it's not clear where the bifurcation is located (15,16,21-23). Interestingly whether the cases are symptomatic or asymptomatic does not seem to be associated with the location of the bifurcation. For example, 3 case reports are similar to our cases with ductal bifurcation located in the body and the tail, of which 2 are asymptomatic (14,17), and one had acute severe pancreatitis (20).

In our cases there is always one aberrant and one normal duct, the aberrant bifurcated ductal system shows signs of previously occurred pancreatitis. This finding is also mentioned in three other case reports. Kawakubo et al. presents a case of pancreatic duct bifurcation in the head of the pancreas with an aberrant upper and normal lower Wirsung (with patent Santorini) (13). Koyasu et al. and Weiner et al. both present a case with a bifurcation of the pancreatic duct in the tail, including bifurcation of the parenchyma, their cases both present with acute pancreatitis in a single limb (26, 28). This finding could suggest pathophysiology. Some authors of the symptomatic cases claim that the presence of a bifurcated pancreatic duct can alter the flow characteristics of pancreatic juice in the pancreatic ducts or results in more vulnerable smaller ducts, thus increasing the risk of pancreatitis (10, 21, 26). In our 2 cases the location of the pancreatitis (neck and head region) is considered downstream or distal from the bifurcated pancreatic duct. No clear explanation can be provided or found in other case reports.

Very little is known about the management of bifurcated pancreatic duct and in asymptomatic cases no therapy seems needed. We found 3 cases in which endotherapy is performed because of symptoms. Agha et al. used sphincterotomy for a patient with complete bifurcated pancreatic duct with joining ducts near the papilla major, the patient, suffering from recurrent abdominal pain, was asymptomatic during 1 year follow-up (8). Kawakubo et al. performed sphincterotomy and stone removal for a bifurcated Wirsung, the clinical impact is not mentioned (13). In another case report sphincterotomy was performed and resulted in clinical improvement of a mild pancreatitis (21).

Conclusion

The clinical relevance of a bifurcated pancreatic duct remains unclear. Since the diagnosis is often difficult, the incidence is probably underestimated (both in asymptomatic and in symptomatic patients). Because of the limited number of published cases it is difficult to make clear conclusions about the causality between a bifurcation of the pancreatic duct and acute or recurrent pancreatitis. However more than half of the found published cases, including our two cases, were symptomatic. This indicates that a bifurcated pancreatic duct might be a possible cause of pancreatitis, but the underlying pathophysiology is not clear and a large group of patients remains asymptomatic. Additionaly patient related factors could influence the development of pancreatitis in these patients. For the same reason the management of patients with bifurcated pancreatic duct is not yet established. The importance of the diagnosis is

highlighted in cases where surgery is performed, as the surgery in these patients is complex and with high risk for post-operative complications.

In summary, the question whether this anatomical anomaly is an accidental finding in patients with idiopathic pancreatitis is unsolved so far. For this reason it is important to further report cases to generate more knowledge about the clinical relevance of this rare condition.

Conflict of interest

The authors declare that they have no competing interest.

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