

CASE REPORT

Role of Endoscopic Ultrasound in the Diagnosis of Agenesis of the Dorsal Pancreas

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ABSTRACT

Context Complete agenesis of the dorsal pancreas is an unusual congenital anomaly. Nowadays, the diagnosis is based on four imaging studies: transabdominal ultrasonography, computed tomography, magnetic resonance cholangiopancreatography and, the gold-standard, endoscopic retrograde cholangiopancreatography. Sometimes the ability of these studies are limited to distinguishing agenesis of the dorsal pancreas from other congenital abnormalities. Endoscopic ultrasound is a minimally invasive technique which permits us to obtain high resolution images of the pancreatic parenchyma and ductal system.

Case report We report the case of a 23-year-old woman with recurrent episodes of acute pancreatitis and non-conclusive classic imaging studies. Complete agenesis of the dorsal pancreas was demonstrated by endoscopic ultrasound.

Conclusion Endoscopic ultrasound may be useful in the diagnosis of agenesis of the dorsal pancreas.

INTRODUCTION

Complete agenesis of the dorsal pancreas is an unusual congenital anomaly. Nowadays

the diagnosis is based on four imaging studies: transabdominal ultrasonography (US), computed tomography (CT), magnetic resonance cholangiopancreatography (MRCP) and, the gold-standard, endoscopic retrograde cholangiopancreatography (ERCP) [1]. Sometimes the ability of these studies is limited to distinguishing agenesis of the dorsal pancreas from another congenital abnormalities [2]. Endoscopic ultrasound (EUS) is a new technique which permits us to obtain high resolution images of the pancreatic parenchyma and ductal system [3, 4]. We present a case of complete agenesis of the dorsal pancreas, with non-conclusive classic imaging studies, in which EUS played an important role in the diagnosis.

CASE REPORT

A 23-year-old woman, without any history of diabetes, was admitted to our hospital to assess recurrent episodes of acute pancreatitis. An episode of severe biliary acute pancreatitis with a complicated pancreatic pseudocyst at the head of the pancreas had been reported 4 years earlier. Cholecystectomy, cystogastrostomy and cystoduodenostomy were performed after the episode. One year later, the patient developed a new episode of acute pancreatitis. An abdominal CT revealed a pancreatic pseudocyst (2.3 cm) at the head of the pancreas (Figure 1). An ERCP showed the main pancreatic duct with a length of 30 mm,



Figure 1. Abdominal CT showing pancreatic pseudocysts at the head of the pancreas.

sharp-ended and with multiple normal side branches (Figure 2). With these findings the original diagnosis was pancreas divisum. In a subsequent MRCP, the head of the pancreas was prominent having a short main pancreatic duct without any evidence of a dorsal pancreas (Figure 3). Different etiologies such as toxins and metabolic or autoimmune disorders were excluded. Intermittent epigastric pain with normal values of serum amylase and lipase had been present for more than 2 years. A transabdominal US showed a cyst (2 cm) at the head of the pancreas. A secretin-stimulated MRCP demonstrated a pseudocyst at the head of the pancreas and a short, irregular main pancreatic duct. The accessory pancreatic duct and the pancreatic dorsal parenchyma were not visible. A new ERCP showed a normal common bile duct

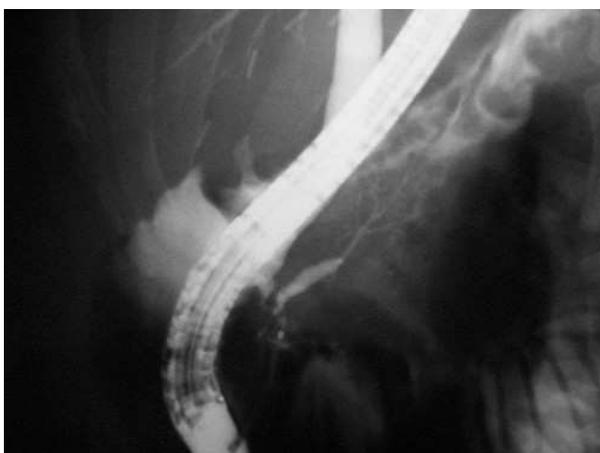


Figure 2. Endoscopic retrograde cholangiopancreatography showing a short main pancreatic duct with sharp-ended and multiple normal side branches.

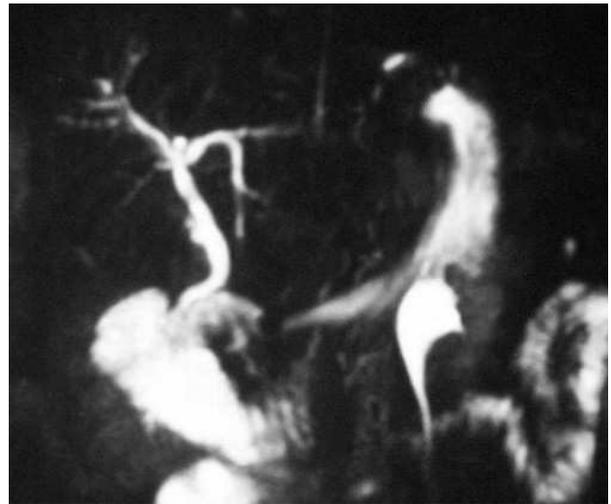


Figure 3. Magnetic resonance cholangiopancreatography showing a prominent short main pancreatic duct.

and a short main pancreatic duct with multiple side branches. A small prominence on the mucosa was detected in the descending part of the duodenum, but cannulation of this structure was impossible. An oral glucose tolerance test and Van de Kamer test (5.9 g/24h; reference range: 1-7 g/24h) were within normal limits but the fecal elastase concentration was low (103 U/g; reference range: 0-200 U/g). Finally, an EUS was performed. It showed the ventral pancreas and a main pancreatic duct of normal caliber, increased duct wall echogenicity and cystic lesion (1.6x2.2 cm). EUS did not show the accessory pancreatic duct and the dorsal pancreas. The splenic vessels were in contact with the posterior wall of the stomach (Figure 4).

The diagnosis of our patient was controversial because of the small prominence on the mucosa of the duodenum. An EUS and the review of the original imaging studies permitted us to conclude that the patient presented agenesis of the dorsal pancreas.

DISCUSSION

The pancreas is formed by two separate anlagen, each from a ventral and a dorsal endodermal bud. The ventral bud forms the lower portion of the pancreatic head and the uncinata process while the dorsal bud forms

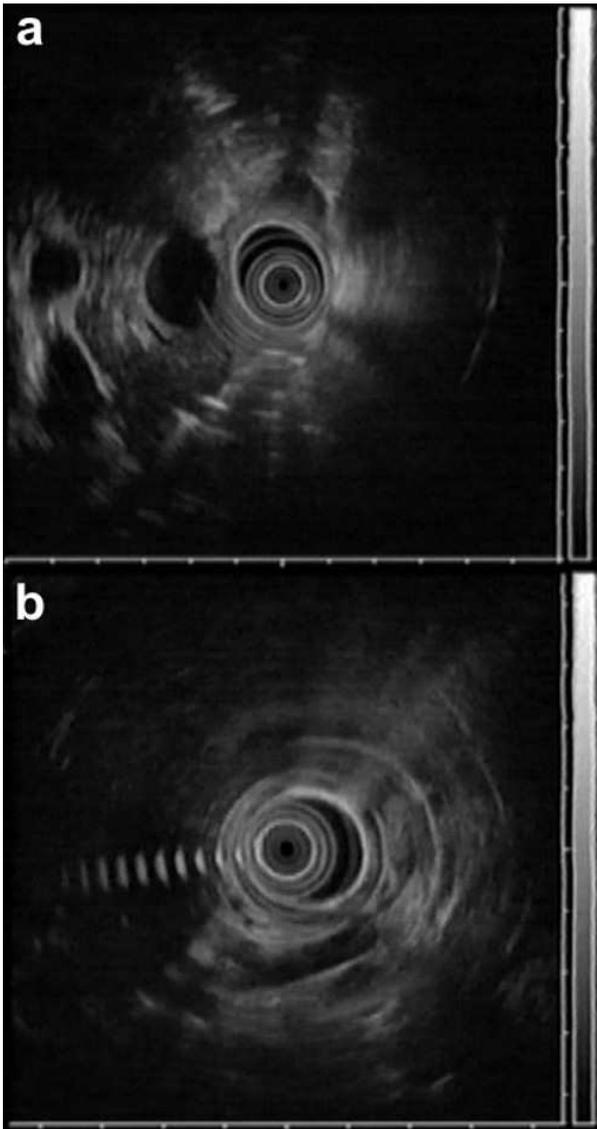


Figure 4. Endoscopic ultrasound. **a.** Ventral pancreas and main pancreatic duct having a normal caliber, increased duct wall echogenicity and a cystic lesion (1.6x2.2 cm). **b.** Absence of the accessory pancreatic duct and the dorsal pancreas. The splenic vessels are in contact with the posterior wall of the stomach.

the upper head, neck, body and tail of the pancreas. Each anlagen possesses its own duct system. The pancreas presents a complicated embryogenesis between the 5th and the 7th week of gestation [5, 6]. At the 6-7th week of gestation, the ventral pancreas fuses with the dorsal pancreas. During the fusion, the ventral and the dorsal ducts form the main pancreatic duct. The accessory pancreatic duct is formed from the portion of the dorsal bud which gives rise to the upper pancreatic head. A disorder during the complicated embryologic-

al development of the pancreas can lead to congenital abnormalities.

Complete agenesis of the pancreas and agenesis of the ventral pancreas are unknown congenital abnormalities because complete agenesis of the pancreas is incompatible with life [7] and the agenesis of the ventral pancreas is extremely rare [1].

Agenesis of the dorsal pancreas is an uncommon congenital abnormality originating from a defect in early embryogenesis which may also be associated with other congenital abnormalities [8, 9]. Primary dysgenesis and ischemic injury to the development of the pancreas could play a role in the etiology [10]. Family cases have been reported in the literature, but the genetic transmission remains unclear [11]. Agenesis of the dorsal pancreas may be partial or complete; partial agenesis is more frequent than complete agenesis. In complete agenesis, the minor papilla, accessory pancreatic duct, body and tail of the pancreas are absent. In partial agenesis, the papilla minor with a remnant of the accessory pancreatic duct and the body of the pancreas are present [12].

We have reviewed 16 recent case reports of complete agenesis of the dorsal pancreas [1, 2, 11, 12, 13, 14, 15, 16, 17, 18, 19, 20, 21].



Figure 5. Endoscopic ultrasound in a healthy person: the body and the tail of the pancreas are visualized between the posterior wall of the stomach and the splenic vessels.

To assess the clinical presentation we excluded 2 cases with several concomitant pathologies (tumor in the head of the pancreas and diffuse peritoneal carcinosis of the adenocarcinoma) [19, 20]. In the other 14 cases, 6 men and 8 women with a mean age of 42 years, the most frequent clinical events were abdominal pain (13 cases, 92.9%) and the development of diabetes mellitus (6 cases, 42.9%). Acute pancreatitis episodes, present in our patient, were described in 7 cases (50.0%). Some authors suggest that abdominal pain is more common in patients with partial agenesis and diabetes mellitus is more common in patients with complete agenesis of dorsal pancreas [12, 15]. The relationship between agenesis of the dorsal pancreas and exocrine pancreatic insufficiency remains unclear; only one case has been reported [13]. The fecal elastase concentration of our patient was low, but there were no other findings at clinical presentation, function tests or diagnostic imaging to confirm the diagnosis of exocrine pancreatic insufficiency.

The diagnosis of agenesis of the dorsal pancreas requires a high index of suspicion in addition to imaging studies. Before the use of modern diagnostic imaging techniques, the diagnosis was made by autopsy or laparotomy [11]. Nowadays, transabdominal US, CT, MRCP and ERCP allow the diagnosis of agenesis of the dorsal pancreas. However, there are some patients, such as ours, in which the classical imaging techniques are not able to differentiate between agenesis of the dorsal pancreas and other congenital abnormalities. Transabdominal US in our patient did not reveal any diagnostically suspicious findings. Congenital abnormalities can only occasionally be identified by transabdominal US because the bowel gas obscures the pancreas. However, a hyperechoic line of demarcation between the head of the pancreas and the retroperitoneal fat with the presence of a small hypoechoic mass in the pancreas, just ventral to the portal confluence, has been reported in agenesis of the dorsal pancreas [14, 15]. Contrast-enhanced CT has frequently been used with ERCP to evaluate

pancreatic abnormalities [13]. In complete agenesis of the dorsal pancreas, only the pancreatic head is visible [13, 14]. A dorsal pancreatic parenchyma was not detected in our patient at either CT examination. In recent years, MRCP has been used as a non-invasive alternative to ERCP for the evaluation of the pancreatic ductal system and congenital abnormalities [22]. On MRCP, the presence of a short main pancreatic duct in the normal head of a pancreas with the absence of pancreatic dorsal tissue and an accessory pancreatic duct, such as in our patient, are the characteristics of complete agenesis of the dorsal pancreas [1, 15]. The inability of performing biopsies and interventional approaches are the disadvantages of MRCP. ERCP is an invasive technique with an important morphologic assessment of the pancreatic ductal system, and is considered the gold standard for agenesis of the dorsal pancreas [12]. Furthermore, brush cytology and therapeutic intervention are possible during ERCP. Invasivity, dependency on the operator and difficulty in the identification and cannulation of the papilla minor are the disadvantages of ERCP [1]. In our patient, the presence of multiple side branches in the main pancreatic duct and the small prominence in the mucosa detected in the second segment of the duodenum did not allow us to reach a definitive diagnosis.

EUS is a relatively new imaging technique which provides direct visualization of the total pancreatic parenchyma and the ductal system [3]. The role of EUS in the identification of agenesis of the dorsal pancreas has not been evaluated, but it may be as good as ERCP. The head of the pancreas, uncinate process, ampulla and distal pancreatic ducts (main and accessory ducts) can be examined with the EUS positioned in the first and second part of the duodenum [23, 24]. "Stripping" is necessary in order to visualize the body and the tail of the pancreas. "Stripping" is the retraction of the EUS device from the duodenum to the gastric antrum. With the EUS positioned in the gastric antrum, the body and the tail of the pancreas are visualized between the posterior

wall of the stomach and the splenic vessels [4] (Figure 5). Agenesis of the dorsal pancreas may be suspected, as in our patient, if the accessory pancreatic duct and the dorsal pancreas (without any structure between the stomach and the splenic vessels) are not visualized. The most crucial differential diagnosis in the agenesis of the dorsal pancreas is a pancreatic carcinoma with proximal atrophy [15]. Electronic linear array EUS has a significant impact on the diagnosis of pancreatic carcinoma, it provides excellent resolution images of the pancreatic parenchyma and the ductal system and the possibility of performing fine-needle aspiration [3, 4].

In summary, complete agenesis of the dorsal pancreas can be mimicked by other congenital abnormalities requiring different clinical management. EUS is a minimally invasive technique which may be useful in the diagnosis of agenesis of the dorsal pancreas.

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Keywords Digestive System Abnormalities; Endosonography; Pancreatitis

Abbreviations CT: computed tomography; ERCP: endoscopic retrograde cholangiopancreatography; EUS: endoscopic ultrasound; MRCP: magnetic resonance cholangiopancreatography; US: ultrasonography

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References

1. Guclu M, Serin E, Ulucan S, Kul K, Ozer B, Gumurdulu Y, et al. Agenesis of the dorsal pancreas in a patient with recurrent acute pancreatitis: case report and review. *Gastrointest Endosc* 2004; 60:472-5. [PMID 15332050]
2. Gold RP. Agenesis and pseudo-agenesis of the dorsal pancreas. *Abdom Imaging* 1993; 18:141-4. [PMID 8439753]
3. Kahl S, Glasbrenner B, Zimmermann S, Malferteiner P. Endoscopic ultrasound in pancreatic diseases. *Dig Dis* 2002; 20:120-6. [PMID 12566614]
4. Palazzo L. Echoendoscopy of the pancreas. *Gastroenterol Hepatol* 2002; 25:26-34. [PMID 11835870]
5. Lankisch PG, Banks PA. General considerations: Embryology. *Pancreatitis*, 1st edition. Berlin-Heidelberg, Germany: Springer-Verlag, 1998:1-14.
6. Tuchmann-Duplessis H, Haegel P. Embryologie: Travaux pratiques et enseignement dirigé. Spanish version, 2nd edition. Barcelona, Spain: Masson, 1982:34-39.
7. Voldsgaard P, Kryger-Baggesen N, Lisse I. Agenesis of pancreas. *Acta Paediatr* 1994; 83:791-3. [PMID 7949819]
8. Soler R, Rodriguez E, Comesana ML, Pombo F, Marini M. Agenesis of the dorsal pancreas with polysplenia syndrome: CT features. *J Comput Assist Tomogr* 1992; 16:921-3. [PMID 1430442]
9. Gurson CT, Tahsinoglu M, Yakacikli S, Ertugrul T. A case of agenesis of the dorsal pancreas with interventricular septal defect in an infant. *Helv Paediatr Acta* 1970; 25:522-6. [PMID 5493568]
10. Sener RN, Alper H. Polysplenia syndrome: a case associated with transhepatic portal vein, short pancreas, and left inferior vena cava with hemiazygous continuation. *Abdom Imaging* 1994; 19:64-6. [PMID 8161909]
11. Wildling R, Schnedl WJ, Reisinger EC, Schreiber F, Lipp RW, Lederer A, Krejs GJ. Agenesis of the dorsal pancreas in a woman with diabetes mellitus and in both of her sons. *Gastroenterology* 1993; 104:1182-6. [PMID 8462806]
12. Schnedl WJ, Reisinger EC, Schreiber F, Pieber TR, Lipp RW, Krejs GJ. Complete and partial agenesis of the dorsal pancreas within one family. *Gastrointest Endosc* 1995; 42:485-7. [PMID 8566643]
13. Klein WA, Dabezies MA, Friedman AC, Caroline DF, Boden GH, Cohen S. Agenesis of dorsal pancreas

in a patient with weight loss and diabetes mellitus. *Dig Dis Sci* 1994; 39:1708-13. [PMID 8050322]

14. Deignan RW, Nizzero A, Malone DE. Case report: agenesis of the dorsal pancreas: a cause of diagnostic error on abdominal sonography. *Clin Radiol* 1996; 51:145-7. [PMID 8631173]

15. Macari M, Giovanniello G, Blair L, Krinsky G. Diagnosis of agenesis of the dorsal pancreas with MR pancreatography. *AJR Am J Roentgenol* 1998; 170:144-6. [PMID 9423620]

16. Terruzzi V, Radaelli F, Spinzi GC, Imperiali G, Minoli G. Congenital short pancreas. Report of a new case observed during the course of a recurrent acute pancreatitis. *Ital J Gastroenterol Hepatol* 1998; 30:199-201. [PMID 9675659]

17. Oldenburg B, van Leeuwen MS, van Berge Henegouwen GP, Koningsberger JC. Pancreatitis and agenesis of the dorsal pancreas. *Eur J Gastroenterol Hepatol* 1998; 10:887-9. [PMID 9831413]

18. Fukuoka K, Ajiki T, Yamamoto M, Fujiwara H, Onoyama H, Fujita T, et al. Complete agenesis of the dorsal pancreas. *J Hepatobiliary Pancreat Surg* 1999; 6:94-7. [PMID 10436244]

19. Nakamura Y, Egami K, Maeda S, Hosone M, Onda M. Solid and papillary tumor of the pancreas complicating agenesis of the dorsal pancreas. *J Hepatobiliary Pancreat Surg* 2001; 8:485-9. [PMID 11702261]

20. Nonent M, Linard J, Leveque E, Larroche P, Bobeuf J, Senecail B. Dorsal pancreas agenesis: computed tomography appearance with three-dimensional volume rendering reconstruction. *Surg Radiol Anat* 2003; 25:161-3. [PMID 12851784]

21. Rakesh K, Choung OW, Reddy DN. Agenesis of the dorsal pancreas (ADP) and pancreatitis - is there an association? *Indian J Gastroenterol*. 2006; 25:35-6. [PMID 16567893]

22. Fulcher AS, Turner MA. MR pancreatography: a useful tool for evaluating pancreatic disorders. *Radiographics* 1999; 19:5-24. [PMID 9925389]

23. Savides TJ, Gress FG, Zaidi SA, Ikenberry SO, Hawes RH. Detection of embryologic ventral pancreatic parenchyma with endoscopic ultrasound. *Gastrointest Endosc* 1996; 43:14-9. [PMID 8903811]

24. Sahai AV. EUS and chronic pancreatitis. *Gastrointest Endosc* 2002; 56(4 Suppl):S76-81. [PMID 12297754]