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MR in complete dorsal pancreatic agenesis: Case report and review of literature

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Abstract

The morphogenesis of the pancreas is a complex process having a very low frequency of anatomic variation. The congenital anomalies are rare. Complete pancreatic and ventral pancreatic agenesis are incompatible with life. Dorsal pancreatic agenesis is exceedingly rare with less than 100 cases reported in the world literature. Patients with this anomaly may be asymptomatic or may present with abdominal pain, hyperglycemia, diabetes mellitus, and acute or chronic pancreatitis. Such anomalies are rarely reported; therefore, clinical awareness of agenesis of the dorsal pancreas as a cause of these symptoms can expand the differential diagnosis and improve patient management.

Key words: Dorsal pancreatic agenesis; embryogenesis; hyperglycemia

Introduction

Complete pancreatic agenesis or ventral pancreatic agenesis is incompatible with life.[1] Agenesis of the dorsal pancreas is very rare and described in literature as case reports only. It describes a congenital malformation in which either the entire dorsal pancreas or a part of it fails to develop (complete agenesis or partial agenesis, respectively). The patient is usually asymptomatic but may present with varied conditions. The diagnosis of complete dorsal pancreatic agenesis is inconclusive without demonstration of the absence of the dorsal pancreatic duct. USG, computed tomography (CT), and magnetic resonance (MR) imaging are only suggestive; however, endoscopic ultrasound (EUS), endoscopic retrograde cholangiopancreatography (ERCP), and magnetic resonance cholangiopancreatography (MRCP) give detailed pancreatic ductal anatomy for diagnostic confirmation.

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Case Report

We report a 42-year-old male who presented with the complaint of having repeated epigastric pain after heavy meals for 1 year. On USG, he had cholelithiasis, but there was no evidence of cholecystitis or cholangitis. The common bile duct could not be seen in its distal-third due to overlying gut air. Rest of the USG findings were reported to be normal, but images were not available. On upper gastrointestinal endoscopy, no peptic ulcer disease was found. He underwent MRI and MRCP (Avanto 1.5T, Siemens) to exclude associated choledocholithiasis before undergoing cholecystectomy. On MRI, the neck, body, and tail of the pancreas were not visualized [Figures 1 and 2A-F]. On MRCP, the accessory duct could not be visualized [Figure 3]. These findings were compatible with complete dorsal pancreatic agenesis. Routine blood analysis including the fasting blood sugar was normal. The patient underwent cholecystectomy. However, his symptoms persisted. Thereafter, his diet was modified to low-fat diet with frequent light meals. The frequency of pain reduced drastically following this dietary modification. Hence, the cause of abdominal pain was dorsal pancreatic agenesis rather than gallstones and

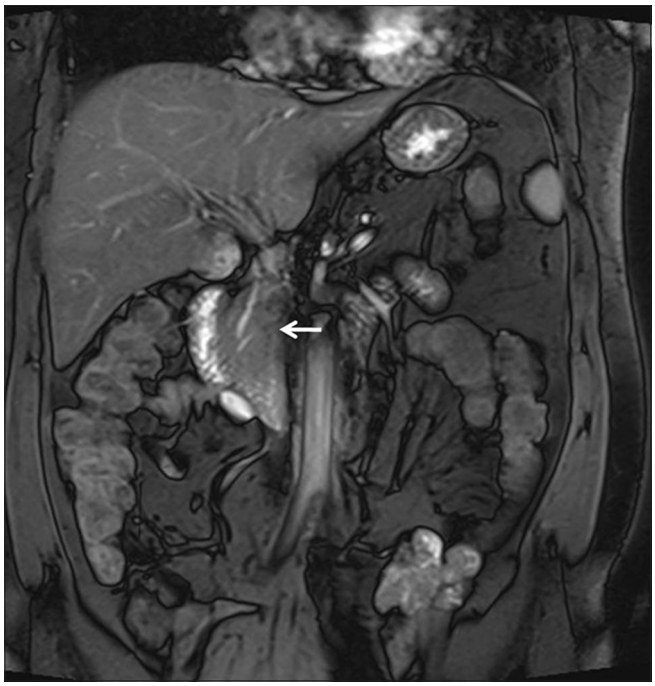

Figure 1: Coronal image (TR-4.05, TE-1.72) shows pancreatic head[white arrow]with non-visualization of rest of the pancreas

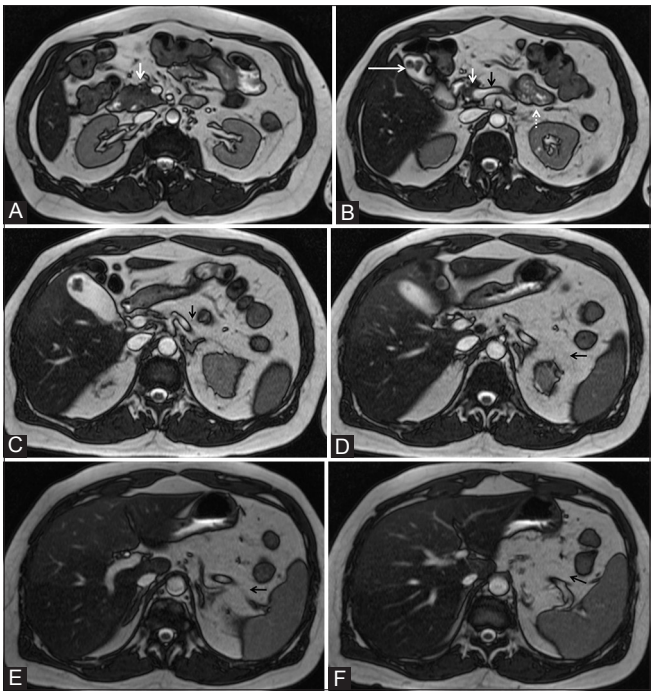

Figure 2 (A-F): (A-F) Tru-fisp (TR-4.05, TE-1.72) serial axial images show pancreatic head and uncinate process which are well developed [white arrow]. The neck, body, and tail of the pancreas are not seen[black arrow]. Loop of small gut is seen in the pancreatic bed anterior to the splenic vein (dependent intestine sign) [dotted white arrow]. Gallstones are visualized in the gall bladder [long white arrow]

the patient was managed accordingly. Since the patient became symptom free, invasive procedures like ERCP were not considered.

Discussion

Complete agenesis of the dorsal pancreas is a rare congenital anomaly thought to be due to abnormal embryogenesis. The embryogenesis of this organ is relatively complex. The pancreas develops from two separate endoderm-lined dorsal and ventral buds of the duodenum. The dorsal bud forms upper part of the head, neck, body, and tail of the pancreas, with its drainage through the accessory duct of Santorini and minor papilla. The ventral bud forms the major part of the head and uncinate process. During the 7th week of gestation, the ventral bud rotates dorsally around the duodenum to fuse with the dorsal bud and form the mature gland.[2] [Figure 4A-D] In complete dorsal pancreatic agenesis, the structures derived from the dorsal bud are undeveloped, that is, the neck, body, and tail of the pancreas, duct of Santorini, and the minor duodenal papilla are not formed. In partial agenesis, pancreatic body is of variable size while remnant of the duct of Santorini and minor duodenal papilla are seen.[1]

The first case of dorsal pancreatic agenesis was reported as an autopsy finding in 1911.[3] Since then, less than 100 cases have been reported in the literature.[4] The condition may be sporadic or may show autosomal dominant or X-linked dominant inheritance. It has been reported to occur with very rare conditions including heterotaxy[2] and polysplenia[5] syndrome. The possible explanation for this is the close proximity of the developing pancreas with spleen in the dorsal mesogastrium.[6] It is also associated with congenital heart defects like septal defects, tetralogy of Fallot, or pulmonary artery stenosis.[7] The exact mechanism and etiology of dorsal pancreatic agenesis are unknown. Recently, alterations in some signaling pathway (retinoic acid and hedgehog) have been shown to play a role. These signaling pathways have also been implicated in the pathogenesis of pancreatic ductal adenocarcinoma.[8]

This disorder is usually asymptomatic, but patient may present with abdominal pain, hyperglycemia, diabetes mellitus, and acute or chronic pancreatitis. The commonest symptom is pain abdomen which is seen in 92.9% of the cases. It is usually localized to epigastrium and aggravated following meals.[4] The possible explanation for this could be lack of papillary muscles.[9] Acute and chronic pancreatitis may also be seen, the causative agents being sphincter of Oddi dysfunction, compensatory enzyme hypersecretion, hypertrophy of the ventral gland, and higher pancreatic duct pressures.[9] Hyperglycemia is seen in approximately 50% of the affected individuals.[3] It may be due to loss of islet cell mass which is predominantly seen in the body and tail of the pancreas.

On imaging, this disorder needs to be differentiated from pancreatic carcinoma with proximal atrophy, pancreatic

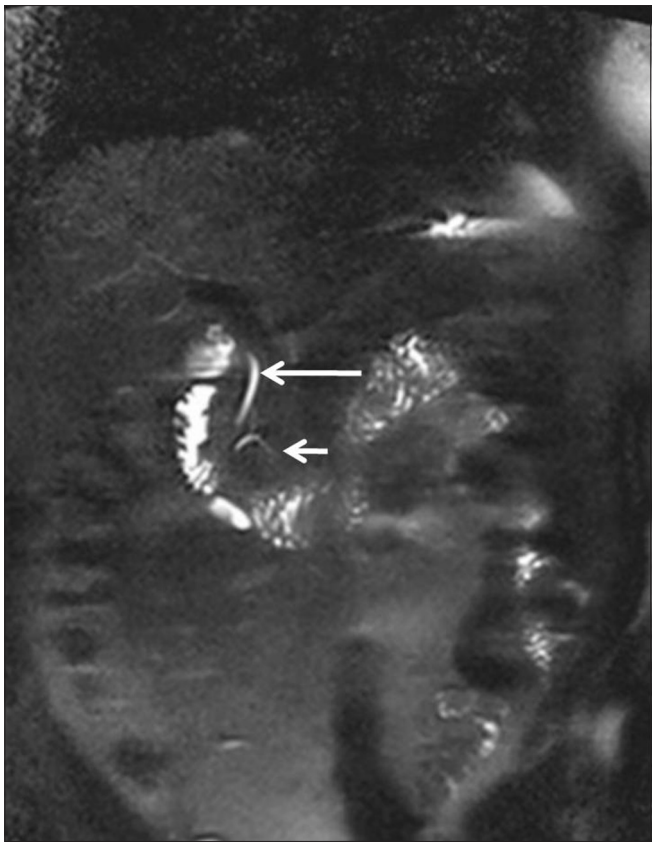

Figure 3: MRCP source image shows absent dorsal duct of Santorini. Common bile duct [long white arrow] and ventral duct of Wirsung are seen clearly [short white arrow]

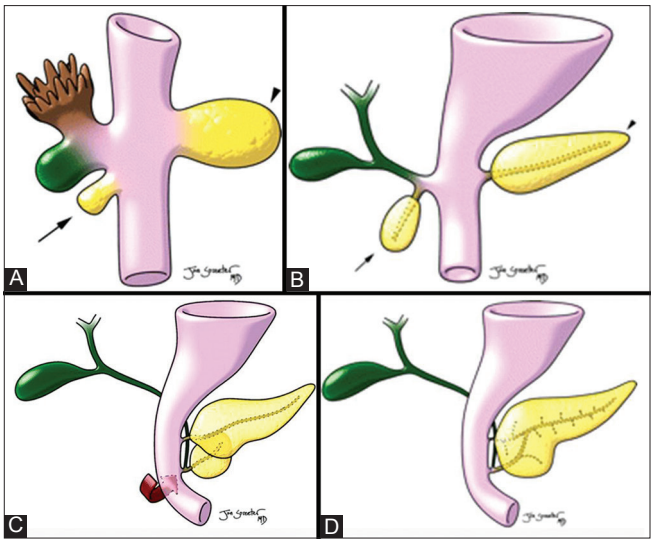

Figure 4 (A-D): Drawings illustrate the normal embryologic development of the pancreas. The ventral pancreatic bud (arrow in A and B) arise from the hepatic diverticulum. The dorsal pancreatic bud (arrowhead in A and B) arises from the dorsal mesogastrium. (C) After clockwise rotation of the ventral bud around the caudal part of the foregut, there is fusion of the dorsal pancreas (located anterior) and ventral pancreas (located posterior). (D) Finally, the ventral and dorsal pancreatic ducts fuse, and the pancreas is predominantly drained through the ventral duct.
Source: Figure has been taken from reference 2 with prior permission from RSNA

divisum, autodigestion from chronic pancreatitis, pancreatic masses, distal pancreatic lipomatosis, and pseudoagenesis. In pseudoagenesis of the pancreas, the pancreatic body and tail are atrophic due to repeated attacks of pancreatitis and replaced by fat. So, it simulates dorsal pancreatic agenesis. The differentiating point is absent or short dorsal duct of Santorini in dorsal pancreatic agenesis while it is demonstrable in pseudoagenesis and lipomatosis.[4]

USG is the first modality, but sometimes pancreas is not adequately visualized due to morbid obesity or excessive overlying bowel gases. CT depicts parenchymal abnormality very well. It shows the deficient part of the pancreas and the stomach or gut loops lying in the pancreatic bed anterior to the splenic vein, called the dependent stomach or dependent intestine signs.[4] But with CT imaging, the information regarding ductal anatomy is limited without which the diagnosis is questionable. Proper description of the pancreatic ductal anatomy is mandatory for diagnostic confirmation. Therefore, evaluation with MRCP or ERCP is required. MRI including MRCP is a noninvasive method which can evaluate the pancreatic ductal anatomy clearly and helps in diagnostic confirmation. CT analogues of dependent stomach or dependent intestine sign can be seen on MRI. Pancreatic head and collapsed bowel loops in the distal pancreatic bed may have similar signal on T1W images, which can be mistaken as the distal body and tail of the pancreas in the absence of intervening gap. So, T2W should be used for the interpretation. ERCP has superior resolution in delineating ductal anatomy. However, it is operator dependent and requires catheterization of the minor duodenal papilla. So, it being an invasive procedure and not devoid of morbidity risk,[1] should be performed if the radiological diagnosis is in doubt.

Keeping this entity in mind, the patient could be managed with gastrointestinal decompression or dietary modifications like total parenteral nutrition and low-fat diet. Further, lengthy and expensive investigations and the mental stress which the patient undergoes in such procedures can be avoided. However, there is an apparent increased risk of developing pancreatic cancer in these patients.[10] The definitive treatment in such cases is total pancreatectomy. But the patient has to suffer the consequences of radical surgery – diabetes mellitus with insulin therapy and exocrine pancreatic insufficiency.[10] Hence, whether this association of dorsal pancreatic agenesis with ductal adenocarcinoma should change the management of the patient from conservative to surgical intervention is still a clinical dilemma.

Conclusion

Clinical awareness of this rare condition is a must for the better understanding of its association with various syndromes. Our case is unique not only because of its rarity but also because of its clinical implication for proper patient management.

References

1. Pasaoglu L, Vural M, Hatipoglu HG, Terekliogl G, Koparal S. Agenesis of the dorsal pancreas. World J Gastroenterol 2008;14:2915-6.
2. Mortelé KJ, Rocha TC, Streeter JL, Taylor AJ. Multimodality Imaging of Pancreatic and Biliary Congenital Anomalies. Radiographics 2006:26;715-31.
3. Schnedl WJ, Piswanger-Soelkner C, Wallner SJ, Krause R, LippRW. Agenesis of the dorsal pancreas. World J Gastroenterol 2009;15:376-7.
4. Mohapatra M, Mishra S, Dalai PC, Acharya SD, Nahak B, Ibrarullah M, *et al*. Imaging Findings in Agenesis of the Dorsal Pancreas. Report of Three Cases. JOP 2012;10:108-14.
5. Schnedl WJ, Piswanger-Soelkner C, Wallner SJ, Reittner P, Krause R, Lipp EW, *et al*. Agenesis of the dorsal pancreas and associated diseases. Dig Dis Sci 2009;54:481-7.
6. Low JP, Williams D, Chaganti JR. Polysplenia syndrome with agenesis of the dorsal pancreas and preduodenal portal vein presenting with obstructive jaundice: A case report and literature review. Br J Radiol 2011;84:e219-22.
7. Shimodaira M, Kumagai N, Sorimachi E, Hara M, Honda K. Agenesis of the dorsal pancreas: A rare cause of diabetes. Intern Emerg Med 2012;7:83-4.
8. Rittenhouse DW, Kennedy EP, Mascaro AA, Brumbaugh JL, Stein LH, Rosenberger LH, et al. The Novel Triad of Dorsal Agenesis of the Pancreas with Concurrent Pancreatic Ductal Adenocarcinoma and Nonalcoholic Chronic Calcific Pancreatitis: A Case Series and Review of the Literature. J Gastrointest Surg 2011;15:1643-9.
9. Uygur-Bayramiçli O, Dabak R, Kiliçoglu G, Dolapçioglu C, Oztas D. Dorsal Pancreatic Agenesis. JOP 2007;8:450-2.
10. Dumitraşcu T, Scarlat A, Diaconescu A, Ionescu M. Dorsal pancreas agenesis and ductal adenocarcinoma: Surgical implications of an extremely rare association. Chirurgia 2012;107:389-92

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