Dorsal pancreas agenesis – A case report and review of literature

Yan Ying Nicola Lee*

Department of Radiolgoy, Ng Teng Fong General Hospital, Singapore

***Correspondence:** Yan Ying Nicola Lee, Department of Radiolgoy, Ng Teng Fong General Hospital, Singapore

Nicola_Lee_Yan_Ying@nuhs.edu.sg

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ABSTRACT

Dorsal pancreas agenesis is a rare congenital anomaly that presents unique challenges in the realm of digestive and endocrine function. While the condition is uncommon, understanding its impact on pancreatic function and implementing appropriate medical interventions can significantly improve the quality of life for affected individuals. This is a case of a 74-year-old male patient with diabetes mellitus who presented with a history of loss of weight and loss of appetite.

CASE REPORT

CASE REPORT

A 74-year-old man with a known history of non-ischaemic cardiomyopathy, congenital heart block, diabetes mellitus, and gout presented with a chief complaint of loss of approximately five kilogrammes in weight associated with loss of appetite and generalized fatigue over the course of a month. He claimed to be compliant to his medications and did not notice any haematuria, change in bowel habits, cough, or any acute symptoms.

Full physical examination was unremarkable. Lab results showed elevated creatine kinase (4122 U/L) and total white cell count (14.45 $*10^9$/L) as well as deranged liver function markers (ALT 258 U/L, AST 175 U/L, ALP 129 U/L). He was then admitted for further evaluation and concern for potential infection.

Total white count was on a downward trend, thought to be a self-resolving occult infection since there were no localizing signs or symptoms. Hepatitis screen was negative. The issues of elevated creatine kinase and hepatocellular transaminitis was therefore thought to be secondary to prescribed medication and bloods were to be repeated and medication list reviewed in a few weeks.

Computed tomography (CT) of the thorax, abdomen, and pelvis was performed for the evaluation of loss of weight and loss of appetite. There was no relevant imaging for comparison at the time of the scan. Although there was no overtly suspicious abnormality identified to account for the patient's presenting complaint, it was noted that the pancreatic head and uncinate process appeared unremarkable and the rest of the pancreas appeared to be absent. The left kidney was not visualized. No significant or suspicious liver abnormality was identified. The spleen was unremarkable.

The patient subsequently defaulted appointments for additional studies including magnetic resonance imaging (MRI) scheduled for further evaluation.

DISCUSSION

Etiology & Demographics

Agenesis of the dorsal pancreas is of a congenital aetiology, resulting from embryological failure of formation of the pancreatic body and tail due to absence of regression of the dorsal pancreatic bud.

Agenesis of the dorsal pancreas may be partial or complete. In the case of partial agenesis, the minor papilla, duct of Santorini, and a variably sized pancreatic body remain present. The aforementioned components in addition to the pancreatic neck are absent in complete agenesis of the dorsal pancreas [1].

Agenesis of the dorsal pancreas is an extremely rare condition, with only 53 cases documented according to a PubMed search, and 23 cases reported between 1913 and 2007 [2]. As of 2006, only five cases have been reported in the Republic of Korea [3].

Clinical & Imaging findings

Clinical presentation of dorsal pancreas agenesis can vary among affected individuals. While some patients may be asymptomatic, the vast majority tend to present with nonspecific abdominal pain [4] which is often difficult to distinguish from acute pancreatitis or other abnormalities including duodenal obstruction [1][5].

The absence or underdevelopment of the dorsal pancreas which contains most of the islet cells is thought to contribute to the development of diabetes mellitus [6], a condition often diagnosed in affected individuals [7].

Abdominal pain may be more common when agenesis of the dorsal pancreas is partial, and diabetes may be more common when it is complete [8].

Dorsal pancreas agenesis has also been reported as part of the polysplenia syndrome [9][10]. This was thought to be due to the proximity of the developing pancreas and spleen in the dorsal mesogastrium.

Discovery of the absent dorsal pancreas is usually incidental when imaging studies are obtained for evaluation of abdominal pain. Transabdominal ultrasound may be deployed in the first instance as a cost effective method of evaluation for such nonspecific symptoms, although it is of limited diagnostic value since the pancreas may often be obscured by overlying bowel gas or inference from patient and technical factors. CT is efficient in delineation of the deficient or absent pancreatic parenchyma with preservation of the pancreatic head, as well as any associated findings. Some studies have noted a related enlargement of the pancreatic head [7]. CT is however sometimes limited in evaluation of the detailed anatomy of the pancreatic duct. Endoscopic retrograde cholangiopancreatography (ERCP) is considered the reference standard for evaluation of the pancreatic and biliary ducts, although this method is very much operator dependent and identification and cannulation of the minor papilla can be difficult in addition to the procedure being relatively invasive, with procedural failure rates of up to ten percent [11]. Magnetic resonance cholangiopancreatography (MRCP) therefore serves as a noninvasive method of diagnostic confirmation of the absence of regular pancreatic ductal morphology.

Treatment & Prognosis

Management of dorsal pancreas agenesis focuses on alleviating symptoms and addressing associated complications. Nutritional support, including pancreatic enzyme replacement therapy and dietary modifications, can aid in managing malabsorption issues. Individuals with diabetes may require insulin therapy to regulate blood sugar levels. A multidisciplinary approach involving gastroenterologists, endocrinologists, and nutritionists is often essential to provide comprehensive care.

The prognosis for individuals with dorsal pancreas agenesis depends on the severity of symptoms and the presence of associated complications. With appropriate medical management, many individuals can lead relatively normal lives. Regular monitoring of blood glucose levels and nutritional status is essential to prevent and manage complications effectively.

Differential Diagnoses

In cases where the diagnosis of agenesis of the dorsal pancreas is suggested, it is also necessary to exclude diagnostic considerations including fat replacement of the pancreas and atrophy secondary to infection/inflammation or downstream carcinoma. In the absence of the dorsal pancreas, the pancreatic bed can be occupied by the stomach (dependent stomach sign) or intestine (dependent intestine sign) which abuts the splenic vein [12]. These signs differentiate agenesis from other diagnostic possibilities such as pancreatic lipomatosis or atrophy. It is however important to correlate with any prior history of distal pancreatectomy, as the splenic vein would be absent in this group of postsurgical patients [13].

TEACHING POINT

Agenesis of the dorsal pancreas is a rare congenital anomaly that is usually asymptomatic but may present with related conditions. Accurate recognition of imaging findings and its association with various syndromes is necessary for effective clinical and patient management.

Questions

Question 1

Which structures are absent in complete agenesis of the dorsal pancreas?

Answer choice 1: pancreatic uncinate process
Answer choice 2: minor duct (applies)
Answer choice 3: duct of Santorini (applies)
Answer choice 4: pancreatic body (applies)
Answer choice 5: pancreatic head

Explanation for question 1: [In the case of partial agenesis, the minor papilla, duct of Santorini, and a variably sized pancreatic body remain present.]

Question 2

What is the most common presenting complaint in affected patients?

Answer choice 1: none
Answer choice 2: diabetes
Answer choice 3: pancreatitis
Answer choice 4: abdominal pain (applies)
Answer choice 5: bowel obstruction

Explanation for question 2: [The vast majority tend to present with nonspecific abdominal pain.]

Question 3

What is the embryological origin of the dorsal pancreas?

Answer 1: ventral pancreatic bud
Answer 2: dorsal pancreatic bud (applies)
Answer 3: liver
Answer 4: stomach
Answer 5: gallbladder

Explanation for question 3: [Agenesis of the dorsal pancreas is of a congenital aetiology, resulting from embryological failure of formation of the pancreatic body and tail due to absence of regression of the dorsal pancreatic bud.]

Question 4

Which is the best modality for delineation of pancreatic duct anatomy?

Answer 1: ultrasound

Answer 2: CT

Answer 3: MRCP (applies)

Answer 4: Nuclear medicine

Answer 5: Barium

Explanation for question 4: [Magnetic resonance cholangiopancreatography (MRCP) therefore serves as a noninvasive method of diagnostic confirmation of the absence of regular pancreatic ductal morphology.]

Question 5

What is the management of agenesis of the dorsal pancreas?

Answer 1: depends on associated conditions and complications (applies)

Answer 2: mainly supportive – fluids, antibiotics

Answer 3: pancreatic transplant

Answer 4: regular monitoring of glucose and nutrition (applies)

Answer 5: none

Explanation for question 5: [Management of dorsal pancreas agenesis focuses on alleviating symptoms and addressing associated complications (…) Regular monitoring of blood glucose levels and nutritional status is essential to prevent and manage complications effectively.]

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FIGURES

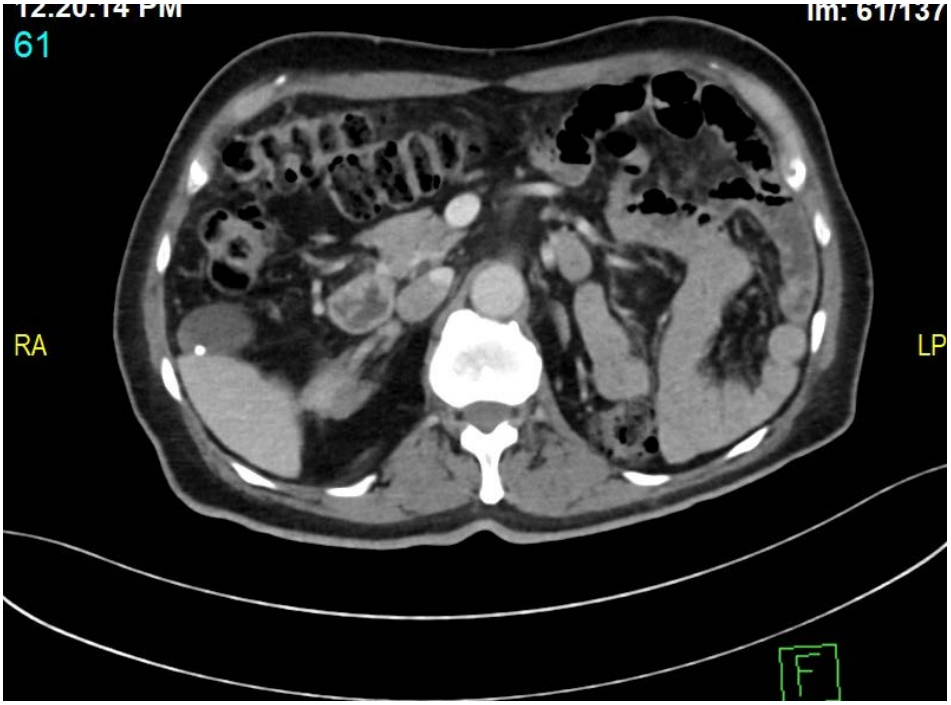

Figure 1: Contrast enhanced CT of the abdomen. Axial view shows the presence of the pancreatic head and uncinate process.

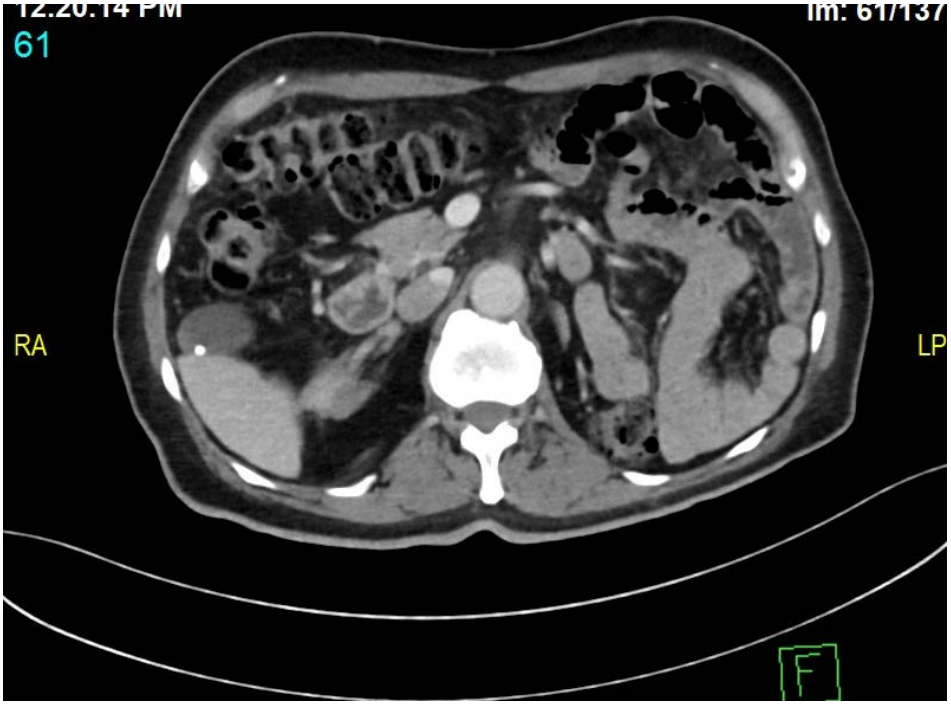

Figure 2: Contrast enhanced CT of the abdomen. Axial view a few slices above the previous image shows the absence of the rest of the pancreas, with the pancreatic bed occupied by surrounding bowel (dependent intestine sign).

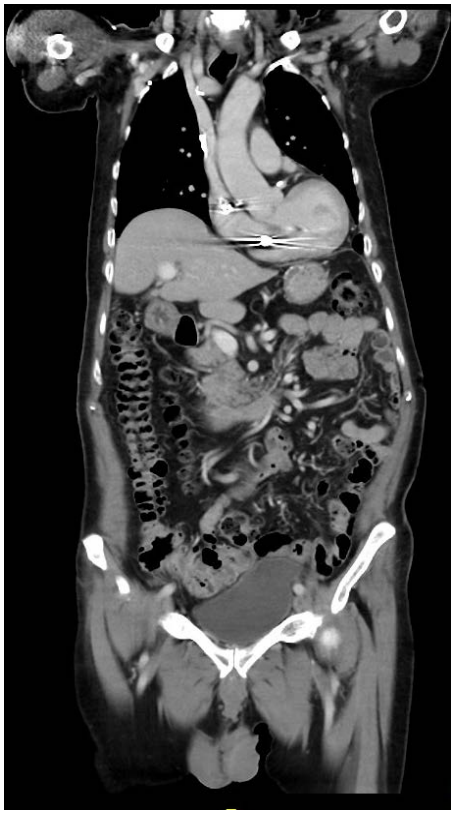

Figure 3: Contrast enhanced CT of the abdomen. Coronal view shows the presence of the pancreatic head and uncinate process.

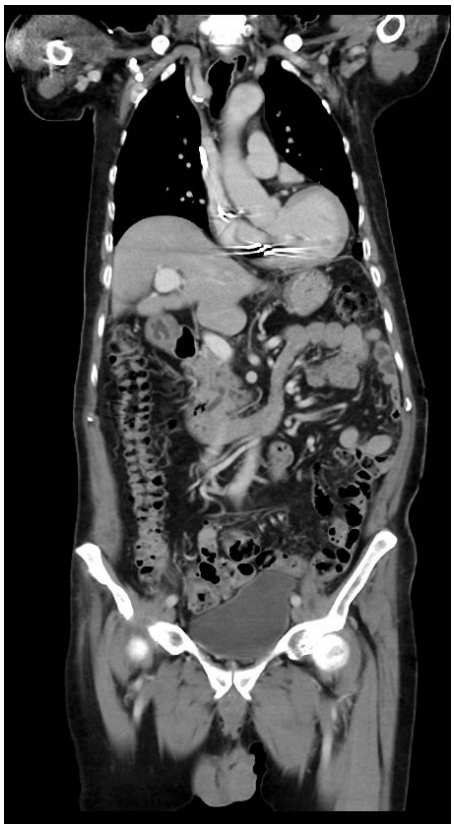

Figure 4: Contrast enhanced CT of the abdomen. Coronal view shows the presence of the common bile duct but no communicating pancreatic main duct.

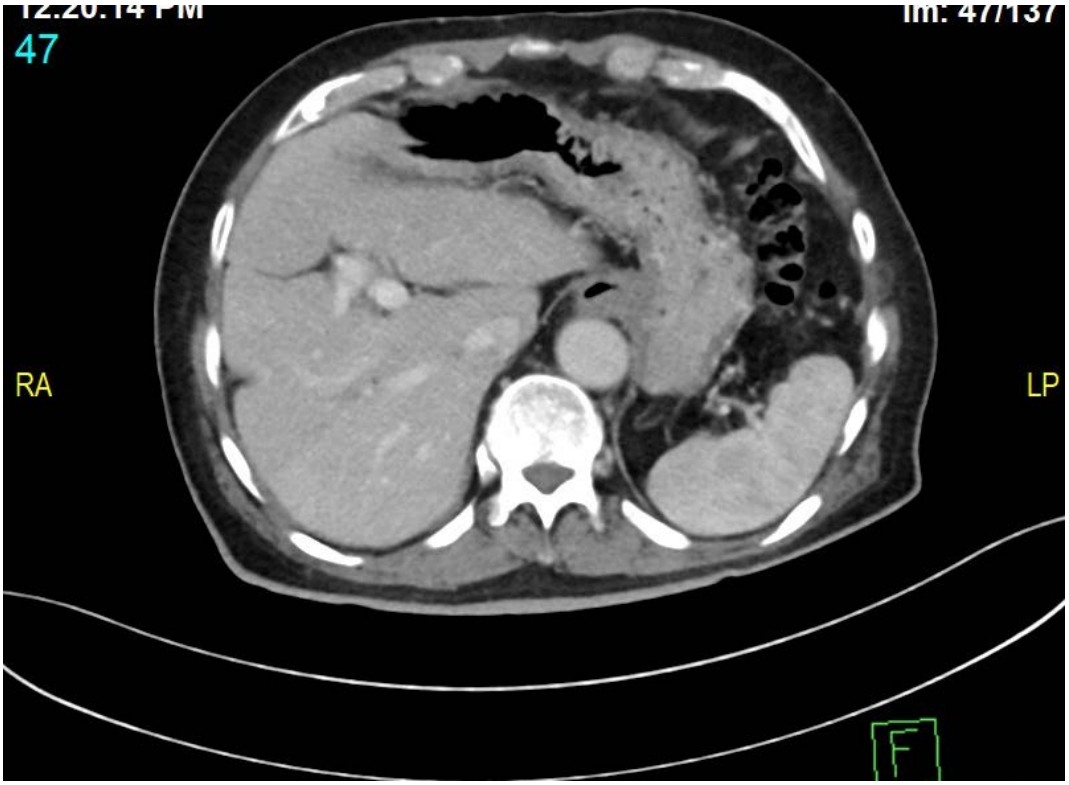

Figure 5: Contrast enhanced CT of the abdomen. Axial view shows the spleen and representative liver to be largely unremarkable. Mild heterogeneity of the spleen is likely due to contrast phase.

KEYWORDS

Agenesis of the dorsal pancreas; Dorsal pancreas agenesis; Pancreas; Computed tomography

ABBREVIATIONS

ALT = Alanine Transaminase
AST = Aspartate Transaminase
ALP = Alkaline Phosphatase
CT = Computed Tomography
MRI = Magnetic Resonance Imaging
ERCP = Endoscopic Retrograde Cholangiopancreatography
MRCP = Magnetic Resonance Cholangiopancreatography