Investigaciones Medicoquirúrgicas
ÓRGANO OFICIAL DEL CIMEQ

2025; 16: e921

Case presentation

Agenesis of the dorsal pancreas

Agenesia del páncreas dorsal

Amarilis Duarte Fernández [1] https://orcid.org/0000-0003-0331-1560
Luis Enrique Concepción Portales [1] https://orcid.org/0000-0002-4348-5963
Daryl Rodríguez Denis [1] https://orcid.org/0000-0002-8690-9869
Virginia Hernández Vargas [1] https://orcid.org/0000-0002-4986-7065

[1] Medical and Surgical Research Center. Havana, Cuba.
*Corresponding author. Email:amarilisduartefernandez@gmail.com

SUMMARY

Introduction: Dorsal pancreatic agenesis is a rare congenital anomaly. Diagnosis depends on a combination of imaging tests. Endoscopic ultrasound has proven to be effective and reliable in diagnosis.

Objective: To present a rare case of agenesis of the dorsal pancreas.

Clinical case: A 29-year-old female patient with dyspeptic symptoms over the past 10 years, which progressively worsened, including slow digestion, early fullness, nausea, vomiting, and a feeling of heaviness in the upper abdomen. She also reported frequent diarrhea with steatorrhea. The abdominal ultrasound described three rounded images in projection of the spleen, with a nodular appearance, confirmed in the contrast-enhanced computed axial tomography as nodular images, with hyperenhancement in the left hypochondrium, with thickening of the uncinate portion and the pancreatic area. The cholangioresonance showed shortening of the pancreas with a short body and a non-

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visualized tail, which suggested partial agenesis of the dorsal pancreas. The endoscopic ultrasound showed a decrease in the size of the body and absence of the tail of the pancreas. Treatment with pancreatin was indicated, which achieved improvement of the symptoms.

Conclusions: Imaging studies are useful to confirm the presence of this congenital anomaly.

Keywords: endoscopic ultrasound; abdominal pain; computed tomography; pancreas.

RESUMEN

Introducción: La agenesia del páncreas dorsal es una anomalía congénita infrecuente. El diagnóstico depende de la combinación de exámenes de imagen. La ecoendoscopía en el diagnóstico ha demostrado ser eficaz y fiable.

Objetivo: Presentar un caso poco frecuente de agenesia del páncreas dorsal.

Caso clínico: Paciente femenina de 29 años de edad con síntomas dispépticos en los últimos 10 años, que empeoraron de forma progresiva, dados por: digestiones lentas, plenitud precoz, náuseas, vómitos y sensación de peso en hemiabdomen superior; además, refirió diarreas frecuentes con esteatorrea. En el ultrasonido abdominal se describieron tres imágenes redondeadas en proyección del bazo, de aspecto nodular, confirmadas en la tomografía axial computarizada contrastada como imágenes nodulares, hipercaptantes en hipocondrio izquierdo, con engrosamiento de la porción unciforme y del área pancreática. En la colangiorresonancia. Se observó acortamiento del páncreas con el cuerpo corto y la cola no se visualizó, lo que hizo pensar en una agenesia parcial del páncreas dorsal. En la ecoendoscopía se observó disminución del tamaño del cuerpo y ausencia de la cola del páncreas. Se indicó tratamiento con pancreatina, con lo cual se logró la mejoría de los síntomas.

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Conclusiones: Los estudios de imagen son útiles para confirmar la presencia de esta anomalía congénita.

Palabras clave: ecoendoscopía; dolor abdominal; tomografía axial computarizada; páncreas.

Received: 12/30/2024
Accepted: 06/01/2025

INTRODUCTION

Dorsal agenesis of the pancreas is a rare congenital anomaly. Most cases are asymptomatic and are diagnosed incidentally. [1] In symptomatic cases, clinical manifestations vary from abdominal pain (the most frequent symptom), pancreatitis and diabetes mellitus to exocrine pancreatic insufficiency with steatorrhea. [2]

Abdominal pain may be due to sphincter of Oddi dysfunction and/or chronic pancreatitis accompanied by elevated pancreatic intraductal pressure. This symptom and diabetes are the clinical manifestations that most commonly reflect exocrine and/or endocrine insufficiency, since most of the islets of Langerhans are located in the tail of the pancreas. [3, 4, 5]

The aim of this work is to present a rare case of agenesis of the dorsal pancreas.

CLINICAL CASE

A 29-year-old female patient with dyspeptic symptoms over the past 10 years, which progressively worsened, including: slow digestion, early fullness, nausea, vomiting, feeling of heaviness in the upper abdomen and frequent diarrhea with steatorrhea. She went to her health

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area and was prescribed an abdominal ultrasound in which they described the presence of three rounded images in projection of the spleen, with a nodular appearance, which is why she was referred to the General Surgery consultation.

The physical examination revealed that the left hepatic lobe extended beyond the midline and occupied a large part of the left hypochondrium. A contrast-enhanced computed tomography (CT) scan of the abdomen was performed to better assess the images reported in the ultrasound (Figures 1, 2 and 3).

Plain CT report: The presence of 6 rounded images is striking, the largest measuring 4 cm and the others approximately 2 to 3 cm. Located in the left hypochondrium. Kidneys, pancreas and adrenals are unchanged. Liver of homogeneous density with a prominent left lobe.

CT report with intravenous contrast: the presence of moderately hyperenhanced nodular images in the left hypochondrium with the appearance of splenic nodules is noted. There is an impressive thickening of the uncinate portion and the pancreatic area, which should be assessed by cholangioresonance. In the arterial phase, the presence of small and multiple vessels projecting at the level of the falciform ligament, as well as an enhancement of regional intestinal loops, is striking. There is an impressive anomalous vascularization at the level of the uncinate process.

Magnetic resonance cholangiopancreatography report: Gallbladder septate towards the fundus. Extrahepatic bile ducts of normal appearance. The existence of isointense images in the T1 and T2 sequence is confirmed in the left hypochondrium in relation to the reported splenic nodules. It is striking that there is shortening of the pancreas with a short body and the tail is not visualized, which suggests partial agenesis of the dorsal pancreas.

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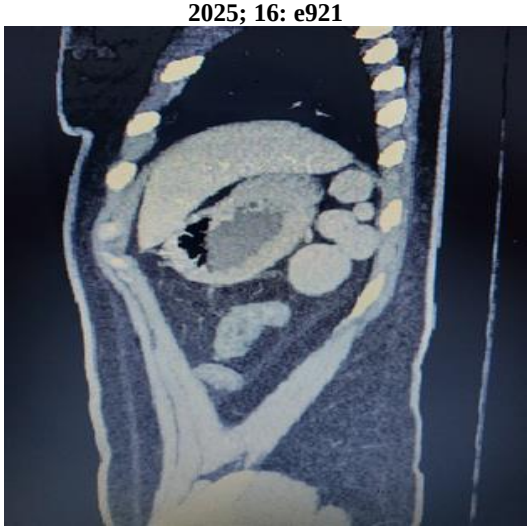

Fig. 1- Contrast-enhanced CT, lateral section. Images compatible with accessory spleens are observed.

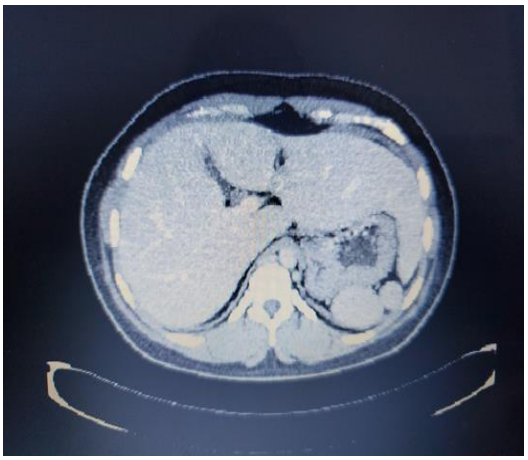

Fig.2- Axial CT scan. Prominent left hepatic lobe is visible.

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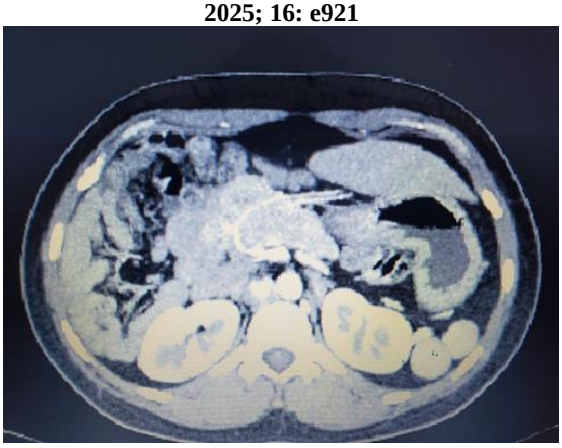

Fig. 3- Axial section with contrast CT. Short pancreatic body and absence of the tail are observed. The patient was consulted with the Gastroenterology service and underwent abdominal ultrasound with gallbladder emptying and endoscopic ultrasound.

Abdominal ultrasound with gallbladder emptying: thin-walled, septate and angled gallbladder. Volume 56 x 21 x 36 mm. Volume: 22,8 cm ^{3}. Volume of the gallbladder after two hours: 6, 7 cm ^{3}

Pancreatic endoscopy: The decrease in the size of the body and the absence of the tail of the pancreas are striking, without adequately visualizing the main pancreatic duct. Three rounded hypoechoic lesions are observed, which appear to be related to accessory spleens. A collective discussion was held between the General Surgery and Gastroenterology services and it was decided to prescribe treatment with pancreatin 25,000 U, one tablet before meals, which achieved improvement in symptoms.

COMMENTS

Dorsal pancreatic agenesis is a rare congenital anomaly. The first case was recorded in 1911 during an autopsy and to date less than 100 cases have been reported in the world literature. [2, 6, 7]

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It may be associated with a tumor or with other anomalies, such as polysplenia, ectopic spleen, cardiac anomalies, intestinal malrotation and has been thought to be part of a syndrome. [7, 8]

In complete dorsal agenesis, the anterior part of the head, neck, body and tail of the pancreas, the minor papilla and the accessory pancreatic duct of Santorini are absent and the pancreatic bed anterior to the splenic vein is occupied by the stomach and intestinal loops. In partial dorsal agenesis, a variable amount of pancreatic tissue is absent, but the minor papilla is present along with a remnant of the accessory pancreatic duct. [2] In the reported case, partial agenesis of the dorsal pancreas is described; the patient has a short body and an absent tail.

This entity has been reported in children and adults. It may be associated with an autosomal dominant mutation of the hepatocyte nuclear factor 1B (HNF1B) gene, although in most cases it is sporadic. [3]

Pancreatic development is a complex process and results from the fusion of the ventral and dorsal buds. The ventral bud forms the head and uncinate process, while the dorsal bud forms the upper part of the head, isthmus, and tail of the body of the pancreas. Dorsal agenesis occurs when there is abnormal development of the dorsal pancreatic bud, but there is regular development of the ventral bud. [6] Agenesis of the ventral pancreas and complete agenesis of the pancreas are lethal conditions. [9]

Diagnosis depends on the combination of imaging tests, including plain or contrast-enhanced CT and endoscopic retrograde cholangiopancreatoduodenography or magnetic resonance cholangiopancreatography, demonstrating the absence of the pancreatic body, the tail, and also the absence of the Santorini duct. Endoscopic ultrasound in diagnosis has proven to be effective and reliable. [10, 11]

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Treatment is not necessary for asymptomatic patients. The primary goal of therapy is to relieve associated symptoms. A low-fat diet and diabetes control are recommended for patients with diabetes. If pancreatitis is present, pancreatic enzymes may be administered to reduce pancreatic secretion and promote pain relief. When pancreatic tumors or other malignant medical problems are suspected in patients with dorsal pancreatic agenesis, surgical therapy such as pancreaticoduodenectomy or total pancreatectomy should be considered. In addition, pancreatin supplementation might be helpful in patients with dorsal pancreatic agenesis complicated by the symptoms of exocrine pancreatic insufficiency. (10)

Dorsal pancreatic agenesis is a rare condition, and it is important to understand it in order to diagnose it, given its variety of presentations, which can range from asymptomatic to pancreatitis or diabetes mellitus. Imaging studies are useful to confirm the diagnosis of this congenital anomaly. In asymptomatic patients, treatment is not necessary, and in symptomatic patients, therapy is aimed at alleviating symptoms. Imaging follow-up may be recommended to detect the possible development of pancreatic neoplasia.

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revinmedquir@infomed.sld.cu

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Conflicts of interest

The authors report no conflicts of interest.