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Partial agenesis of dorsal pancreas. Report of two cases

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ABSTRACT

INTRODUCTION: Agenesis of the dorsal pancreas (ADP) is a rare congenital anomaly resulting in missing corpus and cauda of the pancreas. The possibility of finding this disorder is increasing with the use of advanced radiological techniques like CT scan or MRI.
PRESENTATION OF CASE: We reported 2 cases of a partial ADP as radiological finding: the first one was a 79-year-old asymptomatic patient who presented to perform a CT staging scan for bladder tumor, while the second case was a 73-year-old patient with obstructive jaundice and with suspected common bile duct calculi. In the second patient US, CT scan and MRI were performed, and after that also an ERCP was scheduled.
DISCUSSION: The prevalence of agenesis of the dorsal pancreas is not exactly known; in the literature, only 50 cases have been reported. Its cause and pathogenesis are not fully understood. Some patients experience no symptoms, while others may develop hyperglycemia, diabetes mellitus, bile duct obstruction, abdominal pain, pancreatitis, or other conditions.
CONCLUSION: Considering that dorsal agenesis is sporadically found, often do not have related symptoms and it does not require a specific treatment, whether further examinations are needed to determine the type of agenesis remains questioned.

1. Introduction

Agenesis of dorsal pancreas is an extremely rare congenital anomaly. The first case was described in 1911 and so far around 100 cases have been reported in the world scientific literature [1,2]. Partial or complete agenesis of the dorsal pancreas as a rare congenital anomaly results from embryologic failure of dorsal pancreatic budding in the developing fetus [3–5]. Patients with agenesis of the dorsal pancreas can be asymptomatic or might refer abdominal pain, weight loss, pancreatitis, diabetes mellitus, bile duct obstruction, or duodenal obstruction [6–8]; rarely, they may also develop pancreatic exocrine insufficiency or pancreatic adenocarcinoma [9–13].

Agenesis of the dorsal pancreas can be detected with computed tomography (CT), ultrasound (US), magnetic resonance imaging (MRI), magnetic resonance cholangiopancreatography (MRCP), but the gold-standard technique is endoscopic retrograde cholangiopancreatography (ERCP) [14,15].

The work reported is in line with the SCARE criteria [16]. Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

2. Presentation of cases

2.1. Case 1

A 79-year-old male patient with no symptoms presented to our Radiology Department to perform a Total Body CT scan for staging bladder tumour, previously treated with TURB and BCG instillation. His past medical history was silent for other diseases, as well as his drug history. CT scan, performed by an expert radiologist, revealed the presence of small sized pancreas, due to partial agenesis of its dorsal portion (Fig. 1). The pancreas presented normal head, body and uncinate process, with the absence of body-tail portion and autonomous outlet of the pancreatic duct, as it happens in *pancreas divisum*. After an endocrinological consultation, considering the absence of symptoms, no further exams or any type of therapy were recommended. The patients was followed up as an outpatient, with the advice to perform blood tests annually to monitor pancreatic function.

2.2. Case 2

A 73-year-old male patient with abdominal pain was admitted to the Emergency Department of our Hospital. In his medi-

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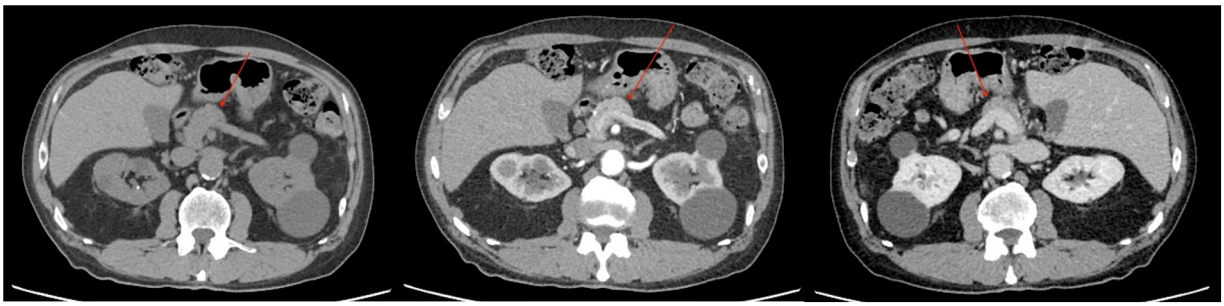

Fig. 1. CT scan revealed a normal-appearing pancreatic head, body, uncinate process and partial absence of the body-tail portion.

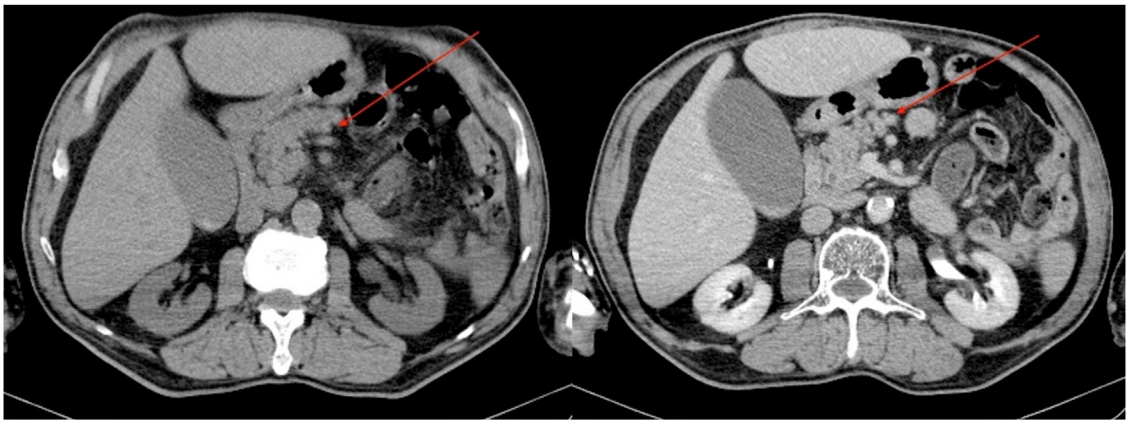

Fig. 2. CT images reveals the absence of the pancreatic tail body-portion.

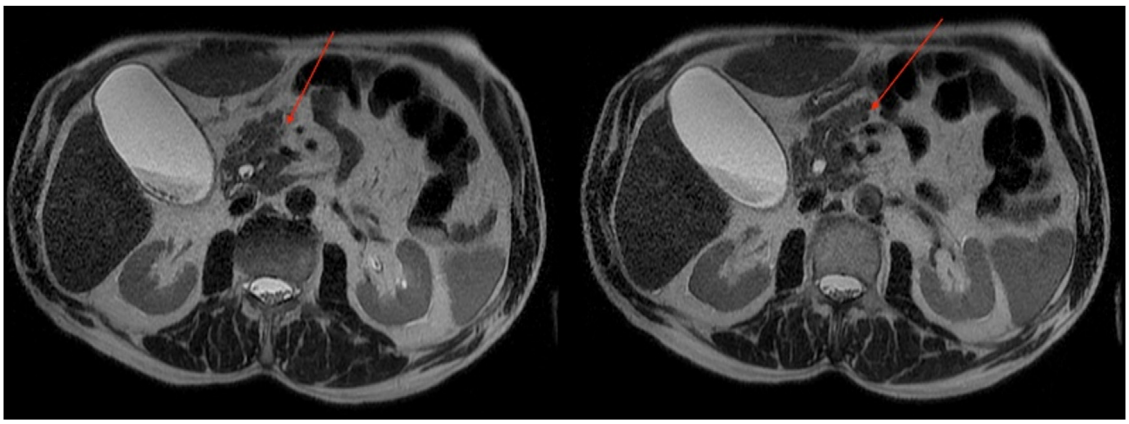

Fig. 3. MRI confirms the CT scan evidence of partial dorsal agenesis of the pancreas and the presence of luminal prepapillary non-obstructive filling defect.

cal history, he referred presence of diabetes mellitus, chronic obstructive pulmonary disease (COPD), benign prostatic hyperplasia (BPH); he had previous cholecystostomy performed for gallbladder empyema. Abdominal US was performed, that revealed the presence of biliary sludge and microlithiasis of the gallbladder. Subsequently, due to the clinical suspect (blood tests result normal except high glucose level of 221 mg/dl), CT scan (Fig. 2) and MRI (Fig. 3) were performed, that confirmed the presence of biliary sludge, microlithiasis of the gallbladder, small and non-obstructive filling defect in the common bile duct, probably due to lithiasis, and also the absence of the pancreatic body-tail portion was confirmed, as partial dorsal agenesis. An ERCP was scheduled: 3–5 mm biliary stones and biliary sludge were removed. Finally, patient underwent laparoscopic cholecystectomy. No other symptoms were referred, so the patient was discharged after surgery with the indication to annually perform blood tests and abdominal ultrasound.

3. Discussion

The pancreas embryologically derives by ventral and dorsal endodermal buds. The dorsal bud forms the upper part of the head, body and tail of the pancreas and drains through the Santorini duct. From the ventral bud originates the major part of the head and uncinate process which drains through the Wirsung duct. During the complex development, congenital abnormalities could occur. Complete agenesis of the pancreas and agenesis of the ventral pancreas are not compatible with life [17].

Agenesis of the dorsal pancreas is described in two forms: partial or complete. In complete dorsal agenesis, the minor papilla, the accessory pancreatic duct, the body and the tail of the pancreas are absent. In partial agenesis, the minor papilla with remnant of the accessory pancreatic duct and the neck and proximal body of pancreas are present [18].

The prevalence of agenesis of the dorsal pancreas is not exactly known; in the literature, only 50 cases have been reported [19]. Nowadays, its cause and pathogenesis are not fully understood. The HNF1B gene is known to regulate pancreatic development and some studies have found a correlation with the phenotype in HNF1B mutation carriers; also GATA6 gene mutations also can lead to pancreatic hypoplasia [20]. Another possible explanation is an ischemic insult to the developing pancreas [21].

Most cases of agenesis of the dorsal pancreas do not have specific symptoms and the diagnosis is incidental during evaluation for other issue. In some cases, patients could refer abdominal symptoms such as pain or bloating, that may be caused by sphincter of Oddi dysfunction or by autonomic neuropathy resulting from diabetes mellitus [19]. Several studies reported an association with acute pancreatitis, maybe due to Oddi sphincter dysfunction, pancreatic head compensatory hypertrophy, increased pancreatic juice secretion and pancreatic duct hypertension [18]. Moreover, most of the islet β cells are located in the tail of the pancreas, and this explains why patients with dorsal agenesis also have diabetes due to insufficient insulin secretion.

A very limited number of pancreatic tumors are reported in association with agenesis of the dorsal pancreas, including a malignant IPMN [22] and cystic lesions [23]. The association is unclear, but chronic pancreatitis is considered one of the risk factors for cancer.

Finally, dorsal agenesis is associated with other organ malformations, including polycystic kidney disease, Kartagener syndrome, multiple splenic deformities, congenital choledochal cysts and biliary atresia [19].

With the development of medical imaging technology, many methods to diagnose agenesis of the dorsal pancreas have been established, such as CT, MRI, US and endoscopic ultrasound (EUS). ERCP is necessary to confirm agenesis of the dorsal pancreas, because it is important to define the anatomy of the pancreatic ducts and to differentiate this anomaly from other diagnostic possibilities, such as *pancreas divisum* or pancreatic neoplasm. Contrast MRCP clearly demonstrates pancreatic duct morphology. Agenesis of the dorsal pancreas is an uncommon condition that typically remains asymptomatic or may present with a variety of signs and symptoms. In the absence of symptoms, no specific therapy is required, neither further in-depth diagnostic exams.

4. Conclusion

Agenesis of the dorsal pancreas, a rare congenital malformation, must be kept in mind when the body and tail of the pancreas cannot be seen during routine examinations. With the availability of cross-sectional imaging, many cases of agenesis of dorsal pancreas have been detected in the last decade. This diagnosis could be missed by US, due to difficulty on visualization of body and tail of pancreas, but MRI combined with MRCP can accurately visualize all the pancreatic duct as well as his whole parenchyma. Considering that dorsal agenesis is sporadically found, often do not have related symptoms and it does not require a specific treatment, whether further examinations are needed to determine the type of agenesis remains questioned.

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Consent

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Author contribution

Sayali Valiyeva, Romano Lucia: Writing the paper.
Schietroma Mario: Data collection and analysis.
Carlei Francesco, Giuliani Antonio: Study concept.

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Declaration of Competing Interest

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