

INTRODUCTION

Pancreas divisum (PD) is the most common congenital anomaly of the pancreas (5-10% in Western countries), resulting of non-fusion of the ventral and dorsal pancreatic duct systems.
Pancreatic tumors may arise in 11.1–12.5% of PD cases. Frequently these tumors correspond to ductal adenocarcinoma, with intraductal papillary mucinous neoplasm (IPMN) being rare in PD.
 We present a case of IPMN arising from dorsal duct of incomplete PD and discuss the relationship between these two conditions.

CLINICAL PRESENTATION

74-year-old man

Past medical history: acute pancreatitis (a unique episode) 18 years before and a prior cholecystectomy (lithiasis).

Referred to our department: slightly dilated main pancreatic duct in an abdominal CT (Fig. 1a) performed due to fever of unknown origin.

Complementary diagnostic exams:

- **Blood tests:** serum amylase level, tumor marker levels, fasting plasma glucose and hemoglobin A1c within normal range.

- **Magnetic resonance cholangiopancreatography (MRCP):** depicted a **incomplete PD**. Dorsal pancreatic duct was dilated (14 mm) with no evidence of pancreatic masses or obstructive causes (Fig. 1b).

- **EUS:** confirmed the **dilatation of the dorsal duct (14 mm)** and **revealed the presence of several mural nodules > 5mm** within the dilated dorsal duct. (Fig. 2).

- **Fine-needle-aspiration** of the dilated dorsal duct was performed with a 22G needle and showed a **carcinoembryonic antigen of 1001.8** (normal range: <5 ng/mL); cytological examination was inconclusive.

Final diagnosis:

Main duct intraductal papillary mucinous neoplasia of the pancreas (MD-IPMN).

After staging showing no distant disease, the patient was proposed to **pancreaticoduodenectomy**.

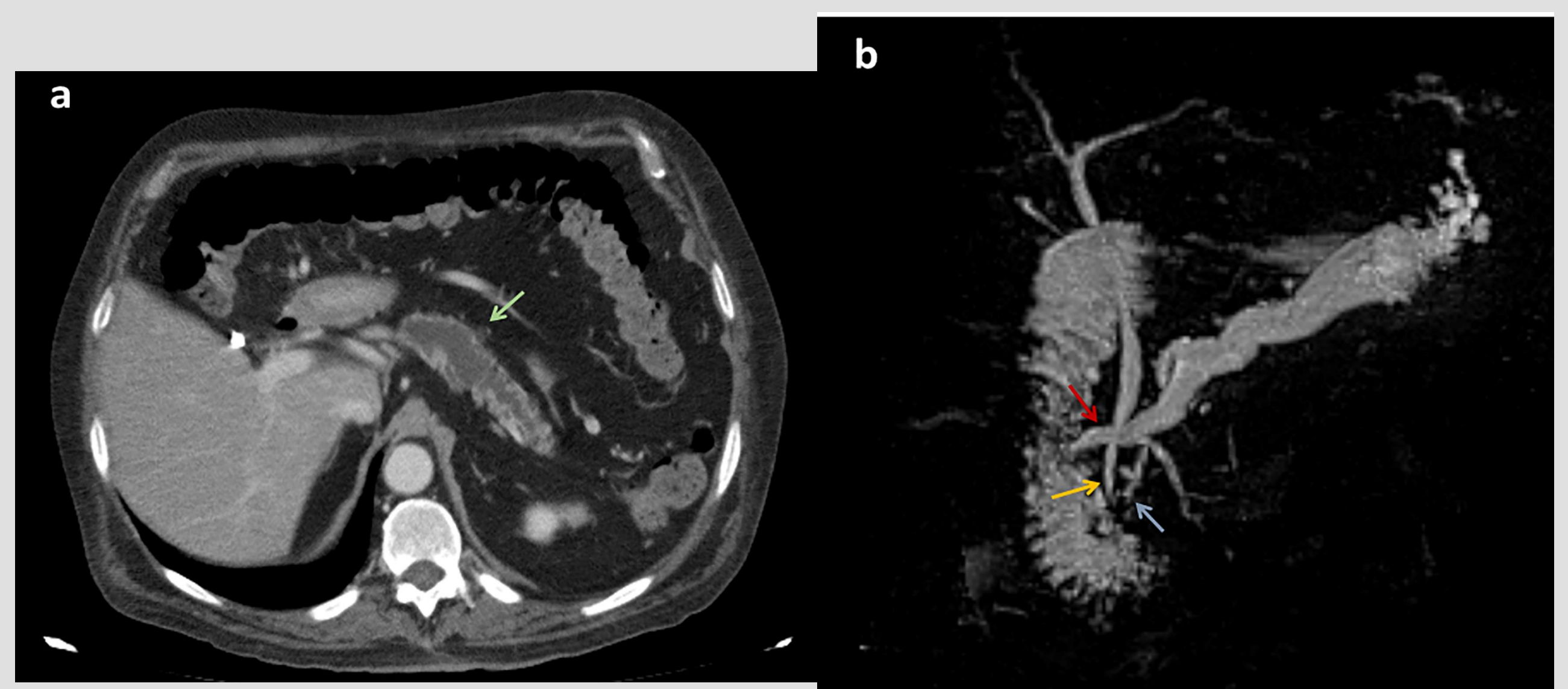


Figure 1. Enhanced CT scan images in axial view (a) showing diffuse dilatation of the main pancreatic duct in the pancreatic body and tail (maximum diameter of 22 mm in pancreatic body), with atrophy of pancreatic parenchyma without any pancreatic mass. **MRCP 3D images** (b) showing incomplete PD in which the dorsal pancreatic duct (red arrow) enters directly in the minor papilla and the ventral pancreatic duct (blue arrow) and main biliary duct (yellow arrow) enter the major papilla. There is also a filamentous connection between dorsal and ventral ducts. Note the diffusely dilated dorsal pancreatic duct.



Figure 2. EUS images showing dilatation of the dorsal duct (a) and presence of digital protrusions within the dorsal duct (b). Ventral duct presented normal caliber (red arrow) and was linked to the dorsal duct (yellow arrow) (c).

DISCUSSION/CONCLUSION

Despite of increasing diagnosis of the PD and IPMN, these conditions continue to be rare and PD associated with IPMN is even rarer. A study from 2015 identified 15 cases of IPMN associated with PD reported in the literature (1). Based on these reports, the following clinical characteristics were identified: 1) female predominance (80 %), 2) predominance of type 1 (complete PD) (80 %), 3) tumor location in the dorsal pancreas (80 %) rather than in the ventral pancreas, 4) a predominance of the branch-duct type (60 %) over the main-duct type and 5) clinical symptoms in more than half of the patients.

Recent studies have showed a tendency for increased pancreatic malignancies associated with PD. Frequently these tumors correspond to ductal adenocarcinoma, with IPMN being rare in PD. It is suggested that chronic obstruction of the pancreatic duct due to relative stenosis of the minor duodenal papilla in PD may lead to recurrent inflammation and therefore promote oncogenesis. Although this mechanism may justify the occurrence of ordinal pancreatic cancer in the dorsal pancreas in patients with PD, as IPMN is considered a cause and not a consequence of pancreatic inflammation, **the relationship between PD and IPMN is currently unclear.** Even so they may not be etiologically related, as both may lead to recurrent pancreatic inflammation, their coexistence may have a synergistic effect on the risk of pancreatic cancer. These are critical issues that warrant further investigation in the future.

Although it remains unclear if PD and IPMN may be etiologically related we emphasize the importance of consider the coexistence of MD-IPMN in patients with PD and dilation of dorsal duct, even in the absence of symptoms.

REFERENCES

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