International Journal of Surgery Case Reports 6 (2015) 169–171

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Selective agenesis of pancreatic isthmus parenchyma with preservation of main pancreatic duct continuity, a very rare entity: Case report

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ARTICLE INFO

Article history:
Received 16 September 2014
Accepted 18 September 2014
Available online 11 December 2014

Keywords:
Pancreatic anomaly
Dorsal pancreatic agenesis
Agenesis of pancreatic isthmus
IPMN
BD-IPMN

ABSTRACT

CONTEXT: Agenesis of the dorsal pancreas is a rare anomaly, mostly associated with other medical conditions. It may be complicated with pancreatic neoplasms.
CASE REPORT: We report the case of a 51-year-old male with selective agenesis of pancreatic isthmus with preservation of main pancreatic duct and branch-duct intraductal papillary mucinous neoplasm (BD-IPMN) with suspicious features of the pancreas.
CONCLUSION: This is probably the first report of isolated agenesis of pancreatic isthmus with conservation of main pancreatic duct.

1. Introduction

Embryologic development of the pancreas is very complex. The dorsal pancreatic bud becomes the body, tail, and isthmus of the pancreas and the ventral pancreatic bud forms the pancreatic head and uncinate process. While glands fuse the duct systems anastomose, then the main pancreatic duct (MPD) is formed too by the fusion of the dorsal and ventral pancreas. Several different anatomic anomalies can develop when pancreatic endodermal buds variously fail to fuse; however they remain very rare entities. The most common congenital abnormality is pancreas divisum, detected in 7% of autopsies.[1] To our knowledge, isolated agenesis of pancreatic isthmus has never been described before. It can be considered as a subtype of dorsal agenesis of the pancreas (DAP), that, in and of itself, it has been reported in about only fifty cases, mostly associated with other clinical conditions.[2–4]

2. Case report

A 51-year-old Caucasian male was referred to our outpatients' pancreatic cyst clinic from his family doctor due to Cholangio-Wirsung Magnetic Resonance (MRCP) findings of dilation of the whole main pancreatic duct associated with multiple cystic lesions. On physical examination, everything was normal. The past medical records revealed that the patients suffered from an acute episode of biliary pancreatitis, treated medically and then surgically (laparoscopic cholecystectomy) 10 years before (another Hospital). There was no family history of pancreatic neoplasms. He did not have history of diabetes, alcohol abuse and he did not use any drugs on regular basis. Biochemical tests did not reveal abnormalities. Oncomarker levels of Ca 19-9 were normal (<37 U/mL). MRCP images (Fig. 1) showed a dilation of MPD (especially at the body, with maximum diameter of 6–7 mm) with three cystic lesions at the uncinate process, body and tail of the pancreas (32 mm, 23 mm and 20 mm maximum diameters respectively), all in connection with main pancreatic duct (Branch-Duct IPMNs – BD-IPMNs). On T2-weighted axial sequences among the three BD-IPMNs, the lesion located at the uncinate process showed an enhancing 7-mm diameter suspicious mural nodule inside (Fig. 2). Endoultrasonographic cytology of the mural nodule was then performed via fine-needle aspiration, with evidence of cytological atypia. At MRCP no findings were compatible with morphologic abnormalities of the main pancreatic duct. On the contrary, we identified a selective glandular agenesis of the pancreatic isthmus (Fig. 3). As a collateral, the patient recently underwent endoscopic resection of a rectal polyp (low-grade adenoma at pathological report), during a screening pancolonoscopy for high familiarity for colon cancer. According to the guidelines of the European Study Group for Cystic Tumors of the Pancreas[5] the patient was considered for surgical resection of pancreatic head in suspicion of degeneration of multifocal BD-IPMN.

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http://dx.doi.org/10.1016/j.ijscr.2014.09.023

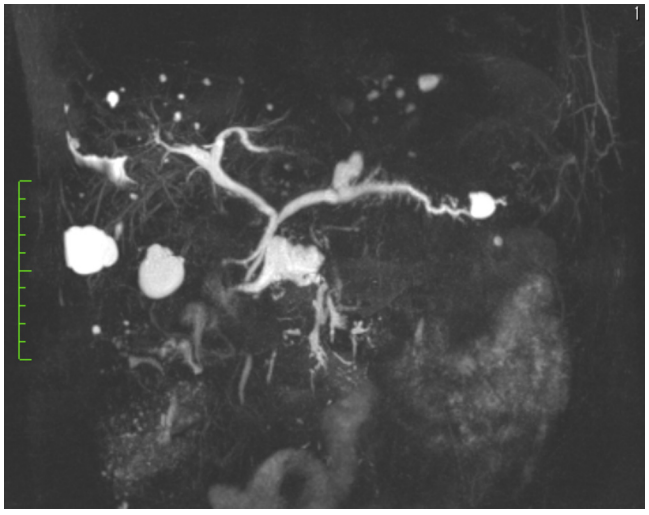

Fig. 1. MRCP scan showing multifocal BD-IPMN with three big cystic lesions (BD-IPMNs) with associated dilation of the main pancreatic duct.

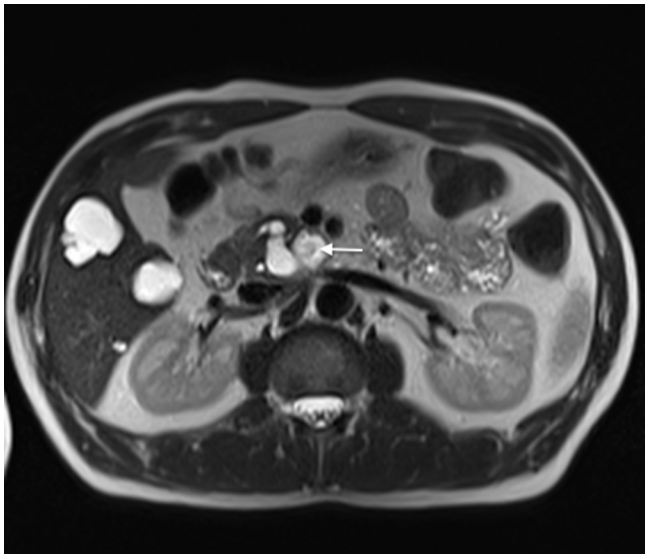

Fig. 2. T2-weighted axial scan demonstrating the worrisome 7-mm diameter mural nodule inside the main BD-IPMN located at the uncinate process.

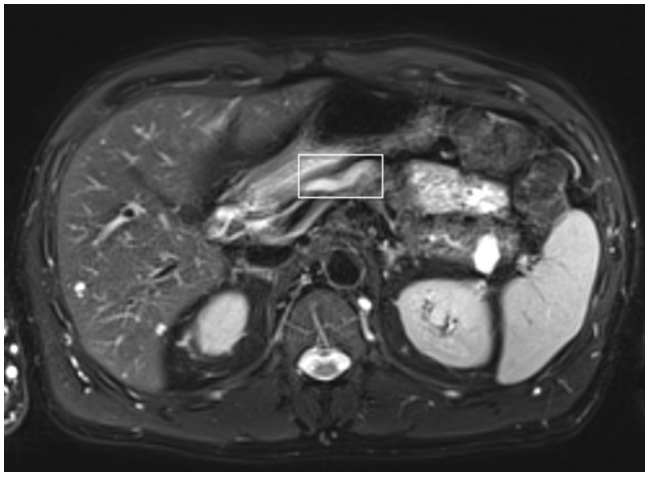

Fig. 3. MRCP scan showing the selective absence of pancreatic parenchyma at the isthmus with preservation of the main pancreatic duct.

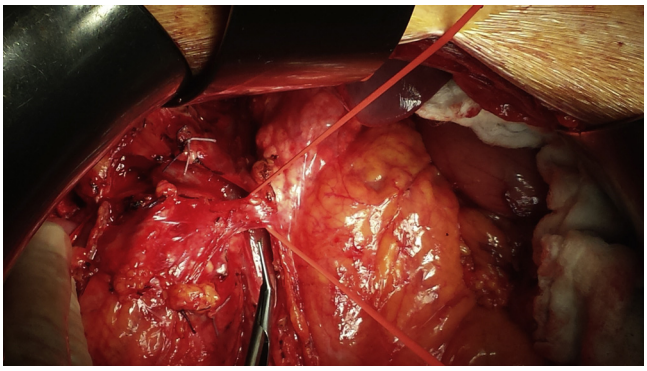

Fig. 4. Intraoperative finding: a 2.5 cm long main pancreatic duct not surrounded by pancreatic parenchyma and ready to be cut for frozen section.

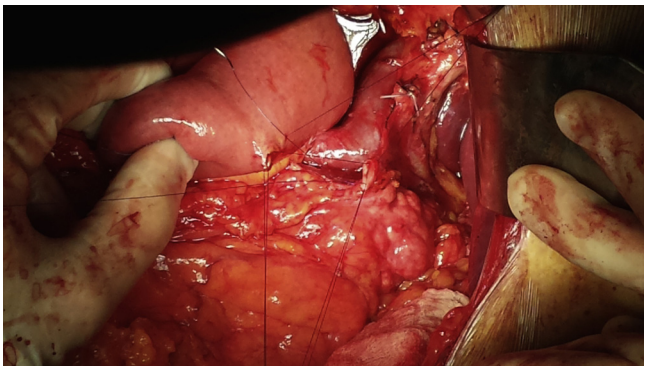

Fig. 5. Duct-to-mucosa pancreaticojejunostomy.

2.1. Perioperative management, intraoperative findings and pathological report

A median laparotomy with xifo-umbilical approach was adopted. After having excluded abdominal secondary lesions, the gastro-colic ligament was opened to access to the pancreatic region. After Kocher maneuver, the pancreatic head was explored. It was enlarged and disconnected from the rest of body parenchyma. The "supposed" isthmus zone was totally occupied only by the naked main pancreatic duct (Fig. 4) for about 2.5 cm in length, with a diameter of about 6–7 mm. An intraoperative ultrasonography was then performed, to assess the nature of the remaining two BD-IPMNs located at the body and at the tail of the gland; once their benign aspect was confirmed they were not removed. A sliver of main pancreatic duct was submitted to frozen section for histologic examination, resulted negative for atypical cells. Hence, a standard pylorus-preserving pancreaticoduodenectomy was performed with duct-to-mucosa pancreaticojejunostomy (Fig. 5), hepaticojejunostomy and duodenojejunostomy. Final pathological examination revealed a High-Grade Mixed-type IPMN, with 22 clear lymph nodes and free resection margins. At the pathological examination the suspected cyst's diameter was 2 cm, with inner papillary proliferations and clear communication with the main pancreatic duct and both presented the same (high) grade of dysplasia. Post-operative course was regular and the patient was discharged to home on post-operative day 7.

3. Discussion

The pancreas is formed by rotation and fusion of ventral and dorsal endodermal buds, at around 6–7th week of gestation. This fusion creates both glandular tissue and main pancreatic duct. During this complex process different anomalies can occur. Complete

agenesis of the pancreas and agenesis of the ventral pancreas are not compatible with life.[6] Dorsal pancreas is variously involved in development abnormalities. Agenesis of dorsal pancreas is mostly diagnosed incidentally, since the most part of patients are asymptomatic. It is a rare entity and it can be complete or partial. Complete agenesis of dorsal pancreas is rare, with about fifty cases reported in English literature, mostly as case reports.[7–9] Usually it results in impairment of pancreatic function and often it passes undetected.[2] Partial agenesis of dorsal pancreas is even more rare, in fact it has been previously reported in literature only in few sporadic cases.[10]

In some cases, dorsal agenesis of the pancreas has been reported in association with pancreatic neoplasms, varying from IPMN, to solid-pseudopapillary tumor and pancreatic adenocarcinoma.[11–13]

Interestingly, Rittenhouse et al. reported three cases of association of dorsal agenesis of the pancreas (DAP) with pancreatic adenocarcinoma (PDAC) and non-alcoholic chronic calcific pancreatitis (NCCP) among a novel triad of diseases. The authors hypothesized that similar molecular processes, involving retinoic acid and hedgehog signaling pathways, are involved in the development of DAP, PDAC and NCCP.[2] Usually, endoscopic retrograde cholangiopancreatography (ERCP) and MRCP are useful to investigate the morphology of the pancreatic ductal system, in order to rule out ductal abnormalities.[9] By the way, in the case we here report the main pancreatic duct was anatomically normal, without signs of discontinuity, the radiological clue being the absence of glandular parenchyma surrounding it. To our knowledge, this is the first report of isolated agenesis of pancreatic isthmus parenchyma with preservation of the normal continuity of the main pancreatic duct. Given its rarity it is not easy to consider it as a subtype of partial DAP.

In conclusion, complete or partial agenesis of the dorsal pancreas is an uncommon condition. In the presence of complete or partial agenesis of dorsal pancreas the presence of pancreatic neoplasm (solid or cystic) should not be excluded. Further data are needed to clarify if in these cases the patients should be considered at a high-risk to develop pancreatic neoplasms.

Conflict of interest

The authors have no conflict of interest.

Funding

None.

Ethical approval

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.

Author contributions

Study design: Salvatore Paiella and Matteo De Pastena; Data collection: Alessandro Esposito and Consuelo Morigi; Writing: Salvatore Paiella and Claudio Bassi.

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