Case report

A giant retention cyst of the pancreas (cystic dilatation of dorsal pancreatic duct) associated with pancreas divisum

ZENICHI MORISE1, KAZUO YAMAFUI1, TADAO TSUJI2, ATSUNORI ASAMI1, KAORU Takeshima1, NORITAKA Hayashi1, HIDEO Baba1, FUMIKO Yoshida1, YUTA Abe1, and YASUYUKI Tokura1

1 Department of Surgery, Saitama Municipal Hospital, Saitama, Japan 2 Department of Internal Medicine, Saitama Municipal Hospital, Saitama, Japan

We describe a rare case of pancreas divisum associated with a giant retention cyst (cystic dilatation of the dorsal pancreatic duct), presumably formed following obstruction of the minor papilla. The patient was treated by pancreatico(cysto)jejunostomy. A 50-year-old man was admitted with complaints of increasing upper abdominal distension and body weight loss. There was no previous history of pancreatitis, gallstones, drinking, or abdominal injury. An elastic-hard tumor-like resistance was palpable in the upper abdomen. Computed tomography and ultrasound (US) examinations revealed a giant cystic lesion expanding from the pancreas head to the tail. Endoscopic retrograde cholangiopancreatography findings showed a looping pancreatic duct which drained only the head and uncinate process of the pancreas to the main papilla. A US-guided puncture to the cystic lesion revealed that the lesion continued to the main pancreatic duct in the tail of pancreas. The lesion was connected to a small cystic lesion, which was located inside the minor papilla, and ended there. The amylase level in liquid aspirated from the cyst was 37869 IU/l, and the result of cytological examination of the liquid showed class II. A pancreatico(cysto)jejunostomy was performed, with the diagnosis being pancreas divisum associated with a retention cyst following obstruction of the minor papilla. The histological findings of a specimen from the cyst wall revealed that the wall was a pancreatic duct covered with mildly inflammatory duct epithelium; there was no evidence of neoplasm. The patient is currently well, and a CT examination 2 years after the operation showed disappearance of the cyst and normal appearance of the whole pancreas.

Key words: pancreatic retention cyst, pancreas divisum, pancreaticojejunostomy

Introduction

Pancreas divisum is a fairly common congenital anomaly of the pancreas, with an incidence of 4%–9% reported in various series.1-5 The anomaly results from a failure of fusion of the dorsal and ventral pancreatic buds during fetal development. Several studies have shown an extremely high incidence of this anomaly, reaching 25%–50%, in patients with idiopathic pancreatitis.2,3 Although there is still controversy about a direct relationship between pancreas divisum and pancreatitis,4-5 such a relationship could be explained by the hypothesis that this anomaly occasionally causes dorsal pancreatitis due to insufficient drainage of the entire dorsal gland through the minor papilla (relative stenosis).1-3 At present, the recurrent acute pancreatitis in patients with pancreas divisum tends to be treated by endoscopic sphincterotomy and/or stenting.6,7 However, Warshaw et al.8 reported that there are patients with pancreas divisum with established chronic pancreatitis and, in these patients, surgical procedures such as pancreaticojejunostomy and pancreatectomy are required. We herein describe a rare case of pancreas divisum associated with a giant retention cyst (cystic dilatation of the dorsal pancreatic duct) presumably formed following obstruction of the minor papilla. The patient was treated by pancreatico(cysto)jejunostomy.

Case report

A 50-year-old man was admitted with complaints of increasing upper abdominal distension that had occurred for a period of 1 year, and body weight loss (6–
1080 Z. Morise et al.: A retention cyst and pancreas divisum

7kg in 6 months). On close questioning, it was revealed that he had had a recent episode of mild steatorrhea. However, he had never had a clinical picture of pancreatitis. There was no previous history of gallstones, drinking, or abdominal injury. He was not icteric or anemic, and an elastic-hard tumor-like resistance (15 × 5cm in size) was palpable in his upper abdomen. On laboratory tests, a mild elevation of serum lipase (300IU/l), and a low urinary paraaminobenzoic acid (PABA) excretion rate (60%) were observed in a pancreatic functional diagnostic (PFD) test for exocrine function. The serum amylase level and the result of the oral-glucose tolerance test were within normal ranges. There was no tumor marker for which the serum level was above the normal limit. Computed tomography (CT) and ultrasound (US) examination revealed a giant cystic lesion, 15 × 5cm in size, expanding from the pancreas head to the tail (Fig. 1). The cyst contents showed clear homogeneous fluid, and the cyst showed no tumor component or irregular wall thickening. Endoscopic retrograde cholangiopancreatography (ERCP) findings showed a looping pancreatic duct (the duct of Wirsung) which drained only the head and uncinate process of the pancreas to the main papilla. There was no connection to the duct which drained the body and tail of the pancreas or to the cystic lesion. The minor papilla was identified, but attempts at cannulation were unsuccessful (Fig. 2). There was no connection to the duct which drained the body and tail of the pancreas or to the cystic lesion. The minor papilla was identified, but attempts at cannulation were unsuccessful. A US-guided puncture to the cystic lesion, followed by cystography, revealed that the lesion continued to the main pancreatic duct (the duct of Santorini) in the tail of the pancreas, and branches of the pancreatic duct protruded directly from the lesion. Near the duodenum, the lesion connected with another, small, cystic lesion, which was located inside the minor papilla, and ended there (Fig. 3). The amylase level of the liquid aspirated from the large cystic lesion was 37869IU/l, and the result of cytological examination of the liquid showed class II. Magnetic resonance cholangiopancreatography (MRCP) revealed the same findings as those of ERCP and the US-guided cystography (Fig. 4).