Recurrent Acute Pancreatitis Secondary to Graft Pancreas Divisum in a Patient with Modified Multi-Visceral Transplant

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Abstract
A patient with modified multivisceral transplant developed recurrent acute pancreatitis (RAP) 1 year after transplant and was found to have graft pancreas divisum with otherwise negative work-up for identifying the etiology of RAP. Endoscopic retrograde cholangiopancreatography was performed with minor papilla sphincterotomy and pancreatic duct stent placement of the graft pancreas. The patient’s symptoms resolved following endotherapy for a follow-up period of 2 years. This is a unique case of graft pancreatitis secondary to pancreas divisum.

Introduction
Composite visceral transplantation (including combined liver/small bowel and multivisceral grafts) is the standard of care in patients with irreversible intestinal failure secondary to short bowel syndrome, abdominal vascular catastrophe, or major abdominal trauma. Multivisceral (MV) grafts include transplantation of the stomach, duodenum, pancreas, liver, and intestine, and are indicated in conditions such as hollow visceral myopathy/neuropathy, GI polyposis, and extensive mesenteric desmoid tumors. This procedure may be modified (modified MV) to contain all of these organs except the liver. Pancreaticobiliary complications including graft pancreatitis (GP) and rejection have been frequently reported in patients with modified MV transplant. The prevalence of such complications is far less when the pancreas is included as a component of a multivisceral graft as opposed to isolated pancreas transplantation.

Herein we describe a unique case of a patient with modified MV transplant who developed recurrent GP secondary to pancreas divisum. The patient provided consent for enrollment in an institutional review board–approved prospective cohort study and was included in a prior published series.

Case Report
A 40-year-old female with history of modified MV transplantation for hollow visceral myopathy presented 1 year after the transplant with recurrent acute pancreatitis (RAP). She had severe attacks of characteristic epigastric pain with concurrent lipase elevation that required hospitalization. Upon initial evaluation, her vital signs were normal. Physical exam showed upper abdominal tenderness to palpation without guarding. She had a lipase level of 625 IU/L (upper limit of normal <200) consistent with acute pancreatitis. Initial work-up, including review of medications, family history of acute pancreatitis, tobacco and alcohol use, serum triglyceride and ionized calcium level, celiac serologies, and right upper quadrant ultrasound was unrevealing. Abdominal MRI showed graft pancreas divisum to be the etiology of the recurrent GP (Figure 1).
Endoscopic retrograde cholangiopancreatography (ERCP) to treat the graft pancreas divisum was performed. The transplanted minor papilla was identified and successfully cannulated (Figure 2). Minor papilla sphincterotomy with pancreatic duct stent placement was then successfully performed. The patient did not develop any subsequent attacks of GP in 2-year clinical follow-up.

Discussion

In the presented case, a modified multivisceral technique was performed, where the graft stomach, duodenum, pancreas, and small intestine were transplanted en bloc with the native pancreas and liver preserved. This technique has been increasingly utilized over the past decade with significant improvement in outcome. However, pancreaticobiliary complications are relatively common after composite visceral transplantation. In a study of 271 patients with 289 composite visceral grafts, 19 patients developed pancreatic complications (6 patients had edematous pancreatitis, 7 had necrotizing pancreatitis, and 6 had pancreatic duct fistulae). Five patients experienced combined biliary and pancreatic complications.

Pancreas divisum is the most common congenital anomaly involving the pancreatic ductal system, with a reported prevalence between 2.7–22%. It is characterized by failure of fusion between dorsal and ventral pancreatic ducts during embryonic development. This condition is usually asymptomatic. Pancreas divisum is an uncommon cause of RAP. Secretin-enhanced MRCP is considered the gold standard imaging modality to diagnose pancreas divisum, but its sensitivity remains modest at 73% with a specificity of 96%.

The treatment of pancreas divisum causing RAP includes ERCP with minor sphincterotomy and stenting. Based on a recent meta-analysis, the pooled response rate of pancreatic endotherapy in patients with recurrent acute pancreatitis and pancreas divisum was 79%. This response rate was higher when compared to response rate of pancreatic endotherapy for patients with chronic pancreatitis (69%) or for pain only in the setting of pancreas divisum (54%).

We describe a unique case of pancreas divisum of the graft pancreas causing RAP in a patient with modified MV transplant. Clinicians treating patients with otherwise unexplained recurrent pancreatitis should have a low threshold to obtain secretin-enhanced MRCP to assess for pancreas divisum, which may be amenable to endoscopic intervention.

Disclosures

Author contributions: H. Nawaz reviewed the clinical data and prepared the manuscript. A. Slivka and GI Papachristou selected the case, reviewed the clinical data, and prepared the manuscript. GI Papachristou is the article guarantor.

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