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Intrapancreatic accessory spleen (IPAS): A single-institution experience and review of the literature

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ABSTRACT

Introduction: Accessory spleens located within the pancreatic parenchyma (intrapancreatic accessory spleen, IPAS) pose a unique clinical challenge. In many cases, despite imaging and other diagnostic studies, malignancy cannot be excluded and patients are subjected to pancreatic resection. We review our experience with the presentation, diagnosis, and treatment of patients with IPAS to provide insight into improving pre-operative evaluation of these patients

Methods: A retrospective chart review identified seven patients who underwent surgical resection of an intrapancreatic spleen at University of Louisville Hospital between 2004 and 2015. Charts were analyzed for presenting symptoms, pre-operative imaging, operative therapy, and final pathologic evaluation. Patients were included in the study if they underwent pancreatic resection for a pancreatic mass and were diagnosed with an IPAS on final pathologic evaluation.

Results: Patient age ranged from 38 to 72 with a median age of 62.5, including five males and two females. Lesions ranged from 1.4 to 7.4 cm in maximal diameter (mean 3.8 cm). All lesions were identified as round, hypervascular, well-circumscribed masses in the pancreatic tail. The most common pre-operative diagnosis was a non-functioning pancreatic neuroendocrine tumor (NF-PNET). The most common operative approach was laparoscopic distal pancreatectomy and splenectomy.

Conclusion: IPAS are benign tumors commonly mistaken for pancreatic neoplasms such as NF-PNET. A combination of CT, MRI and nuclear medicine examinations can confirm the diagnosis of IPAS and prevent unnecessary surgical resection.

1. Introduction

Ectopic splenic tissue, known as an accessory spleen, is present in approximately 12% of patients in autopsy series.[1] While accessory spleens most often reside in the splenic hilum, they can also be present within the pancreatic parenchyma.[2] Accessory spleens are usually of little clinical consequence and found incidentally during surgery for other indications. They can, however, cause symptoms, mostly often hematologic, after a splenectomy has been performed.

Accessory spleens located within the pancreatic parenchyma (intrapancreatic accessory spleen, IPAS) pose a unique clinical challenge. While accessory spleens have no malignant potential, their appearance on computed tomography (CT) as hypervascular masses often raises a concern for malignancy and can be radiologically indistinguishable from neuroendocrine tumors or other hypervascular tumors.[3] In many cases, despite imaging and other diagnostic studies, malignancy cannot be excluded and patients are subjected to pancreatic resection. Unfortunately, the current literature provides conflicting recommendations regarding preoperative discrimination of IPAS vs. tumors of the pancreas.

In this study, we reviewed our experience with IPAS in seven patients who underwent pancreatic resection. As a high-volume pancreatic center, we are referred patients with pancreatic masses of uncertain etiology. By reviewing our experience with the presentation, diagnosis, and treatment of patients with IPAS, in combination with review of the existing literature, we sought to develop recommendations for improved preoperative diagnosis.

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2. Methods

A retrospective chart review was performed of 123 patients who underwent distal pancreatectomy with or without splenectomy at our institution between 2004 and 2015. Charts were analyzed for presenting symptoms, pre-operative imaging, operative therapy, and final pathologic evaluation. A total of seven patients (5.7%) who underwent distal pancreatectomy for a mass that was determined to be IPAS on final pathologic evaluation were included in the study.

3. Case reports

3.1. Patient 1

A 71-year-old, otherwise healthy, white male presented to his primary care physician with a 6-month history of nausea, vomiting, and progressively worsening post-prandial right upper quadrant pain. The patient's physical exam was unremarkable. Right upper quadrant ultrasound demonstrated normal anatomy, minimal cholecystic sludge, and a negative sonographic Murphy's sign. Hepatobiliary (HIDA) scan revealed an ejection fraction of 39% (normal >35%). CT scan of the abdomen and pelvis with contrast demonstrated a solid 2.9 cm mass in the pancreatic tail as well as a 3.5 cm enhancing mass in the spleen. Fine needle aspiration of the pancreatic mass was negative for malignant cells. Serum CA19-9 was 7.2 U/mL and CEA was 0.5 ng/mL. At this juncture, the pancreatic mass was thought to represent a non-functioning pancreatic neuroendocrine tumor (PNET) and the splenic mass was thought to be a hemangioma. Given the suspicion for malignancy, the patient underwent a laparoscopic distal pancreatectomy and splenectomy as well as laparoscopic cholecystectomy. Final pathology confirmed an area of focal congestion in spleen representing a splenic hemangioma and revealed the pancreatic mass to be a collection of mature lymphoid tissue surrounding a central vessel suggestive of splenic tissue, making the diagnosis of IPAS. The patient's postoperative hospital course was unremarkable. He was discharged home on post-operative day 7 in good condition.

3.2. Patient 2

A 55-year-old white male with a past medical history significant for gastroesophageal reflux disease presented to the Emergency Department after choking on food and losing consciousness. Physical exam was unremarkable and the patient denied pain, nausea, vomiting, weight loss, or jaundice. Serologic evaluations revealed an elevated serum amylase level that prompted CT scan of the abdomen, which demonstrated cystic lesions in the pancreas. Endoscopic ultrasonography (EUS) revealed several cystic lesions in the distal pancreas from which mucinous fluid was aspirated. Fine needle aspiration demonstrated no evidence of malignant cells. An MRI of the pancreas demonstrated several cystic masses, the largest of which measured 5.9 × 4.4 × 4.4 cm. The MRI and EUS findings, taken together, suggested a side-branch intraductal papillary mucinous neoplasm (IPMN), but could not exclude cystadenoma or cystadenocarcinoma. Serum CA19-9 was 14 U/mL and CEA was 1.3 ng/mL. Given the concern for malignancy, the patient underwent a laparoscopic-assisted distal pancreatectomy and splenectomy. Intraoperatively, the pancreatic masses appeared to be cystic tumors surrounded by necrotic material, suggestive of underlying pancreatitis. The cysts themselves contained mucinous material. Pathology revealed two ruptured epithelial inclusion cysts associated with accessory splenic tissue and no evidence of malignancy. The patient had an unremarkable postoperative course and was discharged home on post-operative day 5.

3.3. Patient 3

A 38-year-old white male with a past medical history significant for Type 2 diabetes mellitus, obesity, and chronic pancreatitis secondary to hypertriglyceridemia presented to his gastroenterologist for evaluation of persistent post-prandial nausea and abdominal pain. The patient had repeatedly undergone pancreatic stenting over the preceding three years, but had experienced minimal relief of his gastrointestinal symptoms. Prior endoscopic retrograde cholangiopancreatography (ERCP) evaluations had demonstrated a pancreatic duct (PD) stricture in the head of pancreas, which had been dilated repeatedly. The patient had also undergone hepatopancreatic sphincterotomy. His most recent ERCP demonstrated persistent PD stenosis for which he underwent balloon dilation and 7F 12 cm plastic stent placement. Pancreatogram showed a severely diseased duct, worse in the tail, with dilated proximal duct and normal side branch profile. Upper EUS revealed sonographic changes consistent with severe chronic pancreatitis in the head, body, and uncinate process, as well as esophageal and gastric varices. Liver biopsy was performed to rule out cirrhosis. At this point, the patient was referred to a pancreaticobiliary surgeon for further management of severe chronic pancreatitis, since he had failed endoscopic management. Preoperative CT of the abdomen and pelvis without contrast demonstrated a lobulated mass measuring 3.9 × 7.4 cm between the lesser curvature of the stomach and the anterior aspect of the pancreas. This was thought to represent a chronic pseudocyst. No pathologically enlarged lymph nodes or focal liver lesions were detected. Serum CEA was 1.7 ng/mL. The patient subsequently underwent a partial pancreatectomy, pseudocyst resection, splenectomy, and longitudinal Roux-en-Y pancreaticojejunostomy with a Duval modification of Puestow procedure. Pathology confirmed a psuedocyst in the body of the pancreas. Unexpectedly, accessory splenic tissue was found in the distal pancreas, evidenced by a collection of mature lymphoid tissue. The patient was discharged from the hospital on post-operative day 10 but continued to have symptomatic pancreatitis 12 months following procedure.

3.4. Patient 4

A 72-year-old, otherwise healthy, African American male presented to the ED with acute abdominal pain. He denied diarrhea, flushing, diaphoresis, wheezing, or tachycardia. Physical exam was unremarkable except for abdominal tenderness. A CT scan without contrast demonstrated nephrolithiasis. Incidentally, he was found to have a 2 cm round, well-circumscribed lesion in the tail of his pancreas, suspicious for PNET (Fig. 1). There was no evidence of metastatic disease. Fine needle aspiration was negative for malignant cells. Based on the above findings, the pancreatic mass was thought to represent either a non-functional PNET or IPAS. Given the inability to exclude malignancy, the patient underwent diagnostic laparoscopy and a laparoscopic Warshaw-type splenic-preserving distal pancreatectomy. Intraoperatively, an 8-mm accessory spleen was found in the omentum. Examination of the distal pancreas also demonstrated IPAS. Immunohistochemistry confirmed the diagnosis, as the mass stained strongly positive for CD45, a marker of benign lymphoid tissue, but did not stain positive for either chromogranin A or synaptophysin. No malignant cells were evident on final pathology. The patient was discharged on post-operative day 4 in good condition and was recovering well at his 6 month follow-up.

3.5. Patient 5

A 58-year-old white female with a past medical history