Intrapancreatic Accessory Spleen

R. Meiler¹, K.-H. Dietl¹, K. Novák¹, C. Patzel²

¹Department of General, Visceral, and Thoracic Surgery, and ²Department of Radiology, Klinikum Weiden, Weiden, Germany

Intrapancreatic accessory spleen is a rare cause of pancreatic pseudotumors and is located in the pancreatic tail in approximately 1% to 2%. Accessory spleen itself is found in approximately 7% to 15% of the population. Our findings show a case of an intrapancreatic accessory spleen suspected for a malignancy in the pancreatic tail. A 63-year-old man admitted for cholecystitis was incidentally diagnosed with a tumor at the pancreatic tail. On hyperintense magnetic resonance imaging, a solid mass of 1.5 cm in diameter in the pancreatic tail was seen, which contrasted as hyperdense in T2-weighted imaging. Because of inhomogeneous enhancement on the early vascular phase, the diagnosis of an endocrine pancreatic tail carcinoma was suspected. Intraoperatively, an accessory spleen was found in the pancreatic tail. An oncologic left pancreatectomy was performed because of a malignant tumor. Histology showed an intrapancreatic accessory spleen in the pancreatic tail that excluded the presence of cancer. In conclusion, intrapancreatic accessory spleen is a rare cause of unnecessary laparotomy, but the absence of reliable diagnostics for this entity make histologic ascertainment of a benign tumor indispensable. Therefore, we still needed an oncologic tumor resection.

Key words: Accessory spleens – Intrapancreatic ectopic localization – Differential diagnosis

Case Presentation

We report on a 63-year-old male patient who was initially admitted for epigastric pain that recurred for 2 to 3 months before hospitalization. His past medical history was uncomplicated and was noticeable only for a hepatic hemangioma; his family history was without any occurrences of malignancy. At his first admission to internal medicine, the patient was afebrile, had a pulse rate of 84 beats per minute, had a blood pressure of 150/90 mmHg, was...
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Int Surg 2010;95
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Intrapancreatic accessory spleen (IPAS) is a rare cause of pancreatic pseudotumors and is mostly located in the pancreatic tail. The frequency is 1% to 2% of all accessory spleens that may be found in about 7% to 15% of the population according to studies from autopsies.¹⁻⁴ Often, as in our case, those tumors, meant to be of neuroendocrine origin, are suspected to be malignant, which leads to unnecessary surgery. Well-defined diagnostic procedures are actually not present, rendering the differentiation from malignancies quite difficult.

Case Presentation

We report on a 63-year-old male patient who was initially admitted for epigastric pain that recurred for 2 to 3 months before hospitalization. His past medical history was uncomplicated and was noticeable only for a hepatic hemangioma; his family history was without any occurrences of malignancy. At his first admission to internal medicine, the patient was afebrile, had a pulse rate of 84 beats per minute, had a blood pressure of 150/90 mmHg, was...
in a good nutritional condition, but was in a reduced general condition. He presented with epigastric abdominal pain, with muscular defense in the upper abdomen. On abdominal examination there was no evidence of hepatosplenomegaly or ascites. Bowel sounds were normal. Blood investigations showed a bilirubin level of 1.4 mg/dL, lactase dehydrogenase level of 272 U/L, C reactive protein (CRP) of 152.8 mg/L, and leukocytes of 11.7 cells/nL at an elevated level, whereas other parameters had been within normal limits. On ultrasonography (US) we found a hepatic hemangioma, $3 \times 4.6 \times 4.8$ cm in diameter, subcapsular in segment VII, with a concrement of 1.8 cm in diameter in the gallbladder that showed florid signs of inflammation. An acute cholecystitis with known cholelithiasis was diagnosed and was first treated symptomatically and by antibiotic treatment. For a left-sided pain in the upper abdomen during hospitalization, a computed tomography (CT) scan was done for further investigation because of suspected biliary pancreatitis, although amylase was not elevated at 26 U/L. CT showed an intrapancreatic, nonspecific mass in the tail not measurable because of edema secondary to a minimal intense pancreatitis in the inflammatic setting of the gallbladder (Fig. 1).

Because of the undefinable result at CT scan, a magnetic resonance imaging (MRI; Fig. 2 and 3) was performed. The MRI visualized a solid mass of 1.5 cm in diameter in the pancreatic tail that was hyperintense in T2-weighted imaging and that showed an inhomogeneous enhancement on the early vascular phase, as is characteristic for neuroendocrine tumors.

A neuroendocrine tumor was feasible, so we performed an octreotide scan (189MBq 111In-Octreoid intravenously) for diagnosis, which was
negative. Chromogranin A of 83.2 μg/L, glucagon of 37.1 μg/mL, and 5-OH-indolacetic acid of 2.60 mg/24 h were within normal measures, which rendered a neuroendocrine tumor unlikely. Because we suspected a pancreatic neoplasm, a left pancreatic resection was performed that included cholecystectomy because of symptomatic cholecytolithiasis. At instantaneous section, the preparation turned out to be ectopic splenic tissue without any signs of malignancy (Fig. 4A, 4B, 4C, 4D, 4E). Therefore,
the resection was limited to the left side of the pancreas, so the preparation was 4.5 cm in length, 3.5 cm in width, and 2.3 cm in height, and it had a roundish, reddish brown area of 8 mm in diameter, which was intrapancreatic splenic tissue. The postoperative progress of the patient was uncomplicated, so he was discharged on day 8 after the operation.

Discussion

IPAS is, as mentioned above, quite rare, but it is, without any argument, a cause of unnecessary laparotomy. Different groups, such as Meyer et al8 and Dominguez et al,5 propose IPAS as a differential diagnosis in pancreatic neoplasms. This could be considered for pancreatic tumors without any other signs of cancer, such as with tumor markers or positive lymphatic nodes. However, is a diagnosis with the given procedures possible?

There are different approaches of diagnosis present in literature. Schreiner et al7 diagnosed three cases of accessory spleen in the pancreas by using an endoscopic ultrasound (EUS) – guided fine-needle aspiration (FNA) for biopsy and then by performing a CD8 immunostaining of splenic sinus endothelial cells to unveil the splenic tissue. Our question is about displacing tumorous cells in the prick channel in the case of pancreatic neoplasm and the effect on the course of disease. Pancreatic neoplasm, which has an unfavorable outcome, could eventually not be worsened by this procedure, but this may be of discussion.

Kim et al8 found another means of detection. The spleen has a characteristic image by comparing the precontrast and the contrast-enhanced phases in superparamagnetic iron oxide (SPIO) – enhanced MRI as well as in Levovist-enhanced US. Thus, an IPAS shows a significant signal drop, similar to the SPIO-enhanced T2- or T2*-weighted imaging, and shows prolonged enhancement on the delayed hepatosplenic phase of contrast-enhanced US; this could be differentiated from malignant pancreatic neoplasms.8 A contrast-enhanced ultrasound (CEUS) was also proposed earlier by Ota et al9 for diagnosis of those lesions. IPAS is a rare entity and so could be detected in a setting of a pancreatic tumor without any signs of a malignant lesion by using those procedures. The specificity of both methods is important, keeping in mind the deleterious effect of a wrong diagnosis in case of malignancy.

Miyayama et al10 describe computed tomographic arteriography (CTA) as helpful for diagnostic use. Here, the early enhancement of the lesion is similar to the splenic parenchyma. In two cases, Miyayama et al10 could show by multiplanar, reformatted images a deep cleft between the lesion and the pancreas, which, therefore, suggested that the lesion was originally extrapancreatic. Although this is nonspecific, this can only be a help at diagnosis; however, this cannot validate a lesion as having splenic origin.

Weiand et al11 have an approach that is based on physiology of splenic tissue. A scintigram with technetium-99–marked, heat-damaged red blood cells can detect splenic tissue and so can be a valuable diagnostic tool for ectopic splenic tissue. This seems to be the most specific procedure for differential diagnosis in IPAS-suspected lesions.

Thrombocytopenia and leukocytopenia can be due to accelerated degradation in the setting of accessory spleen, ending up in a sort of hypersplenism. Hence, the diagnostics, when platelets and leukocytes are low without other causes, should include the search of hidden accessory spleen tissue. Nevertheless, the current diagnostics are of low specificity and are not effectual enough in such a precarious situation of differential diagnosis. In addition, because the entity of IPAS is so small, it will be very challenging to validate this type of lesion as an important differential diagnosis in pancreatic tumors.

Conclusion

The incidence of intrapancreatic splenic tissue is low in comparison with the incidence of the solitary lesions in the pancreatic tail (e.g., neuroendocrine tumors). However, it is a rare cause of unnecessary laparotomy. In the situation of an intrapancreatic tumor with negative tumor markers, such as carcinoembryonic acid and CA 19-9, and with no signs of suspicious metastasis, intrapancreatic accessory spleen should be seriously discussed in preoperative differential diagnosis. However, the absence of reliable diagnostics for this entity makes histologic ascertainment of a benign tumor indispensable. Furthermore, the mentioned diagnostics are not available ubiquitously. In conclusion, we currently still need an oncologic tumor resection to prevent mistaking a malignancy for a benign lesion. Thus, we support laparotomy and pancreas resection as the gold standard when diagnostics are contradictory and uncertain in this special tumor case. Additional investigation has to be made to find diagnostics to avoid laparotomy in these cases.
References


