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**Case Report**

# Intrapancreatic Accessory Spleen

*A Rare Cause of a Pancreatic Mass*

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## Summary

**Conclusion: The clinical significance of intrapancreatic accessory spleens resides in the mimicry of pancreatic cancer. Radionuclide tests (Octreotide scan and Tc99m sulfur colloid scan) should be undertaken to distinguish these lesions from neuroendocrine tumors, hypervascular metastases and pancreatic carcinoma. If the tests are equivocal, diagnostic laparotomy or laparoscopy is recommended.**

**Background:** Despite its relatively common occurrence, intrapancreatic ectopic splenic tissue is rarely detected owing to its asymptomatic nature.

**Methods:** We report a case of a clinically asymptomatic patient in which abdominal computed tomography (CT) scans revealed a mass of 1.5 cm in diameter in the distal pancreas. The tumor markers CA 19-9 and carcinoembryonic antigen (CEA) were slightly elevated, and pancreatic neoplasm was suspected.

**Results:** Left pancreatic resection and splenectomy were performed. The removed specimen disclosed the presence of an accessory spleen within the pancreatic tail.

**Key Words:** Accessory spleen; ectopic spleen; pancreas; benign; tumor.

## Introduction

Ectopic splenic tissue in the abdominal cavity is a common entity, with a reported incidence of 10% in the general population *(1)*, and can be separated into two categories: splenosis resulting from autotransplantation of splenic tissue, usually after splenectomy; and accessory spleens, which are congenital duplications of splenic tissue in an ectopic location. Intrapancreatic location was found in about 16% of cases in autopsy series *(2)*, but it is rarely detected in clinical practice owing to a lack of symptoms. In contrast, the frequency of detected pancreatic mass on computed tomography (CT) scan is rising, and therefore, accessory spleen within the pancreas can be an important differential diagnosis in these patients. In this article, we describe a patient with accessory spleen in the tail of the pancreas where a pancreatic neoplasm was suspected initially. Treatment consisted of a left pancreatic resection.

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## Case Report

A 71-yr-old man was hospitalized in the dermatology department for exploration of a leukocytoclastic vasculitis. He had a known history of ethanol

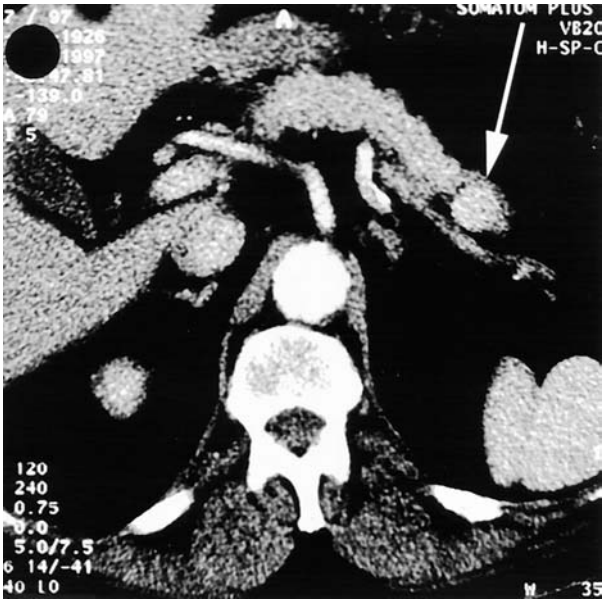


Fig. 1. CT scan of an accessory spleen in the pancreas (arrow) mimicking distal pancreatic neoplasm with homogeneous contrast enhancement. The hypervascular enhancing mass appears to be extrapancreatic and surrounded by fat in the posterior half of the lesion.

abuse of up to 1.5 L of beer/d and a nicotine abuse of 6 yr. During this stay, arterial hypertension was detected. Further investigations revealed an infrarenal aortic aneurysm of 4 cm in diameter and a significant stenosis of the right renal artery. In addition, an abdominal CT scan delineated a hypervascular, enhancing tumor formation of 1.5 cm in diameter in the pancreatic tail (Fig. 1). CT-controlled fine-needle aspiration failed because of the patient's lack of compliance. Therefore, he was referred to our department for surgical exploration.

On admission, he had no complaints, a normal nutritional status, and no jaundice. The abdomen was not tender to pressure, and no resistance or mass was palpable. With the exception of a raised erythrocyte sedimentation rate of 57 mm/h (normal: 20 mm/h), a creatinine value of 119 mol/L (normal range 59–116 mol/L), and an unspecific dysproteinemia, blood sample analyses were unremarkable. The tumor markers CA 19-9 and carcinoembryonic antigen (CEA) were slightly elevated with values of 27.2 kU/L (normal: <22 kU/L) and 3.1 mol/L, respectively (normal <1.5 mol/L); also, determination of the gastrointestinal hormones revealed no abnormal values. Endoscopic retrograde cholangiopancreaticography (ERCP) showed an unremarkable common bile duct and a normal-appearing pancreatic duct with regular side branches. At laparotomy, exploration of the pancreas demonstrated a normal head and body with an enlarged tail region of inconspicuous consistency. Based on the CT findings, a left pancreatic resection and splenectomy was performed to remove the mass completely. An incision into the removed specimen disclosed a round mass of 1.5 cm diameter consisting of spleen-like tissue completely surrounded by pancreatic tissue (Fig. 2). Frozen sections confirmed the presence of an accessory spleen within the pancreatic tail. The postoperative course was uneventful with the exception of intestinal pseudoobstruction, which was successfully treated by endoscopic decompression. The patient was discharged on the 17th postoperative day in good health.

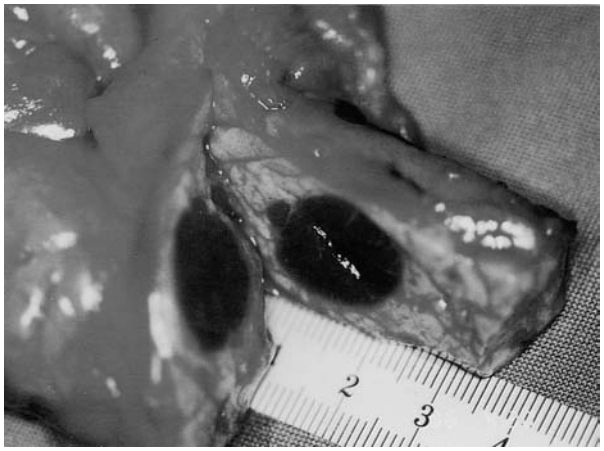

Fig. 2. Gross inspection of the resected specimen disclosed a round mass with a dark-red surface and a clear margin in the pancreatic tail. The suspicion of an accessory spleen was confirmed by microscopic examination.

## Pathological Findings

Pathological gross findings revealed a sharply delineated and encapsulated tumor of up to 1.5 cm in diameter, completely surrounded by lobular pancreatic tissue. There was no direct connection between this tumor and the resected spleen. In particular, the splenic hilum was normal, and there was no visible aberrant splenic artery. On sections, the tissue of this intrapancreatic tumor was dark red with numerous tiny gray dots. Histological examination

of this tumor mass showed the typical structure of splenic tissue with well-developed red and white pulp. A vascular hilum could not be detected, and blood supply consisted of small to medium-sized arteries and veins located within the capsule, an angiologic architecture typical of an accessory spleen. In the adjacent pancreatic tissue, adipocytes were increased with a clustering of islets around the accessory spleen's capsule, whereas the exocrine pancreatic tissue was poorly developed in this region (Fig. 3).

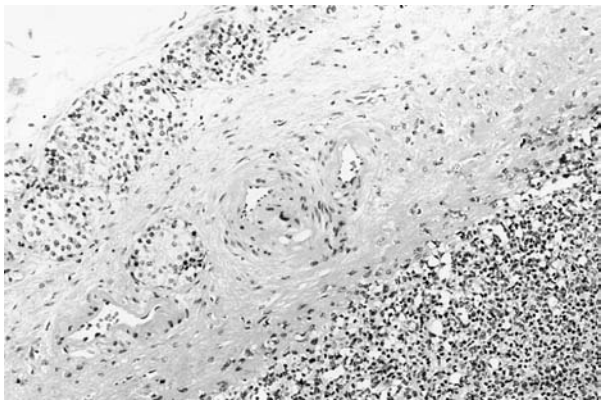

Fig. 3. Peripheral parts of the accessory spleen. Note that the splenic capsule contains small arterial branches, which are typical for this lesion. Adjacent to the capsule, there are a few pancreatic islets (hematoxylin and eosin staining, ×60).

## Discussion

Accessory spleens are characterized by a lack of vascular communication with the main spleen, a missing hilum, and a blood supply through small arteries within the capsule. They are usually located in the region of the splenic hilum, but in about 16% of cases in autopsy series, the accessory spleen is located within the pancreatic tail *(2)*. Particularly high rates of accessory spleens within the pancreas are found in individuals with trisomy 13, where it is one of the distinctive features of this very rare, but lethal genomic aberration *(3)*. In adult patients, accessory spleens within the pancreas are rarely detected owing to their asymptomatic nature. However, their clinical importance resides in the mimicry of pancreatic cancer, which has an increasing incidence in the Western World *(4)*. The specificity of CT scan for pancreatic cancer is rarely better than 80%, and even the combination of CT and ERCP does not allow definition or exclusion of malignancy in about 10% of patients *(5)*. The value of newer techniques, such as magnetic resonance imaging (MRI), has still not been clearly defined, but a prospective comparison of CT and MRI demonstrated that MRI was superior to CT at identifying small tumors *(6)*. However, data determining the accuracy of MRI in differential diagnosis of pancreatic tumors are still lacking. Thus, correct interpretation of a pancreatic mass represents a clinical challenge for the surgeon, especially in asymptomatic individuals.

In our patient, abdominal CT scans revealed the presence of a distal pancreatic mass, and the tumor markers CA 19-9 and CEA were slightly elevated. In contrast, ERCP was normal, and determination of the gastrointestinal hormones failed to demonstrate any alterations that would have raised suspicion of the presence of an endocrine tumor of the pancreas. In this unclear situation, we omitted further diagnostic tests, such as MRI, angiography, and scintigraphy, and performed a surgical exploration with subsequent histopathological examination, which enabled accurate diagnosis. In experienced centers, surgical resection can be performed with an acceptably low risk *(7)*. However, in this patient, a different diagnostic approach may have prevented an operation.

In retrospect, we propose the following diagnostic and therapeutic management: a nonenhancing mass that is hypervascular should for the most part be regarded as an adenocarcinoma of the pancreas until proven otherwise. In such instances, a fine-needle biopsy may be considered if surgery is precluded by clinical criteria. On the other hand, the diagnosis of hypervascular enhancing mass raises the possibility of a neuroendocrine tumor, a hypervascular metastasis or an ectopic spleen. A careful evaluation of other organ systems should provide an indication regarding the nature of any synchronous primary neoplastic disease. A neuroendocrine tumor may be investigated using an octreotide scan, which is accurate in up to 90% of instances, except in the case of insulinoma *(8)*. Under such circumstances, clinical symptomatology should allow for obvious diagnosis. If the possibility of an ectopic spleen, which is an extremely rare condition, is considered, a Tc99m-sulfur-colloid scan should provide an appropriate

diagnosis *(9)*. In the unlikely event of all such tests producing equivocal results, it may well then be necessary to consider a diagnostic laparoscopy or laparotomy to follow if a tissue diagnosis is necessary. Consideration of the patient's fitness for anesthesia and general surgery would obviously dictate the determination of therapy under these circumstances. An important note of clinical caution is the fact that the identification of hypervascular masses in the pancreas should be regarded as an indication for noninvasive study either by radioisotopic imaging techniques or, alternatively, by the use of angiography. Fine-needle aspiration under such circumstances is not warranted and potentially dangerous.

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