Case Report

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Intrapancreatic Accessory Spleen Mimicking a Non-Functioning Pancreatic Neuroendocrine Tumor: A Case Report

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We present a case of a 71-year-old woman with an intrapancreatic accessory spleen. The lesion was originally misdiagnosed as a non-functioning pancreatic neuroendocrine tumor. Therefore, she underwent laparoscopic, spleen-preserving distal pancreatectomy. Histopathological examination revealed an intrapancreatic accessory spleen. Although it is quite rare, this benign anomaly should be included in the differential diagnosis of distal pancreatic masses to avoid unnecessary surgery.

Key words: accessory spleen, pancreas, endocrine tumor, pancreatectomy, non-function

Introduction

An intrapancreatic accessory spleen (IPAS) is a rare, benign, congenital anomaly that is found in approximately less than 5% of the population.[1] It usually presents as a hypervascular tumor mimicking a neuroendocrine tumor. IPAS typically does not require any treatment; therefore, it is important to accurately differentiate it from other pancreatic lesions that require more aggressive treatment. Here we report a patient with IPAS, discuss its differential diagnosis, and provide suggestions for management of this lesion.

Case Report

A otherwise healthy 71-year-old woman presented with left epigastric pain. The pain was not associated with food, fevers, chills, night sweats, nausea, or vomiting.

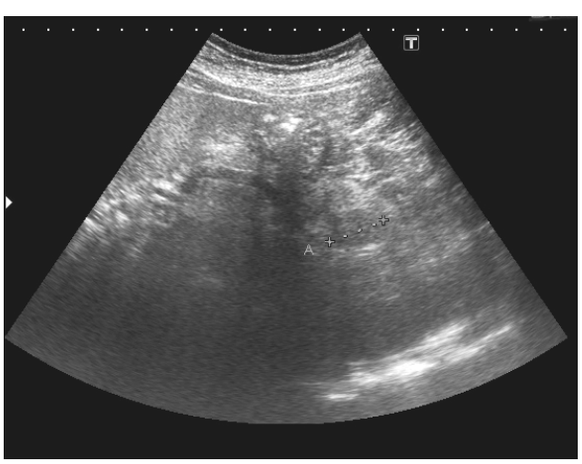

Fig. 1 Transverse ultrasound image showing a 1.8 cm × 2.3 cm, homogeneous, well-defined, round hypoechoic mass in the pancreatic tail.

Physical examination and laboratory data (including peripheral blood counts, blood sugar, and liver function tests) were unremarkable. Tumor markers, including CA19-9, CA125, and carcino-embryonic antigen, were

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within the normal range. Abdominal ultrasound revealed a 1.8 cm × 2.3 cm homogeneous, well-defined, slightly hypoechoic mass in the pancreatic tail (Fig. 1). Magnetic resonance imaging (MRI) revealed a 1.8 cm well-circumscribed solid lesion within the pancreatic tail. The lesion was hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging and showed greater contrast enhancement than the rest of the pancreatic tissue. However, the lesion exhibited signal intensity identical to that of the spleen in all magnetic resonance (MR) pulse sequences (Fig. 2). A non-functioning islet cell tumor of the pancreatic tail was suspected. Subsequently, the patient underwent laparoscopic, spleen-preserving distal pancreatectomy, with an uneventful recovery. Histopathological examination of the surgical specimen confirmed the diagnosis of IPAS.

Discussion

Accessory spleen is a relatively common congenital defect with a reported prevalence at autopsy of 10% – 30%. It is caused by the failure of fusion of the splenic anlage located in the dorsal mesogastrium.[2-4] The most common location is the hilum of the spleen followed by the pancreatic tail. In an autopsy study of 3,000 patients, an accessory spleen located in the pancreatic tail was reported to occur approximately 17% of the time.[4]

IPAS is a benign lesion related to an embryological aberration of splenic development. They are typically detected incidentally in patients undergoing evaluation for non-specific gastrointestinal symptoms. Moreover, preoperative imaging findings may mimic a pancreatic neuroendocrine neoplasm or pseudopapillary neoplasm.[5,6] An accessory spleen usually does not require treatment, and definitive diagnosis of IPAS is generally only confirmed by histopathological examination of the resected tumor. Potential complications associated with biopsy are a major impediment to establishing an accurate preoperative tissue diagnosis.[7] This results in the unnecessary resection of a benign pancreatic lesion in a vast majority of cases. Therefore, radiological modalities play an important role in the preoperative differential diagnosis of pancreatic lesions.

On sonography, IPAS usually appears as a small, well-defined, round, or ovoid mass that exhibits hypo-echogenicity compared with normal pancreatic parenchyma. Its texture is homogeneous, and it exhibits posterior enhancement. Additionally, its echogenicity is identical to that of the main spleen.[8]

Nuclear scintigraphy using technetium-99m-labeled sulfur colloid (SC) or 99mTc-labeled heat-damaged RBC (HDRBC) scan can identify a focal increased concentration of red blood cells. However, the small size of most IPASs and the low anatomic resolution of scintigraphy may limit its usefulness in this setting.

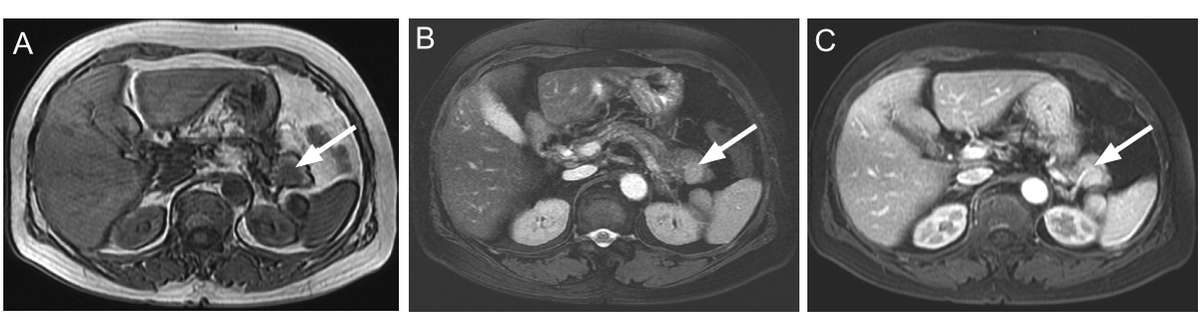

Fig. 2 Magnetic resonance imaging (A) Pre-gadolinium axial FSPGR T1-weighted image, (B) pre-gadolinium axial T2-weighted image and (C) post-gadolinium axial T1-weighted image with fat suppression showing a well-circumscribed, homogeneous, enhanced lesion within the pancreatic tail (arrows) matching the signal intensity and enhancing pattern of the spleen in all phases.

Mortelé et al.[9] reviewed abdominal computed tomography (CT) findings of 1,000 consecutive patients. They reported that most accessory spleens were found at the splenic hilum and found an IPAS in two patients. The characteristic findings suggest that accessory spleens on CT are small (< 2 cm), well-defined, round masses with homogeneous enhancement.

On MRI, an IPAS signal intensity is hypointense compared with the surrounding pancreatic parenchyma on T1-weighted imaging and hyperintense compared with the pancreas on T2-weighted imaging. The hallmark of IPAS is that its signal intensity is identical to that of the spleen on all MR pulse sequences.[10] However, IPAS should be distinguished from other hypervascular, well-enhanced, small pancreatic tail lesions, particularly neuroendocrine tumors. There are multiple imaging modalities and techniques, including CT, MRI, and nuclear medicine, that can help to differentiate IPAS from neuroendocrine tumors. Neuroendocrine tumors have a hypervascular appearance with low signal intensity on T1-weighted imaging, high signal intensity on T2-weighted imaging, and ring-like or homogeneous enhancement that does not match the density or signal intensity of the spleen on all phases and pulse sequences. An IPAS demonstrates a focal increased concentration of red blood cells on SC and HDRBC scans, whereas neuroendocrine tumors can be confirmed with octreotide scintigraphy because they contain somatostatin receptors.[10]

In conclusion, detection of an asymptomatic mass in the pancreatic tail, less than 2 cm in size, with characteristic radiological findings (well demarcated, round or ovoid, homogeneous enhancement, and signal intensity on all MRI pulse sequences identical to that of the spleen), should prompt consideration of IPAS in the differential diagnosis. A brief 2-month delay with subsequent follow-up may help avoid unnecessary surgery.

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