Intrapancreatic Accessory Spleen: The Usefulness of Arciform Arterial Enhancement for non-Invasive Diagnosis

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Dear Sir,

I read with interest the article entitled ‘Intrapancreatic accessory spleen: investigative dilemmas and role of EUS-guided FNA for diagnostic confirmation’ by Krishna et al. in the November 2011 issue of JOP. Journal of the Pancreas [1]. This well written case report and succinct review of the literature outlines the clinical and radiological dilemmas associated with this condition.

Intrapancreatic accessory spleen (IPAS) refers to a focal mass of ectopic splenic tissue located within the tail of the pancreas. It originates embryologically from failure of fusion of the splenic anlage in the dorsal mesogastrium. While found in 10-30% of post-mortem cases, Mortele et al. reported a prevalence of intrapancreatic accessory spleen of 2 in 1,000 consecutive patients that underwent abdominal CT imaging on older generation computed tomography (CT) scanners [2]. This radiological under-call is due to the small lesion size (a significant proportion of intrapancreatic accessory spleen measure 2 cm or smaller in size) making detection more difficult [3, 4]. However, the last decade has borne witness to a technological imaging revolution with rapid improvements in the diagnostic capabilities of cross-sectional imaging modalities such as CT and magnetic resonance imaging (MRI). As these powerful tools are able to detect and resolve lesions with increasing clarity and precision, this has led to a corresponding increase in the detection of incidentalomas on imaging. Many of these incidentalomas are small, asymptomatic and clinically occult. This leaves clinicians with the dilemma of what to make of these incidentalomas, their clinical significance, and how they should be appropriately managed. Within this context, it is reasonable to anticipate that the prevalence of intrapancreatic accessory spleen will increase due to improvements in imaging technology. As a benign asymptomatic condition, intrapancreatic accessory spleen does not require treatment and may be regarded as a ‘do not touch lesion’. The clinical and imaging dilemma lies in that intrapancreatic accessory spleen is frequently misdiagnosed for a pancreatic malignancy. In some cases, this may be due to poor awareness and understanding of the condition. In others, a working diagnosis of intrapancreatic accessory spleen is not entertained for the fear of missing a malignancy. This is regrettable as misdiagnosing intrapancreatic accessory spleen for a pancreatic malignancy leads to unnecessary patient anxiety and inappropriate patient management. This includes performing a distal pancreatectomy which subjects patients to unwarranted postoperative morbidity and risk of postoperative complications.

The differential diagnoses for a small solitary soft tissue mass in the pancreatic tail include intrapancreatic accessory spleen, pancreatic adenocarcinoma, neuroendocrine tumor and pancreatic metastases. These lesions can often be characterized and differentiated on imaging (Table 1). High resolution multiphasic contrast enhanced-CT and/or MRI, widely available in most centers worldwide, can render precise lesion characterization and differentiation. When compared to the spleen, intrapancreatic accessory spleen exhibits a similar attenuation and signal intensity on CT and MRI, respectively, and similar post-contrast enhancement (throughout all vascular phases) [4]. In particular, the arciform pattern of arterial enhancement within the intrapancreatic accessory spleen, similar to that seen in the adjacent spleen, is highly suggestive of
the diagnosis (Figure 1) [4]. The arciform pattern of arterial enhancement is secondary to perfusion differences between red pulp and white pulp present within tissues of splenic origin. This arciform enhancement helps to differentiate intrapancreatic accessory spleen from other focal arterially enhancing pathologies in the pancreatic tail such as neuroendocrine tumor (heterogeneous or ring-like arterial enhancement) and pancreatic metastases (heterogeneous arterial enhancement) from hypervascular neoplasms such as renal cell carcinoma. Solid pseudopapillary tumor may show mild heterogeneous arterial enhancement but it is not typically confused with intrapancreatic accessory spleen as it is generally large at presentation (mean size of 9 cm) [4]. Pancreatic adenocarcinoma is a hypovascular on CECT and CEMRI. Ancillary findings may include parenchymal and vascular invasion, regional adenopathy and distant metastases.

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The differential diagnosis for a small solitary mass in the pancreatic tail. Imaging findings on CT, MRI and nuclear medicine.

<table>
<thead>
<tr>
<th>Imaging modalities</th>
<th>CECT/CEMRI</th>
<th>Nuclear medicine</th>
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<tr>
<td><strong>Intrapancreatic accessory spleen (IPAS)</strong></td>
<td>IPAS typically shows CT attenuation, T1 and T2 MRI signal intensities and enhancement similar to that found in the spleen on all sequences (pre- and post-intravenous contrast)</td>
<td>IPAS typically shows radiotracer uptake similar to that seen in the spleen on $^{99m}$T-HDRBC and $^{99m}$T sulfur colloid</td>
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<td><strong>Presence of arciform arterial enhancement improves diagnostic confidence</strong></td>
<td>On SPIO-enhanced MRI, IPAS shows signal loss similar to that found in the spleen, on T2/T2*-weighted MRI images</td>
<td>Pancreatic adenocarcinoma and its metastases may shows high radiotracer uptake on $^{18}$FDG-PET making PET a valuable modality for staging the disease</td>
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<td><strong>Adenocarcinoma</strong></td>
<td>5-10% of pancreatic adenocarcinomas are located in the pancreatic tail. Adenocarcinoma is a hypovascular on CECT and CEMRI. Ancillary findings may include parenchymal and vascular invasion, regional adenopathy and distant metastases</td>
<td>Most neuroendocrine tumors, aside from some insulinomas, show high radiotracer uptake on $^{123}$I-octreotide imaging</td>
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<td><strong>Neuroendocrine tumor</strong></td>
<td>Neuroendocrine tumors have a variable biologic behavior and morphologic presentation. Tumors may be benign or malignant, small or large, symptomatic (functional tumors) or asymptomatic (non-functional tumors). Tumors may be solitary or multifocal (more common in syndromic conditions such as multiple endocrine neoplasia 1 and von Hippel Lindau disease) Neuroendocrine tumors are typically hypervascular and show heterogeneous or ring-like arterial enhancement followed by portal or delayed phase washout. Occasionally, neuroendocrine tumors may show delayed phase enhancement. When present, the parenchymal and lymph node metastases exhibit similar enhancement to that of the primary pancreatic neuroendocrine tumor</td>
<td>Some poorly differentiated neuroendocrine tumors (mostly non-functional tumors) may show high uptake on $^{18}$FDG-PET</td>
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<td><strong>Pancreatic metastases</strong></td>
<td>A past medical history of a primary extra-pancreatic malignancy should raise the clinical suspicion that a pancreatic mass may represent a metastasis</td>
<td>If the primary extra-pancreatic neoplasm is $^{18}$FDG-PET avid, then its pancreatic metastases will likely show radiotracer uptake on PET</td>
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Examples:
- Pancreatic metastases from renal cell carcinoma are typically hypervascular and show arterial enhancement.
- Pancreatic metastases from lung cancer are typically hypovascular and are hypo-enhancing compared to the normal pancreatic parenchyma.

CECT: contrast enhanced computed tomography; CEMRI: contrast enhanced magnetic resonance imaging; IPAS: intrapancreatic accessory spleen; $^{99m}$T-HDRBC: $^{99m}$T-technetium heat-damaged red blood cell; $^{18}$FDG-PET: $^{18}$fluorodeoxyglucose-positron emission tomography; SPIO: superparamagnetic iron oxide.
perform contrast enhanced ultrasound. When non-invasive diagnostic tests fail, fine needle aspiration biopsy employing image guidance (e.g., CT, ultrasound, endoscopic ultrasound) can be performed for cytological diagnosis [1]. However, this is not a full proof procedure and may yield unsatisfactory results due inadequate biopsy specimens and sampling error. The small size of intrapancreatic accessory spleen and deep location make it a challenging target to both access and biopsy. Furthermore, there is a small risk of biopsy related complications such as hematoma, pancreatitis and trauma to adjacent tissues. It is highly preferable if diagnosis can be made non-invasively, whenever possible.

In conclusion, I would like to reiterate that intrapancreatic accessory spleen, in most cases, can be confidently diagnosed non-invasively on high quality thin-slice multiphasic contrast enhanced CT or MRI. Intrapancreatic accessory spleen shows a similar attenuation, signal intensity and enhancement pattern with the spleen on all sequences. In particular, one should pay attention to the characteristic arciform pattern of arterial enhancement, a feature shared by intrapancreatic accessory spleen and the spleen. This pattern of enhancement permits differentiation of intrapancreatic accessory spleen from neuroendocrine tumors and other arterial enhancing neoplasms in the pancreatic tail, which show heterogeneous or ring like enhancement patterns. In patients with imaging findings characteristic for intrapancreatic accessory spleen, follow-up imaging at 6 and 12 months, for a 1-year duration, is helpful in confirming interval lesion stability consistent with this benign diagnosis.

**Conflict of interest** The study did not involve any grants and there are no disclosures or conflicts of interest.

**References**