World Journal of Surgery and Surgical Research

Case Report
Published: 16 Aug, 2022

Unmasking the Intrapancreatic Accessory Spleen

*Stefanie P Lazow[1] and Ketan R Sheth[2]**

[1]Department of Surgery, Beth Israel Deaconess Medical Center, Harvard Medical School, USA

[2]Department of Surgery, Cambridge Health Alliance, Harvard Medical School, USA

Abstract

Despite the availability of various diagnostic studies including CT, MRI, nuclear medicine images, and endoscopic ultrasound, the differentiation of Intrapancreatic Accessory Spleen (IPAS) from pancreatic neuroendocrine tumor remains challenging. We present the case of a 40-year-old male with an incidentally found intrapancreatic lesion with indeterminate imaging diagnosis. A CT demonstrated accessory spleens at the splenic hilum. Laparoscopic distal pancreatectomy with splenectomy was performed. Pathology resulted with benign IPAS. Despite indeterminate imaging, a high suspicion for IPAS should be maintained in patients with other accessory spleens.

Case Presentation

Initial presentation

The patient is a 40-year-old male with history of type 2 diabetes, hypertension, and obesity who presented to his primary care physician with a 50-pound weight loss after transitioning to a keto diet for diabetes management. This prompted lab work demonstrating a mild transaminitis. Abdominal ultrasound was performed without hepatobiliary anomalies that incidentally noted a left-sided abdominal mass. MRI abdomen showed a 2.3 cm × 1.8 cm pancreatic tail lesion (Figure 1A) with enhancement "not definitively" consistent with an accessory spleen though this could not be excluded. Subsequently, a Tc-99m sulfur colloid nuclear medicine liver/spleen scan with SPECT was performed. This demonstrated homogenous tracer distribution within the spleen and no evidence of abnormal radiotracer uptake at the splenic hilum or within the pancreas to suggest the presence of accessory spleens (Figure 1B).

The patient was referred to general surgery to discuss possible resection given the uncertain diagnosis. The differential included Intrapancreatic Accessory Spleen (IPAS) versus pancreatic Neuroendocrine Tumor (pNET). Endoscopic Ultrasound (EUS) was then performed re-demonstrating a 2.2 cm homogenous hypoechoic mass in the tail of the pancreas with smooth, well-defined borders and a normal pancreatic duct of 3 mm diameter. Fine Needle Aspiration (FNA) sampling, however, could not be performed due to the track of the splenic vessels in the sampling window of the needle.

Given ongoing diagnostic uncertainty, the options of surveillance versus resection were discussed. The patient elected for resection. A preoperative CT abdomen/pelvis re-demonstrated a 2.8 cm × 2.2 cm smooth mass extending posteriorly from the distal tip of the pancreatic tail with mildly reduced density compared to the spleen (Figure 1C). CT also noted several lesions near the splenic hilum consistent with accessory spleens (Figure 1D).

Surgery

The patient underwent laparoscopic distal pancreatectomy. After entering the abdomen and mobilizing the splenic flexure, the lesser sac was entered. Dissection was started along the inferior border of the pancreas working towards the splenic hilum, where a 2 cm nodule was resected. This was sent for frozen pathology, which resulted as an accessory spleen. Upon further inspection of the pancreas, an additional intra-pancreatic tail lesion was visualized so the pancreas dissection was continued (Figure 2). The splenic vein and artery were noted to be intra-parenchymal and could not be safely separated to attempt an enucleation. Therefore, a distal pancreatectomy with splenectomy was performed. The pancreatic resection was completed with an endo-GIA stapling device. The pancreatic margin was sent for frozen pathology, which was negative. A drain was left at the pancreatic staple line.

Pathology

The pathology resulted as a 1 cm IPAS with unremarkable background pancreatic parenchyma

OPEN ACCESS

Correspondence:
Ketan R Sheth, Department of Surgery, Cambridge Health Alliance, Harvard Medical School, 1493 Cambridge Street, Cambridge, MA 02139, USA, Tel: (617) 665-2555; E-mail: Ksheth@cha.harvard.edu

Received Date: *19 Jul 2022*
Accepted Date: *12 Aug 2022*
Published Date: *16 Aug 2022*

Citation:
Lazow SP, Sheth KR. Unmasking the Intrapancreatic Accessory Spleen. World J Surg Surgical Res. 2022; 5: 1400.

Copyright *© 2022 Ketan R Sheth. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.*

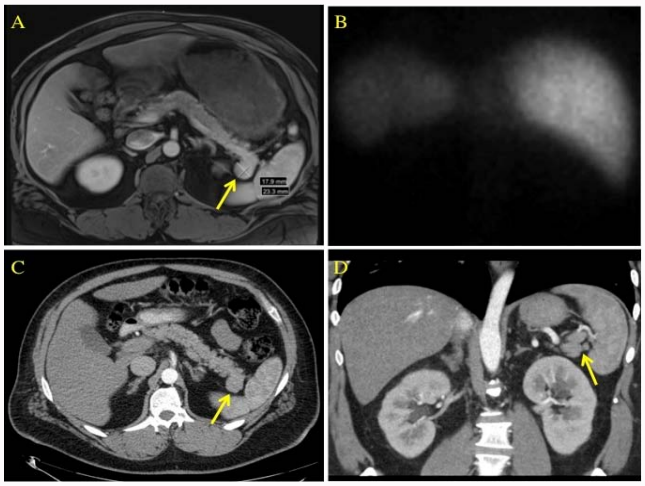

Figure 1A: This is an axial slice from the MRI abdomen (VIBE fat saturated post-contrast) showing a 2.3 cm × 1.8 cm lesion in the pancreatic tail without pancreatic ductal dilation.
Figure 1B: This is a representative image from the Tc-99m sulfur colloid nuclear medicine liver/spleen scan demonstrating homogenous tracer distribution within the spleen and no evidence of abnormal radiotracer uptake at the splenic hilum or pancreas.
Figure 1C: This is an axial slice from the CT abdomen/pelvis with IV contrast. The yellow arrow points to the mass extending posteriorly from the distal tip of the pancreatic tail.
Figure 1D: This is a coronal slice from the CT abdomen/pelvis with IV contrast. The yellow arrow highlights several small masses at the splenic hilum likely consistent with accessory spleens, though of mildly reduced density compared to the spleen.

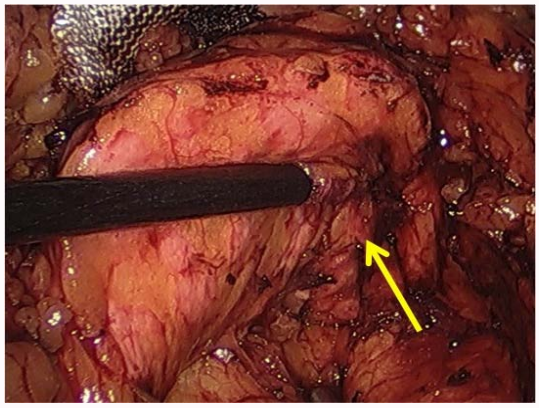

Figure 2: This image shows the pancreas lifted off the retroperitoneum after the dissection of the inferior border with an intrapancreatic mass noted at the tail (arrow).

without malignancy (Figures 3A-3B). There were also three accessory spleens present at the splenic hilum.

Postoperative course

The patient did well postoperatively without immediate complications. He received prophylactic subcutaneous heparin injections three times a day while inpatient. His diet was advanced without change in drain character. His glucose levels were well controlled. He received an inpatient pneumococcal vaccine with the remainder of his post-splenectomy vaccines coordinated outpatient. He was discharged on postoperative day six with the drain in place. Drain amylase was checked in clinic without concern for pancreatic fistula, and the drain was removed on postoperative day 13. He re-presented to the emergency room on postoperative day 16 with complaint of acute vision loss with exam consistent with left superior quadrantanopia. CT and MRI showed a right occipital stroke. TTE demonstrated an atrial septal defect though lower extremity duplex studies were negative for DVT. He was started on aspirin and discharged with occupational therapy.

Discussion

This patient was incidentally found to have an intrapancreatic mass of indeterminate diagnosis despite a variety of imaging modalities including CT, MRI, Tc-99m sulfur colloid, and EUS. EUS FNA could not be safely performed due to the lesion's proximity to the splenic vessels. CT was suspicious for multiple accessory spleens at the splenic hilum, but the Tc-99m sulfur colloid study did not confirm the presence of accessory spleens. Given diagnostic uncertainty and the patient's preference to avoid lengthy surveillance, the patient underwent laparoscopic distal pancreatectomy and splenectomy for definitive diagnosis and to rule out malignancy. Pathology resulted as IPAS, a benign lesion, which was inconsistent with the Tc-99m colloid results. The patient's postoperative course was ultimately complicated by stroke. Despite indeterminate imaging and a negative Tc-99m sulfur colloid test for accessory spleens, a high suspicion for IPAS should be maintained in patients with other accessory spleens suspected by other cross-sectional imaging. Furthermore, patients should be involved in shared decision-making processes to fully understand the options and various risks of surgical intervention.

While accessory spleens are reported in up to 30% of autopsy studies, the majority are found at the splenic hilum (75% to 80%) while only a minority are found within the pancreatic tail (20% to 25%.) [1]. A recent article reviewed 46 published case reports of IPAS in the literature [2]. In this review, IPAS lesions were most commonly described as solitary, solid, and well-defined lesions not exceeding 3 cm in the largest diameter and with similar contrast enhancement to the spleen [2-4]. Despite the availability of high-resolution CT and MRI imaging, the diagnosis of IPAS remains challenging given its similar morphologic and contrast enhancement characteristics to pNETs. In the setting of diagnostic uncertainty, other diagnostic adjuncts can be used including neuroendocrine tumor markers, nuclear medicine studies, and EUS.

While neuroendocrine tumor markers play a role in the diagnosis of a functioning pNET, they will not differentiate a non-functioning pNET from IPAS. These markers were not sent in our patient given he did not have concerning symptoms for a functioning pNET. Furthermore, while octreotide scintigraphy can detect pNET with 70% to 95% sensitivity, there are reported false positive results in the setting of IPAS because the lymphocytes in splenic tissue also have somatostatin receptors [3]. There are similar reported rates of false positive results with DOTATATE PET scans [5,6].

Nuclear medicine scans such as Tc-99m colloid or heat damaged red blood cell scintigraphy are the most sensitive and specific for the localization of splenic tissue and the diagnosis of IPAS [3]. However, their sensitivity can be reduced if lesions are less than 1.5 cm in diameter [7], are embedded in other background tissue such as pancreas [2], or have relatively low uptake of tracer compared to the spleen itself in patients who have not undergone prior splenectomy [7]. All of these factors applied to our patient and likely contributed to his non-diagnostic colloid study. In our patient, the Tc-99m colloid study also did not identify the accessory spleens at the splenic hilum that were ultimately confirmed on surgical pathology, though his

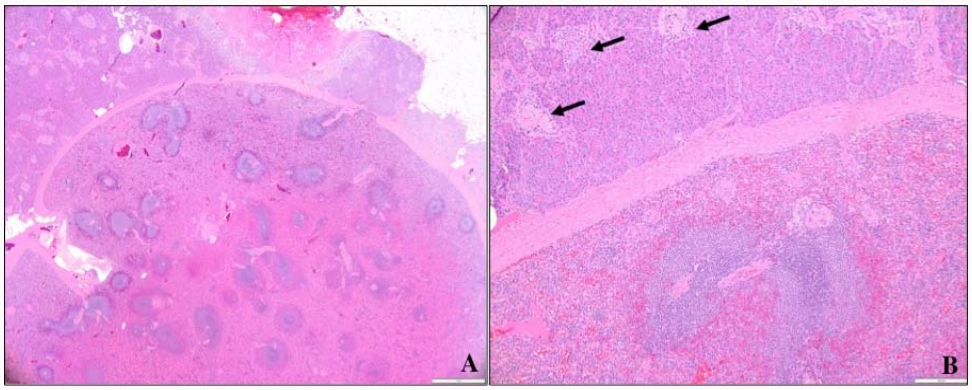

Figure 3A: The accessory spleen is a round, well-circumscribed nodule with a thin fibrous capsule. Benign lobular pancreatic parenchyma and fibroadipose tissue surround the accessory spleen. H&E, 20x magnification.
Figure 3B: The fibrous capsule is seen dividing the pancreatic acinar parenchyma (upper part of image) from the accessory splenic tissue (lower). Scattered islets of Langerhans, the endocrine component of the pancreas, are present (arrows). H&E, 100x magnification.

CT imaging and pathology ultimately demonstrated the presence of multiple accessory spleens in this location.

Another available imaging modality that was used in our patient given diagnostic uncertainty is EUS. While EUS has not been shown to consistently differentiate between IPAS and pNET based upon lesion morphology alone [8], EUS FNA sampling can be diagnostic for IPAS if there is positive CD8 staining to identify splenic endothelial cells [9]. Lymphocyte and/or platelet aggregates can also be seen in IPAS [9]. However, FNA sampling during EUS was not possible in our patient.

Alternative imaging modalities may be further explored to improve the diagnosis of IPAS and prevent unnecessary intervention for this benign pathology. A more recent study suggests that motion-corrected 68Ga-DOTATATE PET/CT scans can more accurately differentiate between IPAS and pNETs [10]. MRI with ferumoxides, which show preferential uptake in hepatic and splenic tissues given their rich reticuloendothelial composition, may also improve IPAS diagnosis [4].

Conclusion

Despite the availability of multiple imaging modalities, the diagnosis of Intrapancreatic Accessory Spleen (IPAS) remains challenging. It is important to maintain a high suspicion that an intrapancreatic lesion may represent IPAS in patients with accessory spleens identified in other locations to preclude diagnosis by surgical intervention and reduce postoperative morbidity.

References

1. Yildiz AE, Ariyurek MO, Karcaaltincaba M. Splenic anomalies of shape, size, and location: Pictoral essay. Sci World J. 2013;2013:321810.
2. Osher E, Scapa E, Klausner J, Greenman Y, Tordjman K, Melhem A, et al. Pancreatic incidentaloma: Differentiating nonfunctioning pancreatic neuroendocrine tumors from intrapancreatic accessory spleen. Endocr Pract. 2016;22(7):773-779.
3. Bhutiani N, Egger M, Doughtie C, Burkardt ES, Scoggins CR, Martin RCG, et al. Intrapancreatic Accessory Spleen (IPAS): A single-institution experience and review of the literature. Am J Surg. 2017;213(4):816-20.
4. Spencer LA, Spizarny DL, Williams TR. Imaging features of intrapancreatic accessory spleen. Br J Radiol. 2010;83(992):668-73.
5. Kripalani S, Patel V, Joneja U, Talwar S, Parikh M, Barshay V, et al. Intrapancreatic accessory spleen diagnosed as neuroendocrine tumor: The dangers of false positives and their implications in subsequent management. Cureus. 2021;13(6):e15891.
6. Lancelloti F, Sacco L, Cerasari S, Bellato V, Cicconi S, Ciardi A, et al. Intrapancreatic accessory spleen false positive to 68Ga-Dotatoc: Case report and literature review. World J Surg Oncol. 2019;17:117.
7. Ekmekci S, Diz-Kucukkaya R, Turkmen C, Adalet I. Selective spleen scintigraphy in the evaluation of accessory spleen/splenosis in splenectomized/nonsplenectomized patients and the contribution of SPECT imaging. Mol Imaging Radionucl Ther. 2015;24(1):1-7.
8. Kim GE, Morris JD, Anand N, DePalma F, Greenwald BD, Kim RE, et al. Recognizing intrapancreatic accessory spleen via EUS: Interobserver variability. Endosc Ultrasound. 2019;8(6):392-7.
9. Renno A, Hill M, Abdel-Aziz Y, Meawad H, Lenhard A, Nawras A. Diagnosis of intrapancreatic accessory spleen by endoscopic ultrasound-guided fine-needle aspiration mimicking a pancreatic neoplasm: A case report and review of literature. Clin J Gastroenterol. 2020;13(2):287-97.
10. Liberini V, Kotasidis F, Treyer V, Messerli M, Orita E, Engel-Bicik I, et al. Impact of PET data driven respiratory motion correction and BSREM reconstruction of 68Ga-DOTATATE PET/CT for differentiating Neuroendocrine Tumors (NET) and intrapancreatic accessory spleens (IPAS). Sci Rep. 2021;11:2273.