

Contents lists available at [ScienceDirect](http://www.sciencedirect.com)

International Journal of Surgery Case Reports

journal homepage: www.casereports.com

Post-splenectomy intrapancreatic accessory spleen mimicking endocrine tumor of the pancreas



Hong-xu Zhu, Wen-hui Lou, Tian-tao Kuang, Dan-song Wang*

Department of General Surgery, Zhongshan Hospital Fudan University, Shanghai 200032, China

ARTICLE INFO

Article history:

Received 10 September 2014
Received in revised form 30 October 2014
Accepted 10 November 2014
Available online 15 November 2014

Keywords:

Intrapancreatic accessory spleen
Pancreatic neuroendocrine tumor
Splenectomy
Pancreas

ABSTRACT

INTRODUCTION: Intrapancreatic accessory spleen is an uncommon congenital abnormality of the spleen with no indication for surgical intervention. Among the few cases reported, IPAS coexisted with a normal spleen. We here report the first case of IPAS arising a couple years after splenectomy with the appearance of an endocrine tumor of the pancreas.

PRESENTATION OF CASE: A 62-year-old female presented with a one-week history of left upper quadrant discomfort. She had splenectomy for the treatment of hypersplenism caused by cirrhotic portal hypertension two years before this admission. Her physical examination was unremarkable and laboratory data was within the normal range. Both the ultrasonography and magnetic resonance image revealed a small oval-shaped mass in the tail of her pancreas with the diameter 2 cm or less. A distal pancreatectomy was performed for the suspicion of malignant neuroendocrine tumor of the pancreas. An intrapancreatic accessory spleen was confirmed by the pathologic examination.

DISCUSSION: Intrapancreatic accessory spleen is one kind of congenital ectopic spleen without indication for operative intervention. We present the case to support that intrapancreatic accessory spleen may enlarge through a compensatory mechanism, and raise the awareness of this intrapancreatic entity to avoid unnecessary surgical operation.

CONCLUSION: IPAS should be highly considered as a differential diagnosis while the lesion is no more than 2.5 cm in diameter and/or other accessory spleens show around the splenic hilum.

© 2014 The Authors. Published by Elsevier Ltd. on behalf of Surgical Associates Ltd. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/3.0/>).

1. Background

1.1. Introduction

Accessory spleen is a congenital defect which is caused by the fusion failure of the splenic anlage during embryology. As the name suggests, it is called intrapancreatic accessory spleen (IPAS) when the accessory spleen locates within the pancreas. IPAS is a benign lesion and rarely causes any symptom, and requires no surgical treatment unless it is associated with hematological disorders such as idiopathic thrombocytopenic purpura. Therefore it is crucial to differentiate IPAS with neoplastic diseases of the pancreas. There are only a few cases of IPAS reported according to the literature and most were identified only after surgical resection, which is performed for suspected endocrine tumor. Here we present a case of IPAS mimicking endocrine tumor of the pancreas with a history of splenectomy.

1.2. Case presentation

A 62-year-old female with a history of hepatic cirrhosis and portal hypertension for which she had been previously submitted to surgery and had spleen removed. Cholecystectomy was performed for the treatment of cholecystolithiasis at the same time. Before this admission, she presented with left upper quadrant discomfort for one week. The patient had no other complaints, such as nausea, vomiting, jaundice, or symptoms of hypoglycemia. Physical examination was unremarkable and laboratory data including peripheral blood counts, blood glucose and tumor biomarkers was within the normal range.

The ultrasonography to investigate her abdominal symptom revealed a 17 mm × 15 mm hypoechoic mass posterior to the stomach and superior to the left adrenal gland. The abdominal magnetic resonance image (MRI) further confirmed the 16 mm × 12 mm lesion in the pancreatic tail. Compared with the pancreatic tissue, the intrapancreatic mass presented lower intensity on T1-weighted MRI and higher intensity on both T2 and diffused weighed images (Fig. 1). The lesion showed persisting enhancement during the late phases of dynamic MRI study. An extensive serum pancreatic hormone screen showed hormone levels within the normal limit after admission to our department. The computed tomography (CT)

* Corresponding author at: 180# Fenglin Road, Xuhui Area, Shanghai City 200032, China. Tel.: +86 13601675641.

E-mail address: zhuhongxu007@163.com (D.-s. Wang).

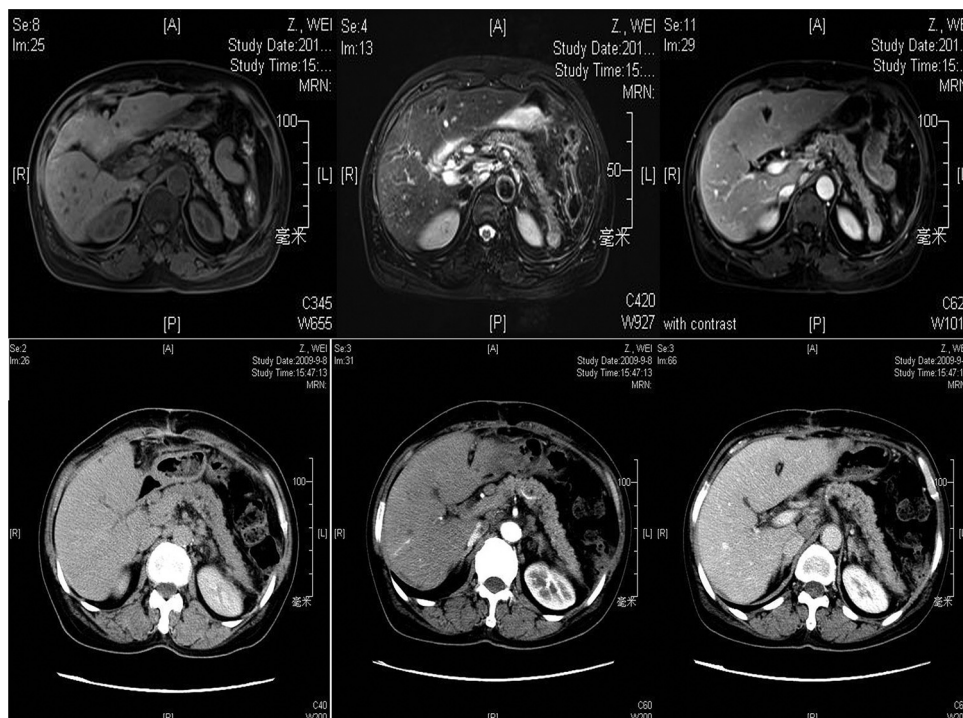


Fig. 1. Radiologic examination results. An intrapancreatic mass presented lower intensity on T1-weighted MRI and higher intensity on both T2 and late phase of dynamic MRI study. The CT scans detected no intrapancreatic space-occupying lesion after her splenectomy two years ago.



Fig. 2. Histologic staining of the mass showing a spleen surrounded by pancreatic tissue (H&E stain, original magnification 40 \times).

scans after her splenectomy two years ago were also reviewed and no bump was shown on both of the plain and enhanced scans (Fig. 1). Thus, a non-functional pancreatic neuroendocrine tumor (PNET) was suspected after completion of this workup, and distal pancreatectomy was conducted because of the possibility of malignancy with the evidence that several slightly enlarged peri-pancreatic lymph nodes were revealed on the MRI.

The surgical specimen contained a well-demarcated dark-red soft nodule, 2.0 cm \times 1.5 cm in size, surrounded by pancreatic tissue on macroscopic appearance. Microscopically, the nodule was composed of lymphoid follicles and sinusoid-like structures, and no apparent neoplastic component was founded (Fig. 2). Thus, an intrapancreatic accessory spleen was confirmed by pathologic diagnosis. The patient recovered from surgery without complications and was discharged five days later. Both the platelet count and

blood glucose level were within the normal limits on her follow-up visit three months after the operation.

1.3. Discussion

IPAS is an uncommon congenital abnormality of the spleen with the incidence around 2%.¹ It is a benign lesion without indication for operative intervention. However, in the majority of the cases reported from the literature, IPASs were resected surgically with no therapeutic effect, because they were difficult to make an accurate diagnosis preoperatively and were usually misdiagnosed as hypervascular pancreatic neoplasms, such as PNETs.

The incidence of PNETs ranges up to 15% of pancreatic neoplasms in surgical series. Nearly 70% of PNETs are classified as nonfunctional and all the PNET are malignant with the exception of minute incipient neoplasms that occur in some syndromes like type-1 multiple endocrine neoplasia.^{2,3} The initial imaging study used to identify a PNET is a high-quality contrast-enhanced multi-detector CT scan. PNETs are typically hyperdense and spherical on the arterial phase of imaging. MRI is more sensitive in the detection of smaller PNETs. PNETs are especially well visualized on T1 and T2-weighted images with fat suppression and typically have high signal intensity on T2-weighted images.⁴ The multimodality treatment for PNET includes surgery, hepatic-directed techniques and systemic chemotherapy. The overall 10-year survival rate of PNET ranges from 60% to 70% as reported.³

It is essential to differentiate nonfunctional PNETs from IPASs because surgery is the preferable form of management for the former while the latter usually require no operative intervention. Unfortunately, there has been no special laboratory test with great usefulness to confirm or exclude the diagnosis of IPAS as yet. Current image techniques may provide useful information in the differentiation between the two kind of lesions. Compared with the spleen, the IPAS more frequently shows homogeneous enhancement and signal intensity than the PNET does on dynamic CT scans and MRI respectively. However, it remains difficult to accurately

evaluate heterogeneous enhancement in IPAS smaller than 1 cm in diameter by using contrast-enhanced CT or conventional MRI. Jang et al.⁵ evaluated the diffusion-weighted MRI (DW-MRI) imaging in differentiation of an IPAS from a small solid pancreatic tumor and found that combined review of both DW-MRI with apparent diffusion coefficient maps and conventional morphologic MRI can prevent the misdiagnosis of IPAS. Superparamagnetic iron oxide-enhanced MRI is a sensitive and specific modality for the diagnosis of IPAS. Another noninvasive and specific test for detecting ectopic splenic tissue is ^{99m}Tc scintigraphy which is based on the phagocytosis of ^{99m}Tc-labeled red blood cells by macrophages of the reticuloendothelial system in the spleen.⁶

Except for the non-invasive methods mentioned above, EUS-guided fine-needle aspiration biopsy is an invasive approach to confirm the diagnosis of IPAS. Since Schreiner et al. reported the first case of IPAS diagnosed by EUS-FNA, there have been a few case reports of ectopic spleen diagnosed by EUS-FNA cytology.⁷ The predominant cytological features of IPAS include a polymorphous population of lymphocytes, traversing small vascular structures, and CD8 positive immunostaining of endothelial cells in cell block sections. As immunostaining of CD8 is negative in systemic endothelial cell and hemangioma but specifically highlighted in endothelial cells of splenic sinuses, EUS-FNA with the aid of CD8 immunostaining may make it possible to diagnose IPAS preoperatively and thus avoid unnecessary surgery.^{7,8}

Although a handful of IPASs have been reported, here we report the first case of IPAS after splenectomy according to the literature. The surgeons did not find any visible or palpable lesions during the previous operation. The IPAS had not been found until the patient underwent MRI for following examination after his splenectomy two years later. This phenomenon can be explained as compensation according to the literature.⁹ In other words, the IPAS in our case did exist invisibly and enlarged after her splenectomy. We do not know whether the IPAS would continue to grow bigger if not been resected, but our rare case should raise the awareness of IPAS to avoid unnecessary surgery.

1.4. Conclusion

IPAS has no indication for operative intervention in the most cases, it is important to differentiate this benign lesion from pancreatic neoplasm to avoid unnecessary surgery. While the lesion is no more than 2.5 cm in diameter and other accessory spleens show around the splenic hilum, IPAS should be highly considered as a differential diagnosis.

Conflict of interest

All authors declare no competing interest.

Funding

The authors have no extra or intra-institutional funding to declare.

Ethical approval

Written informed consent has been obtained from the patient for publication of this case report and its accompanying images. A copy of the written consent is available for review by the Editor of this journal.

Author's contributions

Hong-xu Zhu reviewed the medical records and drafted the manuscript; Wen-hui Lou and Tian-tao Kuang assisted with interpretation and presentation of radiological images and pathological specimens; Dan-song Wang helped in study design and manuscript edition; Wen-hui Lou and Tian-tao Kuang performed the operation.

All authors read and approved the final draft submitted.

References

- Halpert B, Gyorkey F. Lesions observed in accessory spleens of 311 patients. *Am J Clin Pathol* 1959;**32**:165–8.
- Rha SE, Jung SE, Lee KH, Ku YM, Byun JY, Lee JM. CT and MR imaging findings of endocrine tumor of the pancreas according to WHO classification. *Eur J Radiol* 2007;**62**:371–7.
- Reid MD, Balci S, Saka B, Adsay NV. Neuroendocrine tumors of the pancreas: current concepts and controversies. *Endocr Pathol* 2014;**25**:65–79.
- Milan SA, Yeo CJ. Neuroendocrine tumors of the pancreas. *Curr Opin Oncol* 2012;**24**:46–55.
- Jang KM, Kim SH, Lee SJ, Park MJ, Lee MH, Choi D. Differentiation of an intrapancreatic accessory spleen from a small (<3-cm) solid pancreatic tumor: value of diffusion-weighted MR imaging. *Radiology* 2013;**266**:159–67.
- Ota T, Tei M, Yoshioka A, et al. Intrapaneatic accessory spleen diagnosed by technetium-99m heat-damaged red blood cell SPECT. *J Nucl Med* 1997;**38**:494–5.
- Schreiner AM, Mansoor A, Faigel DO, Morgan TK. Intrapaneatic accessory spleen: mimic of pancreatic endocrine tumor diagnosed by endoscopic ultrasound-guided fine-needle aspiration biopsy. *Diagn Cytopathol* 2008;**36**:262–5.
- Lin J, Jing X. Fine-needle aspiration of intrapancreatic accessory spleen, mimic of pancreatic neoplasms. *Arch Pathol Lab Med* 2010;**134**:1474–8.
- Dodds WJ, Taylor AJ, Erickson SJ, Stewart ET, Lawson TL. Radiologic imaging of splenic anomalies. *AJR Am J Roentgenol* 1990;**155**:805–10.

Open Access

This article is published Open Access at sciendo.com. It is distributed under the [IJSCR Supplemental terms and conditions](#), which permits unrestricted non commercial use, distribution, and reproduction in any medium, provided the original authors and source are credited.