Clinical Case Report

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# Significance of imaging findings in the diagnosis of heterotopic spleen—an intrapancreatic accessory spleen (IPAS)

## Case report

Bin Yang, MS[a], Bimbadhar Valluru, MD[a], Ya-ru Guo, MS[a], Chunmao Cui, MS[a,*], Ping Zhang, MS[b,*], Wenshuai Duan, MS[a,*]

**Abstract**

**Rationale:** Accessory spleen (Heterotopic/Ectopic) or splenunculus has been attributed to the failure of the fusion of splenic primordial buds-derived from dorsal mesentery (mesodermal mesenchymal in origin) during the 5th week of embryonic organogenesis or to an extreme degree of splenic lobulation with pinching off of the spleen tissue. The most common locations for accessory spleens are the hilum of the spleen followed by adjacent to the tail of the pancreas. The patients usually present with no clinical symptoms.

**Patient concerns:** A 49-year-old female undergoing a routine medical examination- Abdominal Ultrasound revealed a pancreatic mass. She was admitted into the hospital for 3 days and was put under observation. There are no specific findings during the physical examination or any related abnormalities in the laboratory investigations.

**Diagnosis:** Heterotopic spleen—an intrapancreatic accessory spleen (IPAS).

**Interventions:** Noncontrast CT of the abdomen demonstrated a soft tissue mass with a clear boundary in the tail of the pancreas. On contrast examination—the arterial phase, it was markedly enhanced, homogenous congruity similar to that of spleen; on magnetic resonance imaging (MRI)-T2WI with fat suppressed sequence, it demonstrated a regular round clear edged mass in the pancreatic tail. On Diffusion Weighted Imaging (DWI), a mass with a clear boundary was observed within the parenchyma of the pancreatic tail. The mass showed a high signal on noncontrast MRI, while on contrast examination, the mass showed a strengthening signal with homogenous enhancement as that of spleen.

**Outcomes:** Heterotopic spleen presentation is a very rare asymptomatic clinical condition. During the routine medical examination - it presents mostly as a solitary benign round or oval mass with a clear boundary or as an ectopic focus, either in the pancreatic tail or adjacent to the pancreatic appendage, as an incidental finding. On Contrast CT, it shows as a homogeneously enhanced density- a strengthening mass lesion, in the pancreatic tail, similar to that of spleen.

**Lessons:** Our case emphasizes the importance of recognizing IPAS radiological characteristics and typical variations in its presentation in an asymptomatic patient that could help the personnel to differentiate it from other mass lesions. Thus, recognizing imaging findings on Plain CT, Contrast CT and MRI plays a key role to form a conclusive diagnosis of an accessory spleen, which has to be clinically associated. So, surgeons should consider IPAS as a differential for which unnecessary resection and an unintended surgical procedure can be avoided.

**Abbreviations:** CT = computed tomography, IPAS = intrapancreatic accessory spleen, MRI = magnetic resonance imaging, PET = positron emission tomography.

**Keywords:** case report, imaging diagnosis, intrapancreatic accessory spleen

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*[a] Department of Radiology, [b] Department of Pulmonology, the First Affiliated Hospital of Dali University, Dali, People's Republic of China.*

*[*] Correspondence: Wenshuai Duan, Ping Zhang and Chunmao Cui Department of Radiology, the First Affiliated Hospital of Dali University, Dali, People's Republic of China (e-mail: dwensh@163.com) [WD], (e-mail: ZhPing2910@sina.com) [PZ], (e-mail: cuichunmao@163.com) [CC]*



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## 1. Introduction

### 1.1. Patient

A 49-year-old female undergoing a routine medical examination—USG abdomen revealed a pancreatic mass. She was admitted into the hospital for 3 days and was put under observation. There are no specific abnormal findings during the physical examination or any related abnormalities in the laboratory investigations data. There were no obvious symptoms. Overall medical, family, and social history was unremarkable. There is no previous abdominal trauma or surgeries or hospitalization due to chronic illness.

## 2. Materials and methods

### 2.1. Imaging examination

All the material and methods were carried out in accordance with guidelines and regulations set by the Affiliated Hospital of Dali

University. All the protocols were approved by the Department of Radiology and Medical Imaging of Affiliated Hospital of Dali University. The patient provided the written consent on standard forms. Plain CT scan demonstrated a soft tissue mass, with a regular boundary, of dimensions 3.28 × 2.9 cm that can be clearly demarcated by its presence in the pancreatic tail (Fig. 1A). On Triphasic contrast examination—arterial phase, it visualizes a markedly enhanced lesion, a homogenous enhancement pattern similar to that of spleen, no peri pancreatic fat stranding is observed; no wash out is seen on late images (Fig. 1B–D).

Precontrast magnetic resonance imaging (MRI), T1WI fat-saturated gradient-echo shows a well-defined mass of low signal intensity that is surrounded by a higher signal intensity pancreas (Fig. 2A). T2WI sequence with fat suppression signals, showed a clear edged round mass in the pancreatic tail, with high intensity, of dimensions 3.28 × 2.9 cm; with a clear boundary within pancreatic parenchyma (Fig. 2B). On DWI, the mass showed a markedly high signal (Fig. 2C). After contrast with injection of gadolinium, the mass demonstrated moderate homogenous strengthening signals similar to that of spleen. The lesion was of similar signal intensity to adjacent spleen on each MR image (Fig. 2D–G).

## 3. Results

### 3.1. Surgery and histopathology

Surgery revealed a nonmovable hard and capsulated mass of size 3.0 × 2.5 cm, located in the tail of the pancreas. The splenic vasculature was isolated and secured with a ligation. The mass was separated from the pancreas and the retro peritoneal tissue, isolated from the pancreatic appendage, and performed a complete successful resection of the mass (Fig. 3A). Postoperative microscopic histopathological examination revealed dark red nodules, splenic tissue was encapsulated within the pancreatic tissue (Fig. 3B) and was concluded as an "accessory spleen."

## 4. Discussion

Intrapancreatic accessory spleen (IPAS) is a congenital anomaly of the splenic tissue[1] formed by the failure of the fusion of splenic primordial buds derived from dorsal mesentery (mesodermal mesenchymal in origin) during the 5th week of embryonic organogenesis or to an extreme degree of splenic lobulation with pinching off of the splenic tissue.[2] It can be mistaken for a hypervascular pancreatic tumor in both CT and MRI so a definitive diagnosis is difficult on radiological findings. Recognition of IPAS and being aware of its presentation and radiological characteristics is important. The most common locations for accessory spleens are the hilum of the spleen followed by adjacent to the tail of the pancreas.[3] Accessory spleens may also be the site of recurrent disease in patients following complete splenectomy because of hematological disorders causing hypersplenism or because of lymphomas. Heterotopic splenunculus or an accessory spleen is a very rare clinical presentation, where patients usually have no clinical symptoms, though occasionally patients present with abdominal pain, or nausea-vomiting with other nonspecific clinical scenarios. During inadvertent physical or medical examinations, it is observed as a benign mass, which is followed-up, generally does not require resection. With low incidence rates, preoperative clinical diagnostic criteria still remain unstudied. However, with the help of diagnostic imaging modalities, it has to be differentiated, particularly if hyper trophic, from other pancreatic tumors-benign pancreatic mass lesions-such as pancreatic endocrine tumors (PETs), solid pseudo papillary tumors, left-sided adrenal tumors, multiple endocrine metastatic neoplasia-pancreatic cancers like insulinoma, gastrinoma, and

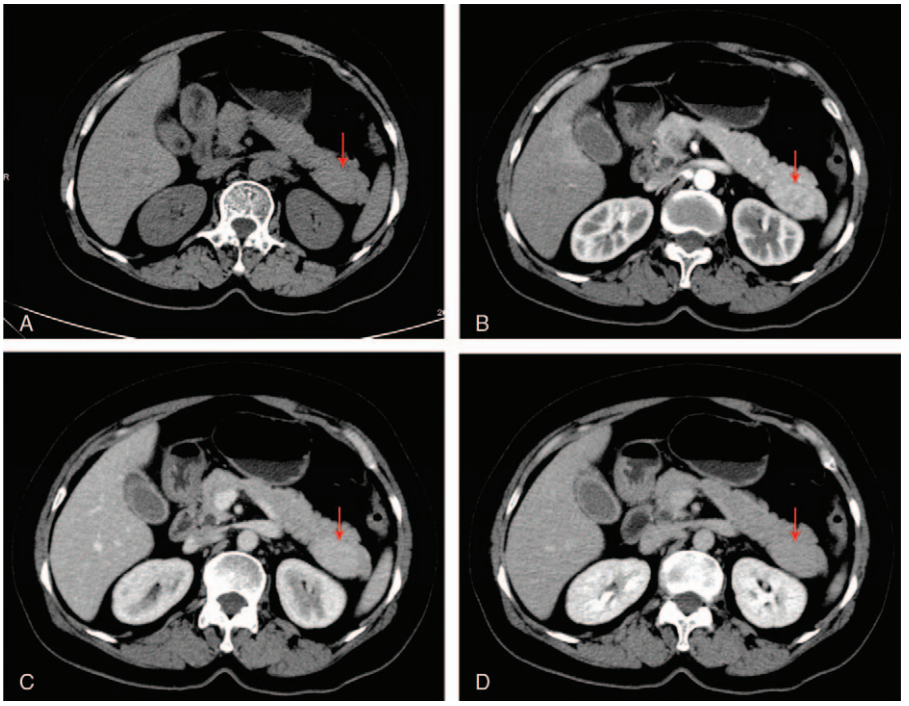


**Figure 1.** (A) A soft tissue mass, with a regular clear boundary, of dimensions 3.28 × 2.9 cm in the pancreatic tail. (B–D) Contrast examination-arterial phase, it showed a markedly enhanced lesion, a homogenous enhancement pattern similar to that of spleen.

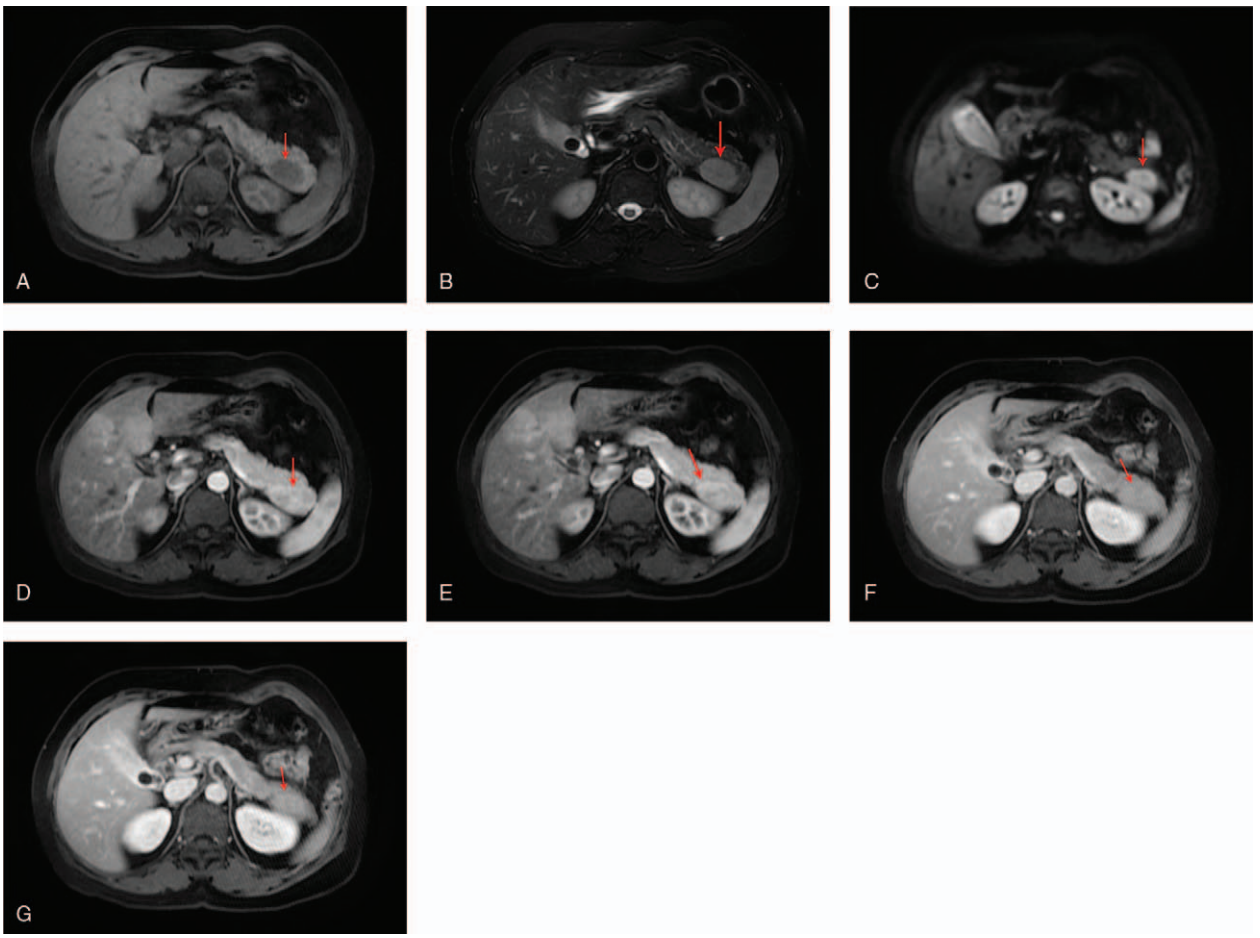


**Figure 2.** (A) MRI T1 WI sequence showing a well-defined mass of low signal intensity that is surrounded by a higher signal intensity pancreas. (B) T2WI sequence showed a clear edged round mass in the pancreatic tail mass with fat suppression signals, of dimensions 3.28 × 2.9 cm; with a clear boundary within pancreatic parenchyma. (C) On DWI, the mass showed a markedly high signal. (D–G) Enhanced sequences showed homogenous strengthening signals similar to that of spleen. MRI = magnetic resonance imaging.

glucagonoma, though they present with specific clinical symptoms.[4]

### 4.1. CT presentation

IPAS presents as solitary or multiple lesions or clustered lesion within the same location—round to oval, solid masses with a clear boundary of varying sizes with microscopic deposits to nodules ranging from few millimeters to centimeters in diameter, often traced within the pancreatic tail. CT scan demonstrated a highly enhanced mass, a high-density mass lesion, compared to pancreatic parenchyma, yet homogenously enhanced mass similar to that of splenic density. On contrast scan—arterial phase, it presents as a homogeneously enhanced density, similar

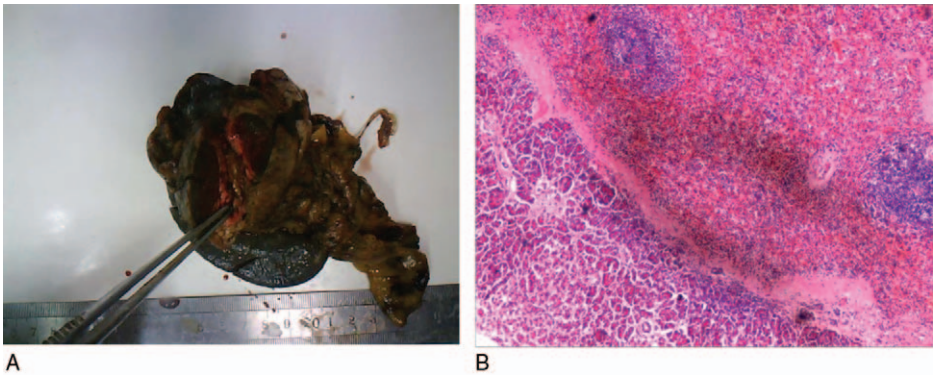


**Figure 3.** (A) Histopathological study—a dark red nodulated mass of dimensions 3 × 2 × 1.1 cm within tail of the pancreas (forceps). (B) Histopathology—spleen and pancreas substance, left lower section of the slide shows gross distribution of the splenic substance (in red), hyperemia, and atrophied tissue (in white). Right upper section of the slice shows a splenic substance encapsulated (dark red) within the pancreatic tissue. H&E (Hematoxylin and Eosin) Section 10 × 10.

to that of splenic pattern; in case of torsion or spontaneous rupture or infarction, rarely presented as a piebald enhancement comparable to small lesions with variable medullary-endothelial contents enhancement ratios. When in doubt, in the delayed phase, the granules are retained especially by heaptosplenic parenchyma allowing the clinician to differentiate between pancreatic tumor and accessory spleen. Arterial phase CT imaging provides significant diagnostic value to confirm IPAS when heterogenous, serpiginous enhancement of normal spleen is also observed. Such pattern is related to the vascular system of spleen and different flow rate through the cords of red and white pulp.[5,6] Apparently, CT arteriography 3D reconstruction of blood vessels shows principle artery is indeed a branch of splenic artery, which confirms the diagnosis of an IPAS.

### *4.2. MRI presentation*

On T1WI, IPAS presents a mass lesion, generally a low signal than that of pancreatic parenchyma, while on T2WI—a higher signal than pancreas, similar to that of splenic signal intensity, but sometimes may express signals higher than splenic intensity. DWI has no restricted diffusion—high signal intensity, which is due to changes in proportion of white and red matrix of the spleen. However, especially in the arterial phase, the lesions show proportionate signals that are always consistent with splenic parenchymal intensity. During the arterial phase, the enhancement degree is always similar to that of a normal splenic artery,[7] which is a significant observation for forming a conclusive diagnosis.

IPAS is a very rare clinical condition which has no obvious specific clinical symptoms.[8] Therefore, it is very important to recognize its imaging findings to avoid unnecessary biopsies and unintended surgical procedures. It is either located in the pancreatic tail or adjacent to the pancreatic appendage, as a solitary solid capsulated round to oval mass with clear boundary having a homogenous enhancement pattern similar to that of spleen; with varying degree of enhancements in Multi-phase contrast CT examinations should be appreciated. The converse of this is that if a mass does not enhance in the same fashion as the adjacent spleen, then the alternative diagnosis should be considered. In case of a suspicion, Superparamagnetic iron oxide based contrast enhanced MRI scan or 99mTC-HDRPC scan can be performed to confirm the diagnosis and remains 'Gold Standard'.[9] In many documented cases, IPAS was not considered and a pre-operative assessment for the differentials was not done.[10]

In conclusion, this is another case documentation of a presence of an IPAS. Our case emphasizes the importance of recognizing IPAS radiological characteristics and typical variations in its presentation in an asymptomatic patient that could help the personnel to differentiate it from other mass lesions. So, surgeons should consider IPAS as a differential for which unnecessary resection and an unintended surgical procedure can be avoided.

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