CASE REPORT

Intrapancreatic Accessory Spleen: A Differential Diagnosis to Neuroendocrine Tumors of the Pancreas on Ga-68-DOTATOC PET/CT

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ABSTRACT

A Seventy-three-year-old Caucasian woman underwent a contrast-enhanced computed tomography to clarify a deep venous thrombosis because of a swollen left leg. The CT revealed a contrast-enhancing 18x20 mm sized lesion in the pancreatic tail. It was suspected to be a neuroendocrine tumor and a Ga-68-DOTATOC-PET/CT showed a corresponding strong focal uptake. Laparoscopic spleen-preserving distal pancreatectomy was performed and histopathological examination diagnosed an intrapancreatic accessory spleen. The presence of IPAS should be kept in mind even in case of positive Ga-68-DOTATOC-PET/CT imaging to avoid unnecessary pancreatic resections.

INTRODUCTION

Ectopic splenic tissue can be a congenital anomaly and is formed during defective embryonic development when it rises from the left side of the dorsal mesogastrium as a result of imperfect fusion of separate splenic masses. On the other hand it can be a consequence of dispersed splenic tissue after surgery (e.g. splenectomy). Accessory spleens are found in about 10% of the population [1]. They are often located intra-abdominal, as nodules near the vessel pedicle of the main body of the spleen or adjacent to the pancreas. In some cases they are found within other organs like the pancreas or the liver [2]. The second most common location of accessory spleens is the pancreatic tail, the so called intrapancreatic accessory spleen (IPAS) [3, 4, 5]. About 68 cases of IPAS are published in the literature since 1980, as well as over 300 cases diagnosed postmortem in autopsy [6-8].

In radiological imaging IPAS appear as a solitary, well-encapsulated and hyper vascular lesion. On computed tomography (CT) and magnetic resonance imaging (MRI) IPAS often show the same characteristics as small neuroendocrine pancreatic neoplasm (pNEN). This is also valid for the sonomorphologic characteristics in endoscopic ultrasonography (EUS). Therefore, it can be challenging to distinguish IPAS from its main differential diagnoses pancreatic neuroendocrine neoplasm (pNEN) [9, 10]. It was recently suggested that Ga-68-DOTATOC-PET/CT is very specific for its enhancement in gastroenteropancreatic neuroendocrine tumors, because it binds specifically to somatostatin-receptors. Here we report the case of a Ga-68-DOTATOC-PET/CT positive IPAS that was misdiagnosed as non-functioning pNEN.

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CASE REPORT

A Sevety-three-year-old woman presented with a swollen painful right leg. The physical examination was otherwise unremarkable, the patient did not complain of any abdominal pain or discomfort. After excluding a deep vein thrombosis by a duplex sonography, an abdominal CT was performed to exclude a lymphatic stasis caused by adhesions after a partial right nephrectomy about ten years ago. The CT showed an 18x120mm contrast-enhancing lesion in the pancreatic tail, suggestive for a pNEN and also a tiny cyst of the pancreas head 9x7 mm **(Figure 1)**.

Laboratory work-up revealed normal values of CEA and CA19-9, pancreatic polypeptide, chromogranin A, gastrin, insulin and glucagon, proinsuline was lightly elevated (11.5 pmol/L, normal range <10 pmol/L). A Ga-68-DOTATOC-PET/CT was performed to further clarification. There revealed a strong focal uptake in the pancreatic tail sized 18x20 mm in diameter corresponding to the lesion visualized on the CT scan **(Figure 2)**, thus interpreted as nonfunctional pNEN.

To further establish the suspected diagnosis of a non-functioning pNEN an endoscopic ultrasound was performed. The slightly hyperechogenic, well-encapsulated lesion in the pancreatic tail was hardly to visualize and so the fine needle aspiration biopsy was not feasible under appropriate risk. A diffusion-weighted resonance imaging was not performed.

The case was discussed in the interdisciplinary tumor board and a recommendation for surgical resection was given for a non-functioning pNEN according to current guidelines [11]. The patient decided to undergo surgery and a laparoscopic spleen-preserving distal pancreatectomy was performed. Intraoperative inspection of the resected tumor gave the macroscopic impression of splenic tissue. Histopathological analysis confirmed an 18mm sized IPAS without any evidence for a pNEN **(Figure 3)**. The postoperative course was uneventful and the patient was discharged at postoperative day 8.

DISCUSSION

Accessory spleens can be found in more than 10% of the population. Approximately 80% are located in the splenic hilus and just around 17% of ectopic splenic tissue is located intrapancreatic. The second most common site

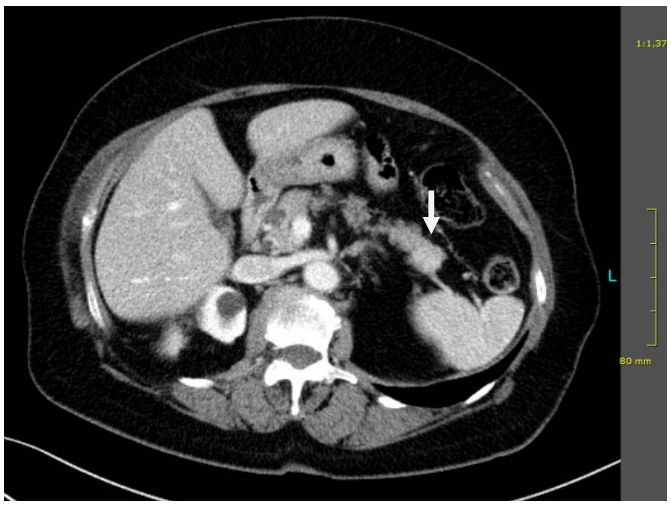

Figure 1. Contrast-enhanced CT scan (coronal plane) with Ultravist showing the homogenous intra-pancreatic lesion (arrow) in the tail of the pancreas with similar attenuation compared to the spleen

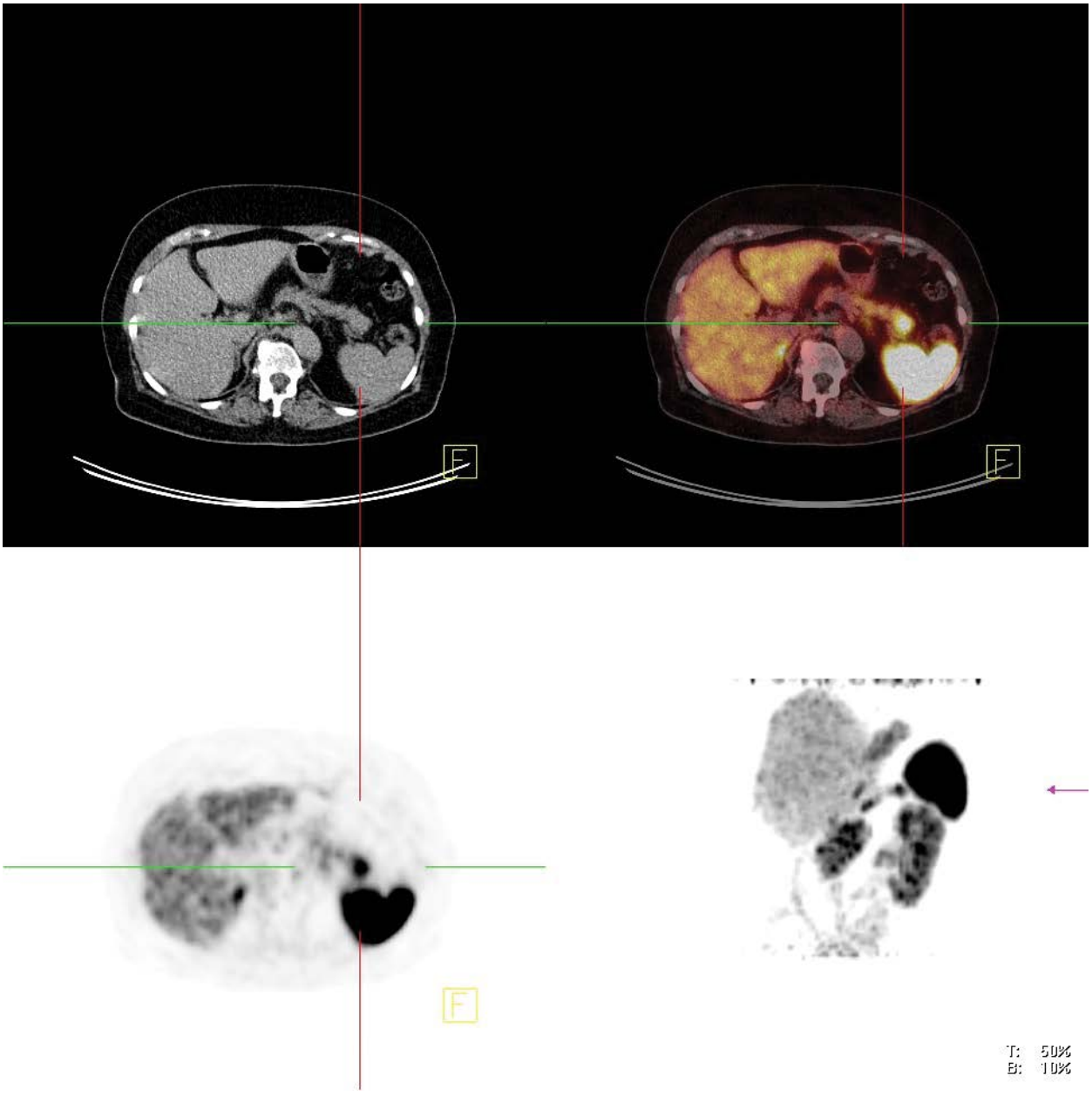

Figure 2. Ga-68-DOTATOC-PET/CT shows an 18x20mm contrast-enhancing lesion in the pancreatic tail. Next to physiologic uptake of Ga-68-DOTATOC-PET/CT in spleen, adrenals, kidneys and urine bladder there was a focal uptake, seen in the suspicious part of the pancreatic tail as seen in the CT scan (arrows).

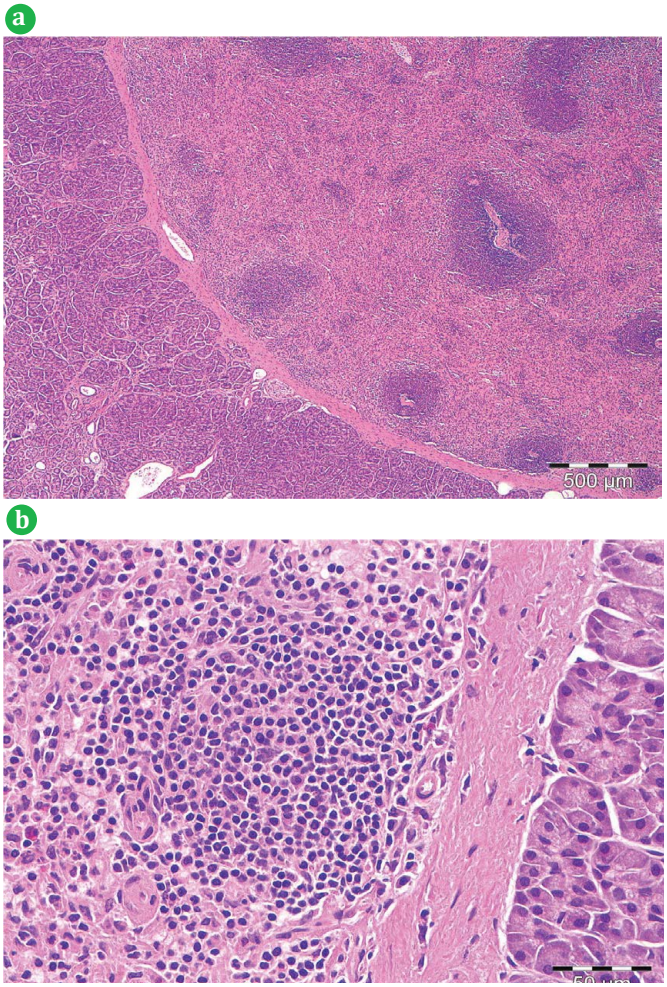

Figure 3. Histopathology confirming an intra-pancreatic accessory spleen.

A HE(40x) of IPAS. Microscopically, normal red and white pulp (right) is separated from the pancreatic tissue (left) by a fibrous capsule.

B HE (400x) of IPAS showing heterotopic splenic tissue with typical white and red pulp (left), thin fibrous capsule and pancreatic acinar tissue (right).

of splenic tissue is the pancreatic tail and IPAS are most frequently described as incidentally during imaging for non-pancreatic symptoms. On imaging like MRI, CT and EUS IPAS have similar characteristics as hyper vascular pNEN [12, 13]. In the presented case the IPAS was also positive on Ga-68-DOTATOC-PET/CT which was yet described in 3 other reports [3, 14, 15, 16, 17]. The fact that IPAS can be positive on Ga-68-DOTATOC-PET/CT should be kept in mind to avoid unnecessary pancreatic surgery.

Diffusion-weighted MRI is a good tool to differentiate between IPAS und pNEN. An IPAS normally appear in MRI hyper intense on T2-weightened sequence and hypo intense on T1-weightened sequence compared to the tissue of the pancreas. In diffusions weighted MRI IPAS appear in 92% is intense to the spleen whereas 84% of pNENs are hypo intense compared to the spleen [18]. Unfortunately, in the present case diffusion-weighted MRI was not performed, because of the Ga-68-positive PET/CT.

EUS with fine-needle biopsy is also a good tool to establish the diagnosis of IPAS [19, 20, 21] but it depends on the investigator's experience and the IPAS location. In the present case the IPAS was hardly to visualize in the splenic hilus and fine-needle biopsy was not feasible under appropriate risk to get tissue for further examination.

A Ga-68-DOTATOC-PET/CT is considered a very specific tool to establish the diagnosis of pNEN. However, as mentioned above, this is the fourth report for a Ga-68-positive IPAS. This is founded in the expression of somatostatine receptors in a relevant amount in the central nervous system as well as lymphocytes and activated lymphocytes. So, there is always a physiological accumulation of Ga-68-DOTATOC in splenic tissue [22, 23, 24].

Actually there is a new tracer labelled with copper (Cu-64). This tracer accumulates in mitochondria of lymphatic tissue. So this could be another possibility to differentiate between a pNEN and splenic tissue, if such a PET/CT is available [25]. Additionally you get information about the oxygenation of the tumor [26].

Bhure *et al*. reported about a contrast enhancing lesion in the pancreatic tail seen on abdominal CT-scan, also positive in Ga-68-DOTATOC-PET/CT and misinterpreted as pNEN. Pathology revealed the diagnosis of an IPAS [3]. Unfortunately the tumor is not specified any more. Barber *et al*. [16] also described a 16mm sized intrapancreatic lesion with a focal uptake in Ga-68-DOTATOC-PET/CT. After a MRI and a Tc-99m-Nanocoll-Szntigraphy it was classified as IPAS and no resection was performed. Also Collarino *et al*. reported about an incidentally detected lesion in the pancreatic tail with an intense tracer uptake in Ga-68-DOTATOC-PET/CT. Because this lesion was similar to the splenic tissue in MRI they performed a Tc-99m-Nanocoll-Szintigraphy that showed ectopic spleen and prevented the patient for unnecessary surgery.

Thus, if it is not possible to clarify the presence of IPAS by DW-MRI, EUS with fine-needle biopsy or Ga-68-DOTATOC-PET/CT a Tc-99m-Nanocoll-Szntigraphy should be considered [9, 27]. Tc-99m-Nanocoll-Szintigraphy is most specific imaging methods for diagnosing ectopic splenic tissue [28-30].

In conclusion IPAS will be a more frequent differential diagnosis of small nonfunctioning pNENs given by the widespread used CT- and MRI imaging. Thus, IPAS should be ruled out by diffusion-weighted MRI, EUS with FNB or Tc-99m-Nanocoll-szintigraphy even in the case of a Ga68 PET/CT positive lesion to avoid unnecessary pancreatic resections.

Conflict of Interest

All authors declare no conflict of interests.

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