

## Case Report

# Intra-pancreatic Accessory Spleen Mimicking Pancreatic Neuroendocrine Tumor on 68-Ga-Dotatate PET/CT

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## Abstract

Neuroendocrine tumors (NETs) are rare tumors, but the incidence is increasing with new diagnostics. A 37-year-old man was admitted to our hospital for an incidental 17-mm nodule in the tail of the pancreas. PET/CT shows indeterminate mass in the pancreatic tail with enhanced uptake of 68-Ga-dotatate. NET was suspected and laparoscopic distal pancreatectomy was performed. Pathologic examination revealed an accessory spleen with a heterotopic location. To the best of our knowledge, this is the first intrapancreatic accessory spleen (IPAS) case in which the positive 68-Ga-dotatate uptake reported in the literature. Our case showed that IPAS is one of the reasons of false positive involvement of 68-Ga-dotatate PET/CT. When PET/CT shows an indeterminate mass in the pancreatic tail with enhanced uptake of 68-Ga-dotatate, surgeons should keep IPAS in their mind for differential diagnosis to avoid false treatment.

**Keywords:** 68-Ga-dotatate PET/CT, intrapancreatic accessory spleen, pancreatic neuroendocrine tumors

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## Introduction

Neuroendocrine tumors (NETs) are very rare tumors, but incidence is increasing with new diagnostics.<sup>1</sup> Neuroendocrine cells are present in nearly all organs, so a primary NET can arise practically any tissue in the body. Approximately one third of gastrointestinal NETs are placed in the pancreas (31% – 34%) presenting with or without clinical symptoms.<sup>2,3</sup> Most NETs do not secrete any hormones<sup>4</sup> and NET are often unspecific symptoms, such as abdominal discomfort or abdominal pain.

Here, we describe a 37 year old man with an incidental 17-mm nodule in the tail of the pancreas found during his work-up for gastroesophageal reflux disease. Associated 68-Ga-dotatate PET/CT result was initially suspected of showing pancreatic NET, but the result was accessory spleen with a heterotopic location in pancreatic tissue. To the best of our knowledge, this is the first IPAS case in which the positive 68-Ga-dotatate uptake mistakenly led to a diagnosis of pancreatic NET.

## Case Report

A 37-year-old man was admitted to our hospital for an incidental 17-mm nodule in the tail of the pancreas. The patient had an unremarkable family, medical, and social history. Physical

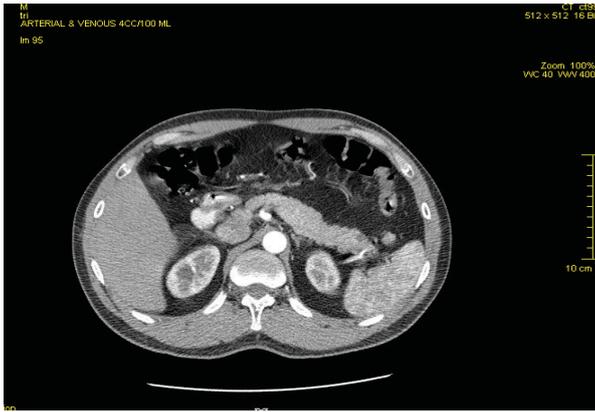
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examination and laboratory data, including peripheral blood counts, blood sugar and liver function tests were all unremarkable. Tumor markers, including Carbohydrate antigen 19–9 (CA19–9), Carbohydrate antigen 125 (CA125), Carcino-Embryonic Antigen (CEA), Alpha-Feto Protein (AFP) and Chromogranin A levels (38 ng/mL) were within the normal range. Abdominal tomography (CT) reveals a well-defined nodular lesion, with 1.7 cm of diameter, on the tail of the pancreas, showing slightly more contrast enhancement in the arterial phase and isodense in the venous and delayed phases which was suspected for neuroendocrine tumor (Figure 1). Endoscopic ultrasonography (EUS) showed a well-delineated nodule, about 1.6 cm in size, relatively homogeneous with well-defined and smooth margins in the tail of the pancreas. However, biopsy couldn't be performed due to localization of the mass. PET/CT shows indeterminate mass in the pancreatic tail with enhanced uptake of 68-Ga-dotatate. Neuroendocrine tumor of the pancreatic tail was suspected by the NET council and council was decided to surgery. Laparoscopic distal pancreatectomy was performed. The surgical resected material was sent to the pathology laboratory for frozen section confirmation of the tumor. Cut surface revealed a round, well-demarcated, smooth, dark-red nodule, 17 mm in diameter, surrounded by pancreatic tissue (Figure 2). Frozen section showed an intrapancreatic accessory spleen in the pancreatic tail, excluding the presence of neoplasia. Permanent sections confirmed the presence of an accessory spleen within the pancreatic tail. Thus, the mass was diagnosed as an Intrapancreatic Accessory Spleen (IPAS), (Figure 3).

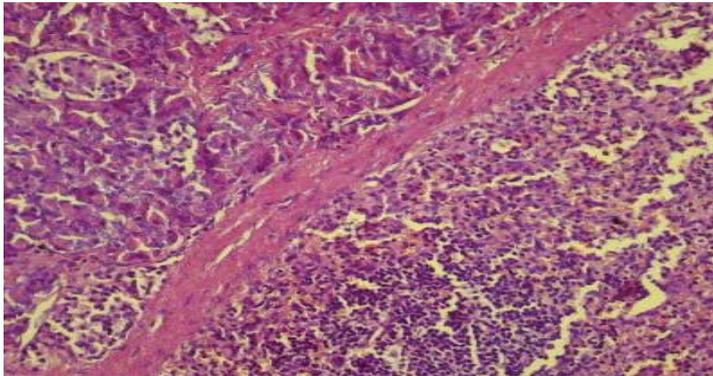
A 68-Ga-dotatate PET/CT images were acquired with a GE Discovery ST PET/CT scanner. PET/CT images were obtained 50 minutes after intravenous injection of 148 MBq 68-Ga-dotatate. Images from the vertex to the proximal femur were obtained while patients were in the supine position. PET images were acquired for 4 minutes per bed position. CT images were also obtained from the patient's integrated PET/CT with the use of a



**Figure 1.** Abdominal tomography (CT): A well-defined nodular lesion with a 1.7 cm of diameter on the tail of the pancreas



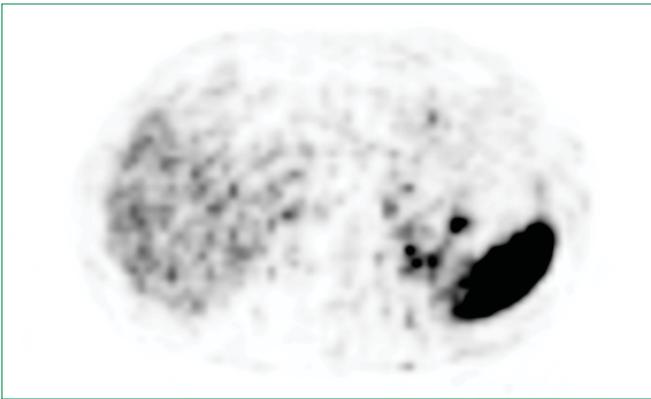
**Figure 2.** Gross pathology of IPAS in the tail of pancreas.



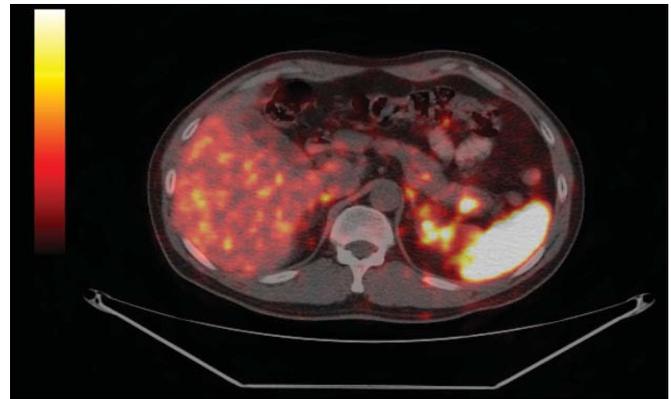
**Figure 3.** Histologic staining: An intra-pancreatic spleen surrounded by pancreatic tissue (hematoxylin-eosin)



**Figure 4.** A CT image from the patient's integrated PET/CT



**Figure 5.** The PET image that shows an indeterminate mass in pancreatic tail.



**Figure 6.** The PET-CT fusion image shows indeterminate mass in the pancreatic tail with enhanced uptake of 68-Ga-dotatate.

standardized protocol of 140 kV, 70 mA, tube rotation time of 0.5 s per rotation, and a pitch of 6 and a slice thickness of 5 mm (Figure 4). Emission PET images were reconstructed with an oral-contrast CT scan. Attenuation-corrected PET/CT fusion images were reviewed in three planes (transaxial, coronal and sagittal) on a Xeleris Workstation (GE Medical Systems). PET/CT images were evaluated and confirmed visually (Figures 5 and 6)

## Discussion

In our patient, 68-Ga-dotatate PET/CT result was initially

suspected of showing pancreatic NET, the result was accessory spleen with a heterotopic location in the pancreatic tissue.

The diagnostic technique for NET patients were imaging techniques like ultrasonography (US), computed tomography (CT), magnetic resonance imaging (MRI) and functional imaging, using octreotide somatostatin receptor scintigraphy (SRS) or 68-Ga-dotatate PET/CT.<sup>5</sup> Neuroendocrine cells secrete Chromogranin-A (CgA), has a sensitivity of approximately 70% – 85% in patients with known NET.<sup>6,7</sup> However, elevated CgA is not enough for diagnosis of NET, because treatment with proton pump inhibitors, atrophic gastritis, liver or renal

insufficiency, and other conditions can also increase the level of CgA.<sup>8,9</sup> Therefore, performing of the 68-Ga-labelled somatostatin derivatives is becoming interesting for NETs.<sup>10</sup> These compounds can be used for positron emission tomography (PET) imaging of tumors expressing somatostatin receptors (SSTR). Up to now five different SSTR (SSTR1–5) subtypes have been identified. NETs have a relatively high expression of SSTR2.<sup>11</sup> Currently, 68-Ga-dotatate, 68-Ga-dotatoc and 68-Ga-dotanoc are the most situated somatostatin receptor PET imagings.<sup>12</sup>

The detection of suspected NET accomplished with a high expression of SSTR with high sensitivity imaging using PET with somatostatin analogs such as 68-Ga-dotatate.<sup>13,14</sup> 68-Ga-dotatate PET/CT is commonly used in well-differentiated to intermediate differentiated NET whereas fluorodeoxyglucose (FDG) PET/CT is preferred for poorly differentiated NET.<sup>15,16</sup> In a study, 29 patients with suspected pancreatic NET, octreotide scintigraphy had a sensitivity of 54% and a specificity of 81%.<sup>17</sup> In another study, 68-Ga-dotatate PET/CT was ruled out a diagnosis of NET with specificity of 90%.<sup>18</sup> Haug, et al. reported that 68-Ga-dotatate PET/CT was identified NETs with a sensitivity of 81% (29/36), and excluded the presence of a NET with a specificity of 90% (61/68).<sup>19</sup>

Accessory spleen (AS), that is a congenital anomaly of splenic tissue,<sup>20</sup> results from the fusion failure of splenic anlage.<sup>21,22</sup> Accessory spleen, a relatively common congenital anomaly, is seen in 10% – 30% of patients at postmortem studies.<sup>20,21,23</sup> Splenic hilus is the most common site of an accessory spleen and followed by the pancreatic tail. In an autopsy study of 3000 patients, accessory spleen was found in the tail of the pancreas 61 of 364 patients (17%).<sup>24</sup> Intrapancreatic accessory spleen can be mistaken for a hypervascular pancreatic tumor in both CT and MRI imaging, so a definitive differential diagnosis is difficult on radiological findings.<sup>25,26</sup> Arterial phase CT imaging may be helpful when heterogeneous, serpiginous enhancement pattern of normal spleen is also observed within the IPAS. This characteristic pattern is likely related to the vascular system of the spleen and the different flow rate through the cords of the red pulp and white pulp.<sup>27,28</sup> Ota and colleagues merit consideration described two approaches when in doubt.<sup>29,30</sup> The first approach is photon emission CT with technetium 99-labelled red blood cells.<sup>29</sup> The second approach is contrast-enhanced ultrasonography using microgranules. In the late phase, the granules are retained almost exclusively by the hepatosplenic parenchyma, permitting the clinician to distinguish between an accessory spleen and a pancreatic tumour.<sup>30</sup> In our case, IPAS was not considered so that the preoperative assessment for the differential diagnosis of IPAS was not performed.

In Conclusion, this is the first IPAS case in which the positive 68-Ga-dotatate uptake mistakenly led to a diagnosis of pancreatic NET reported in the literature as we know. Our case showed that one of the reasons of the false positive involvement of 68-Ga-dotatate PET/CT is IPAS. When PET/CT shows an indeterminate mass in the pancreatic tail with enhanced uptake of 68-Ga-dotatate, surgeons should keep IPAS in their mind for differential diagnosis to avoid false treatment.

### Conflicts of interest

We declare that we have no conflict of interest and patient consent was obtained.

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