

Positive somatostatin receptor scintigraphy in accessory spleen mimicking recurrent neuroendocrine tumor

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We report the case of a female patient who had a neuroendocrine tumor in the pancreatic tail. Followup using ^{68}Ga DOTA-d-Phe(1)-Tyr(3)-octreotide (DOTATOC) positron-emission tomography (PET)/computed tomography (CT) detected a round, well-circumscribed nodular mass that exhibited positive somatostatin receptors. This finding was highly suggestive of an accessory spleen; however, due to the slight elevation of the tumor marker, recurrence of the tumor or lymph node metastasis of the endocrine tumor was considered as well. Ultimately, splenic scintigraphy confirmed an accessory spleen. This case shows the benefit of splenic scintigraphy (SS) in excluding a recurrent neuroendocrine tumor by confirming an accessory spleen.

Introduction

Accessory spleen is a normal anatomic variant that occurs in approximately 10% of the population. Several reports describe an intrapancreatic accessory spleen that was misdiagnosed as a nonsecreting endocrine tumor, since it exhibits positive somatostatin receptors (SSTRs). The accessory spleen poses a diagnostic challenge even in its extrapancreatic location, mimicking a lymph node metastasis or a local recurrence of a pancreatic neuroendocrine tumor. An accurate diagnosis is crucial, since such an accessory spleen does not require surgical treatment.

During the fifth week of gestation, the spleen develops in the dorsal mesogastrium from mesenchymal cells that migrate between the leaves of the mesentery and coalesce. An accessory spleen may arise from isolated cells that stay separated from the main body of the spleen. In a large series of nonselected autopsy investigations, an accessory spleen was found in 10–30%. (1, 2, 3). In 1,000 consecutive patients undergoing contrast-enhanced abdominal CT scan, an accessory spleen was present in 16% (1, 4). In 80%, the accessory spleen is located at or near the splenic hilum. The second most common site is the pancreatic tail (17%) (1, 3).

Case report

A 59-year-old woman underwent pancreatectomy and splenectomy as consequence of well-differentiated neuroendocrine tumor (NET) (pT3, N1, M0). Four years later, she was referred to our department for followup DOTATOC-scintigraphy after a slight elevation of chromogranin A. Other laboratory tests revealed normal CA 19-9 and carcinoembryonic antigen. However, somatostatin receptor scintigraphy using ^{68}Ga DOTATOC-PET/CT showed a positive uptake in a round mass in the upper left abdominal region (Fig. 1).

This finding could have been a sign of recurrence of NET, considering the elevation of chromogranin A. How-

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Positive SRS in accessory spleen mimicking recurrent neuroendocrine tumor

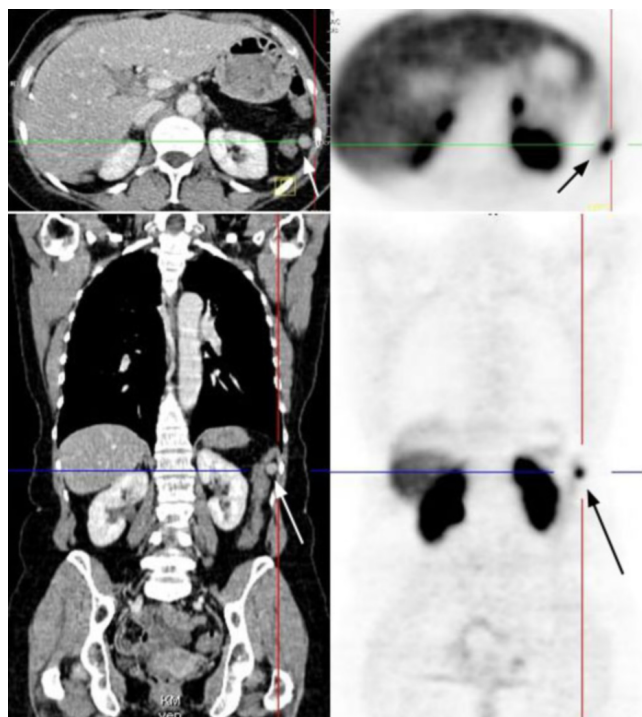


Figure 1. 59-year-old woman with accessory spleen. DOTATOC-PET/CT after intravenous injection of 206 MBq ^{68}Ga DOTATOC; the axial and coronal planes show an increased focal uptake that corresponds with a nodular lesion (diameter, 15 mm) in the left upper abdominal region (arrows).

ever, due to its location and round shape, we felt that an accessory spleen should be strongly considered in the differential diagnosis, especially after splenectomy.

The critical test that enabled us to determine the lesion's origin turned out to be SS. It demonstrated the ability of the described round mass to accumulate $^{99\text{m}}\text{Tc}$ -tagged heat-damaged red blood cells, which is a characteristic of splenic tissue (Fig. 2).

It is also worth mentioning that a $^{99\text{m}}\text{Tc}$ sulfur colloid scan with single-photon-emission computed tomography (SPECT) could accomplish the same goal as $^{99\text{m}}\text{Tc}$ heat-denatured red cells for this purpose.

Consequently, the recurrent NET was excluded in favor of an accessory spleen exhibiting positive SSTRs. This case shows the benefit of using SS as the method of choice in distinguishing SSTR-positive accessory spleen from recurrent NET.

Discussion

Somatostatin receptor scintigraphy (SRS) detects neuroendocrine gastroenteropancreatic (GEP) tumors with a high sensitivity (70% to 95%). It may detect primary and metastatic endocrine tumors not visualized by other imaging techniques, thereby affecting patient management (5).

Somatostatin is a small regulatory peptide that functions as a neurotransmitter and has inhibitory and antiproliferative effects. It binds to somatostatin receptors and has several identified subclasses: SSTR1, 2a, 2b, 3, 4, and 5.7. The commercially available somatostatin analog, pentetreotide, is labeled with ^{111}In Indium.

^{68}Ga Gallium somatostatin analogs, DOTATOC (DOTA0, D-Phe1, Tyr(3)-octreotate DOTATATE, and DOTA 1-Nal3-octreotide (DOTANOC) are widely used for PET imaging with promising results, but are not currently approved for clinical use in the USA (6). However, SRS may also lead to false-positive results; this is due to the presence of SSTRs on the surface of lymphocytes within the splenic tissue, which also attach SSTR ligands with a high affinity and produce images similar to a recurrent endocrine tumor (5).

In Prasad and Baum's study (7), the highest uptake of ^{68}Ga -DOTANOC was found in the spleen. Considering this result and many others, the spleen tissue (spleen and accessory spleen) can definitely be visualized with SRS.

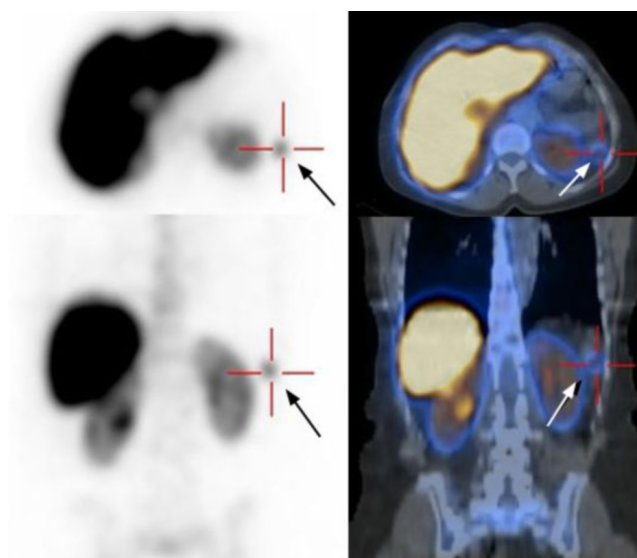


Figure 2. 59-year-old woman with accessory spleen. Splenic scintigraphy using $^{99\text{m}}\text{Tc}$ -tagged heat-damaged red-blood-cell scintigraphy, Scintigraphic image (left) and SPECT/CT fused image (right) demonstrate an uptake in the round nodule (arrows) that already exhibited a significant uptake of ^{68}Ga -DOTATOC, confirming the diagnosis of accessory spleen.

Several reports describe an intrapancreatic accessory spleen that was misdiagnosed as a nonsecreting endocrine tumor (8, 9); our report describes a possible misdiagnosis in case of extrapancreatic accessory spleen.

Since the accessory spleen exhibits positive SSTR, it poses a diagnostic challenge even in its extrapancreatic location, mimicking a lymph node metastasis or local recurrence of a pancreatic NET. However, definitive diagnosis

Positive SRS in accessory spleen mimicking recurrent neuroendocrine tumor

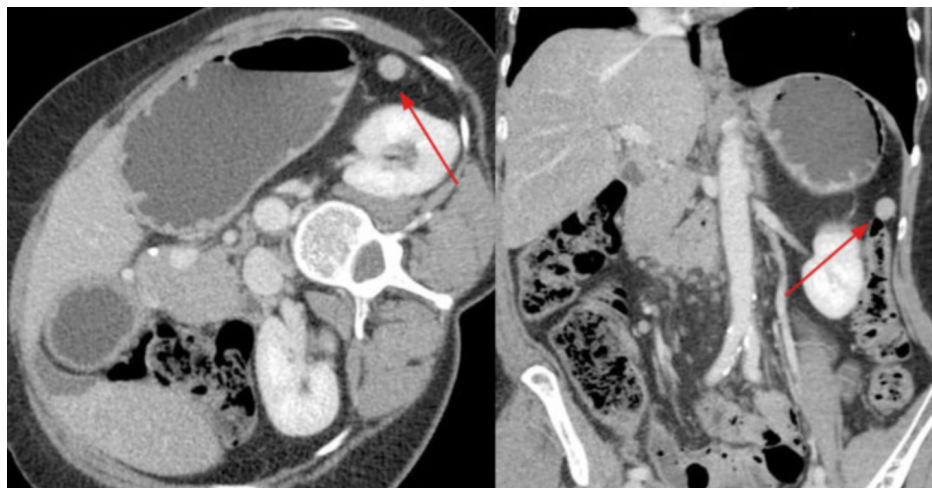


Figure 3. 59-year-old woman with accessory spleen. Oblique position modified contrast-enhanced abdominal CT. Modified oblique axial plane (left) and oblique coronal plane (right) show the confirmed accessory spleen, whose diameter is 15 mm (arrows).

based on conventional imaging modalities can be difficult because CT scan, MRI, and ultrasound images of such accessory spleens can be quite similar to those of recurrent tumor (Fig. 3). The optimal method is to identify the splenic tissue using ^{99m}Tc -tagged heat-damaged red-blood-cell scintigraphy, thus providing a simple, sensitive, and specific method for recognizing the accessory spleen.

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