

Case Report

Thoracic Splenosis Mimicking Metastatic Neuroendocrine Tumor (NET)

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ABSTRACT

A 64-year-old man presented with abdominal pain. CT scan showed a 5mm left ureter stone, mesenteric root mass as well as a left lung pleural-based lesion. Metastatic Neuroendocrine Tumor (NET) was suspected, hence a Gallium-68 Dotatate PET was performed. It showed intense uptake in the mesenteric mass, as well as in the pleural nodule, and no intense uptake in the liver. The liver is the most common site for small bowel NET metastasis. It is unusual to have pleural metastasis without liver metastasis. A review of this patient's past medical history revealed a motor vehicle accident 20 years ago which required an exploratory laparotomy and splenectomy. Gallium 68 Dotatate PET scan can detect splenosis because it has somatostatin receptors. In patients with an existing neoplasm, metastases should be ruled out before confirming the diagnosis of splenosis. CT-guided biopsy of pleura nodule was performed, which confirmed the diagnosis of splenosis.

Keywords: Neuroendocrine tumor; Thoracic splenosis; Gallium-68 Dotatate PET scan.

INTRODUCTION

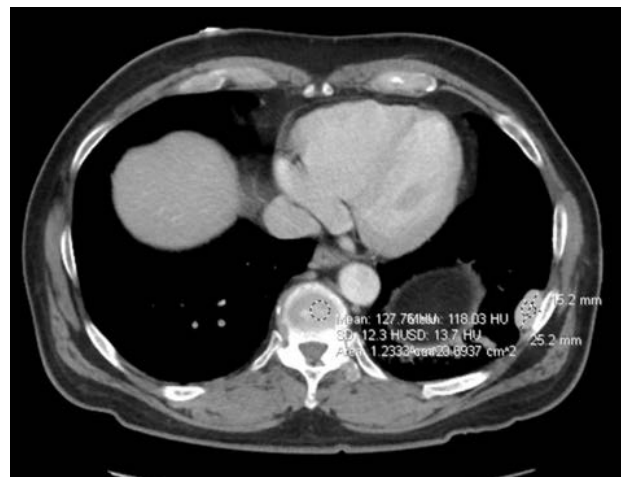
Thoracic splenosis is rare and consists of ectopic implantation of splenic tissue into the chest after concomitant thoracic and abdominal trauma with diaphragmatic injury. It occurs in about 18% of cases of splenic ruptures.¹ Intrathoracic splenosis is usually an asymptomatic, incidental finding.¹⁻³ Patients rarely present with symptoms of hemoptysis, cough, or pleuritic chest pain.³ Thoracic splenosis should be considered in the differential diagnosis of pleural nodules when there is a history of thoracoabdominal trauma and splenectomy.⁴⁻⁶ Metastases should be considered when patients already have neoplasia. If the possibility of malignancy cannot be ruled out safely with imaging, a biopsy must be pursued.¹ The diagnosis of this condition can be challenging as the multiple incidentally found nodules could mimic metastatic malignancy and obtaining a good history is key.⁷ Spleen can often have intense uptake by ⁶⁸Ga-Dotatate because splenic tissue can express somatostatin receptors (SSTR).⁸

CASE PRESENTATION

A 64-year-old man with a past medical history of hyperlipidemia, benign prostate hyperplasia, and renal calculi presented to the emergency department for evaluation of sudden onset of abdominal pain.

Computer Tomography (CT) scan was performed, which showed a 4 × 5 mm renal stone in the proximal left ureter and mild hydronephrosis, CT also showed a mesenteric root mass measuring 4.2 × 3.1 × 4.4 cm, as well as a left lung pleural-based lesion measuring 2.5 cm (Figure 1).

Figure 1: CT scan showing left lung pleural-based lesion measuring 2.5 cm.



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The patient had mild diarrhea and denied any flushing. He had a motor vehicle accident 20 years before the current presentation requiring an exploratory laparotomy and splenectomy. He smoked half a pack of cigarettes per day for 20 years which he had quit 20 years ago. He had no significant family history. His physical examination was unremarkable.

Based on CT findings, a metastatic neuroendocrine tumor (NET) was suspected. Baseline functional imaging using one of the somatostatin receptor-based imaging techniques is generally recommended in patients with advanced NETs. A Gallium 68 Dotatate PET scan was performed, which showed intense uptake in the partial calcified mass at the root of the mesentery as well as in the 2.5 cm pleural nodule. The scan was revealed another pleural-based lesion between the pericardium and descending thoracic aorta (Figure 2,3,4).

Figure 2: Gallium 68 Dotatate PET scan showing intense uptake in the partial calcified mass at the root of the mesentery.

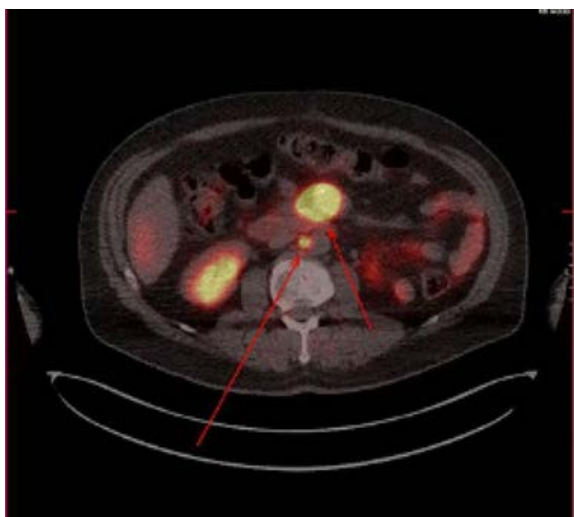


Figure 3: Dotatate PET scan showing intense uptake in the 2.5 cm pleural nodule.

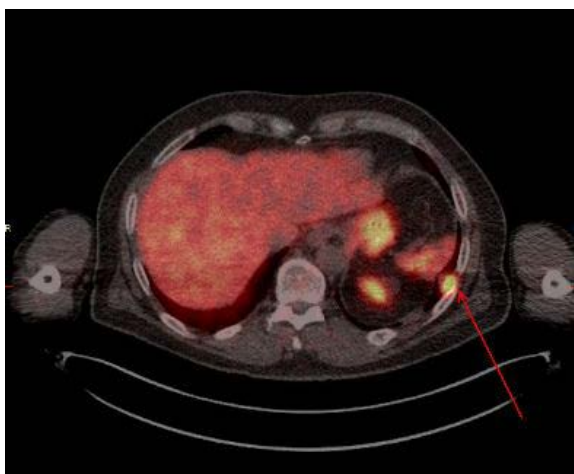


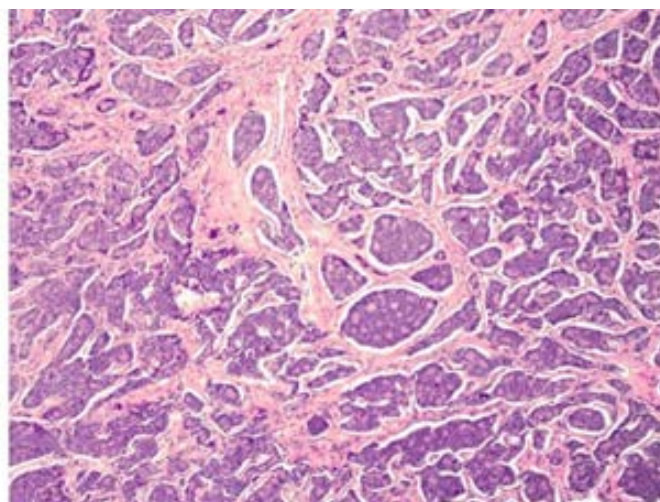
Figure 4: Gallium 68 Dotatate scan showing intense uptake in the pleural nodule.



Pertinent laboratory data included a 24-hour urine 5-HIAA level which was 19.7mg/day (normal is less than 10/24 hour). Serum Chromogranin A level was elevated at 343 ng/mL (reference range 0 -95 ng/mL) and elevated serotonin level of 1425 ng/mL (reference range 56 -244 ng/mL).

The patient had diagnostic laparoscopy, mesenteric lymphadenectomy, and small bowel resection with anastomosis along with cholecystectomy. Surgical pathology showed distal jejunal well-differentiated neuroendocrine tumor (2.0cm), margins were negative (Figure 5).

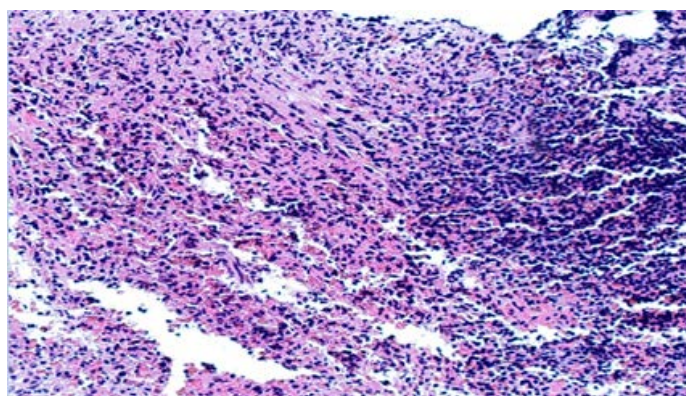
Figure 5: Small bowel tumor biopsy showing distal jejunal well-differentiated neuroendocrine tumor.



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A lymph node biopsy in the abdomen was also performed. One of eight mesenteric lymph nodes was positive for neoplasm and lymphovascular invasion was present. One month after small bowel resection CT-guided biopsy was performed. Biopsy of the pleural-based lesion was consistent with benign splenic tissue (Figure 6).

Figure 6: CT-guided biopsy of the pleural-based lesion showing benign splenic tissue.



Initiation of Sandostatin therapy was reviewed with the patient. He was monitored with close laboratory and imaging surveillance. Six months after diagnosis, 24 hr urine 5-HIAA level was 9 mg/day, which was within normal limits. Chromogranin A level decreased to 255 ng/mL (reference range 0- 95ng/mL) and serotonin level decreased to 1044 ng/mL (reference range 56 - 244 ng/mL). The Gallium 68 Dotatate scan was repeated, showing stable known mesenteric mass.

14 months after diagnosis, a repeat Gallium 68 Dotatate scan showed Interval progression of disease with increased radiotracer uptake and increased size of the partially calcified mesenteric mass, as well as a new focus of radiotracer uptake in right lateral hepatic lobe. Radiotracer uptake was again seen in the left lung base which corresponding to the known biopsy-proven splenic tissue. Repeated 24 hours urine 5-HIAA level was 10.9mg/day (normal is less than 10 mg/day). The patient was recommended to start Sandostatin therapy.

DISCUSSION

Small Bowel Neuroendocrine Tumors (SBNETs) are increasing in incidence and are now the most common primary malignancies of the small intestine.^{9,10} Despite this increase, the vague presentation and slow growth of these tumors lead to long delays in diagnosis, and many patients present with metastases.^{9,10} Localized SBNETs are mostly incidental diagnoses. Although SBNETs are typically small, they tend to induce a pronounced fibrotic reaction in the mesentery and often are accompanied by a mesenteric mass that represents enlarged regional lymph nodes.⁹ Our patient had a mesenteric mass on CT scan which was an incidental finding.

Most patients present with nonspecific abdominal pain rather than with symptoms of excess hormone secretion.¹⁰⁻¹³ It is often difficult to make the diagnosis of midgut NETs at an early stage because the pri-

mary tumors tend to be small and generally do not lead to symptoms. SBNET can present with symptoms of abdominal pain, bleeding or partial obstruction and NET with Liver metastasis usually presents with carcinoid syndrome.¹¹

Baseline functional imaging using one of the somatostatin receptor-based imaging techniques is generally recommended to identify NET primary tumors and their metastases.¹⁰ 68Ga-Dotatate PET was approved by the US Food and Drug Administration in June 2016 and is now widely available. It has several advantages compared with 111Indium-octreotide single-photon emission computed tomography (111In-octreotide SPECT) including better spatial resolution, faster post-injection image acquisition, less radiation exposure, and improved diagnostic accuracy.^{9,10}

Small bowel NET usually metastasizes to the liver.^{9,10,13} It is unusual to have pleural metastasis without liver metastasis. Benign lesions (physiological, tumor, and inflammatory lesions) can accumulate 68Ga-Dotatate because their tissues can express somatostatin receptors (SSTR). Both T-cell and B-cell lymphocytes including monocytes express SSTR. The immune system and its organs, including the spleen with its abundance of T and B cells, therefore can often have intense uptake by 68Ga-Dotatate.⁸

Thoracic splenosis should be considered in the differential diagnosis of left-sided pleural nodules in patients with a remote history of severe thoracoabdominal trauma and splenectomy.^{1,4-6,14} Our patient had exploratory laparotomy and splenectomy 20 years ago after a MVA. Therefore, thoracic splenosis was considered in the differential diagnosis.

Thoracic splenosis is a rare condition of splenic tissue auto transplantation into the chest following severe thoracoabdominal trauma and splenectomy. The time interval between trauma and diagnosis usually ranges from 6 to 46 years, with a mean of 21 years.¹⁵ Chest implantation is less frequent compared to abdominal involvement, and it occurs in approximately 18% of cases of splenic rupture.^{1,16} Generally, splenic tissue implants on serosal surfaces, and when it migrates into the chest, left-sided deposits are common because of the anatomical position of the spleen. Nodules are multiple in 75% of patients and isolated in approximately 25%. Pulmonary parenchyma is an uncommon site of implantation.¹

The true prevalence of these cases is likely underestimated, as most patients are asymptomatic, and the diagnosis is an incidental finding.^{1-3,15} There are few reports of symptoms of recurrent hemoptysis and pleuritic pain associated with this condition.^{1,3,15}

It is important to rule out other conditions that mimic these radiological findings, such as lymphoma, infectious diseases, hamartomas, pleural metastases (lung, breast, and melanoma), and rheumatoid nodules.^{1,15} Metastases should be considered when patients already have an existing neoplasm. If the possibility of malignancy cannot be ruled out safely with imaging, CT-guided biopsy or thoracotomy must be pursued.^{1,15} In our patient with a recent diagnosis of small bowel Neuroendocrine tumor, CT-guided biopsy of the pleural-based nodule was

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performed to rule out metastatic NET, which confirmed the diagnosis of thoracic splenosis. In almost all cases, the management is expectant. Surgery is indicated only in symptomatic patients and in those patients whose diagnosis is questionable, or the exclusion of malignancy is not possible.^{1,15}

CONCLUSION

Thoracic splenosis must be considered in the differential diagnosis of pleural nodules when there is a remote history of severe thoracoabdominal trauma and splenectomy. Splenosis may mimic metastatic lesions on 68 Gallium Dotatate PET scan. While evaluating pleural nodules, metastases should be considered when patients already have an existing neoplasm. Biopsy must be pursued if the possibility of malignancy cannot be ruled out safely with imaging.

CONFLICTS OF INTEREST

None.

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