A rare case of multiple primary extragastrointestinal stromal tumors in the abdominopelvic cavity detected by ^{18}F-FDG PET/CT

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Abstract
An extragastrointestinal stromal tumor (EGIST) refers to the gastrointestinal stromal tumor in any location outside the gastrointestinal tract originates from mesenchymal tumors. The majority of the EGIST appeared as isolated mass, and multiple primary EGIST is quite rare. Here, we report a 36-year-old man of multiple EGIST in the abdominopelvic cavity, which was misdiagnosed as lymphoma on fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) imaging. The patient received adjuvant drug treatment (imatinib, 400mg/day) for three months, and then further resection of all lesions was performed. No recurrence was found in CT follow-up one year after the operation.

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Introduction

Gastrointestinal stromal tumors (GIST) are most frequently found in the stomach (60%-70%), colorectum (10%), and esophagus (<5%) [1]. Although stromal tumors in the gastrointestinal tract commonly metastasize to the omentum and mesentery, they may also occur as primary tumors outside the gastrointestinal tract, especially in the omentum and the mesentery [2]. A number of studies of gastrointestinal stromal tumors have been published in the radiology literature [3-6]; however, few case reports are available on the imaging appearances of multiple extragastrointestinal stromal tumors (EGIST). Here, we report a case of multiple primary EGIST in the abdominal, which was misdiagnosed as lymphoma on fluorine-18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) imaging.

Case presentation

A 36-year-old man complained of abdominal pain for two months, no bloody stools, and altered bowel habits. An outside CT scan of the abdomen showed multiple space-occupying lesions in the abdominal cavity, which suggested malignancy. Physical examination revealed multiple hard masses in the abdominal. Laboratory findings were unremarkable, and tumor markers, including AFP, CEA, CA199, CA125, HE4, and SCC were all within the normal limit. No tumor dwelling inside of the colon and the rectal wall was detected by the enteroscopy examination. Fluorine-18-FDG PET/CT was performed for staging. Positron emission tomography/CT demonstrated multiple space-occupying lesions with abnormal ^{18}F-FDG uptake in the abdominopelvic cavity, and the tumor sizes ranged from 3 to 10cm (mean±standard deviation, 7±3cm). The maximum cross-section of the largest mass was approximately 10.9×8.5cm, and the margin was well defined. The abnormal increase of glucose metabolism was heterogeneous, and the maximum standardized uptake value (SUVmax) was about 9.1 (Figure 1). Fluorine-18-FDG PET/CT imaging suggested lymphoma in the abdominopelvic cavity.

A biopsy of the tumor was performed under laparoscopy. Hematoxylin and eosin staining of these sections of the tumors consisted of epithelial-like cells and spindle cells (Figure 2), which were positive for CD117, DOG-1, and CD34 immunostaining. The Ki67

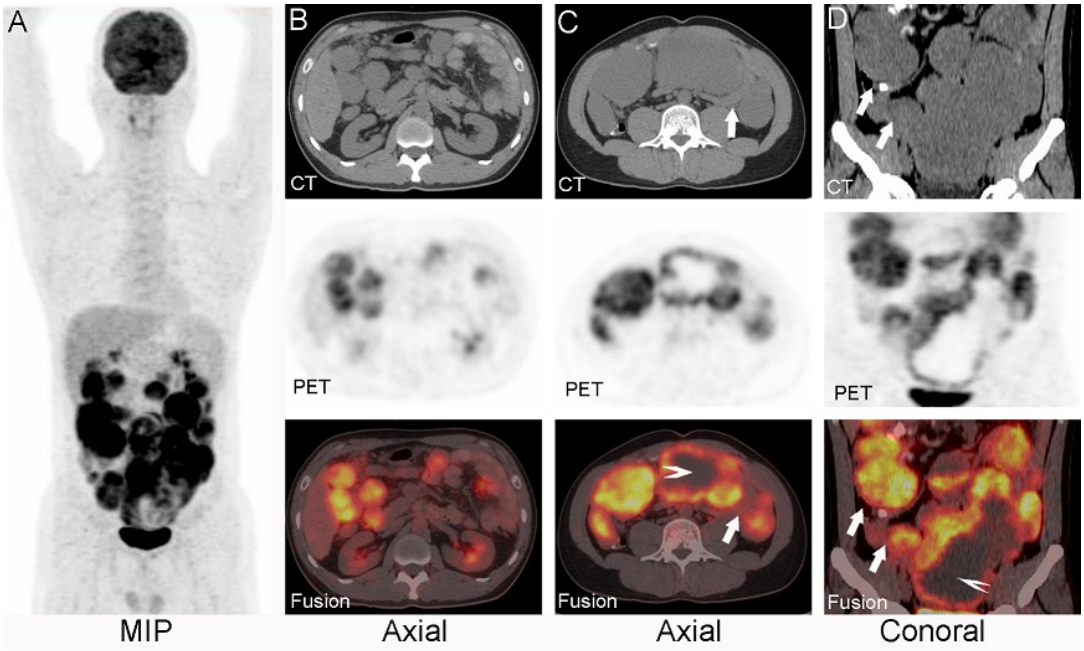

Figure 1. The maximum intensity projection (A) image demonstrated multiple lesions with abnormal ^{18}F-FDG uptake in the abdominopelvic cavity. The transverse images (B) of the abdomen showed multiple space-occupying lesions in the abdominopelvic cavity with an intense ^{18}F-FDG uptake. The transverse images (C) and The coronal images (D) revealed the enormous mass showed peripheral hypermetabolism and internal necrotic area (arrowheads), and the intestinal tract was obviously compressed (arrows).

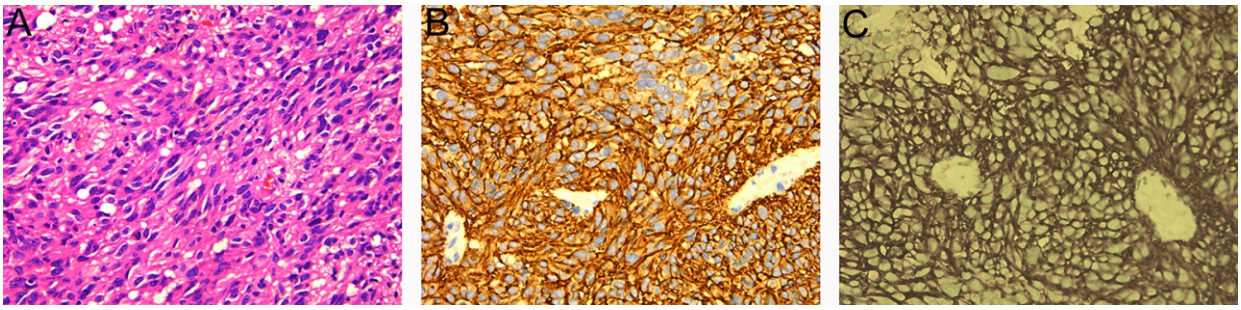

Figure 2. A biopsy of the tumor was performed. Hematoxylin and eosin staining of these sections of the tumors consisted of epithelial-like cells and spindle cells (A, ×400), which were positive for CD117 (B, ×400), DOG-1 (C, ×400).

was 50%. These findings were consistent with a diagnosis of multiple primary EGIST. However, owing to the location and volume of the various tumors, the risk of intestinal canal injury exists for immediate operation. The patient was discharged from the hospital with adjuvant drug treatment (imatinib, 400mg/day) for three months, and then further resection of all lesions was performed, and no recurrence was found in CT follow-up one year after the operation.

Discussion

A GIST is the most common mesenchymal tumor of the gastrointestinal tract, originating from the interstitial cell of Cajal [7]. An extragastrointestinal stromal tumor refers to GIST in any location outside the gastrointestinal tract originates from mesenchymal tumors [8-10]. Extragastrointestinal stromal tumor has a preference for the areas of the prostate [11], bone [12], vagina [13], mesentery [14], and retroperitoneum [15], etc. However, the majority of the EGIST appeared as isolated mass, and a case of multiple primary EGIST in the abdominal has not been reported.

Gastrointestinal stromal tumors occur at any age, with a median age at detection of 65 years, but they rarely occur (< 0.5%) in individuals younger than age 20 years [16]. Gastrointestinal stromal tumors occur with similar frequency in males and females. The median tumor size at diagnosis is approximately 6cm^2, but it may be >20cm^2. The most common clinical manifestation of symptomatic stromal tumors in the gastrointestinal tract is gastrointestinal bleeding caused by mucosal ulceration [17]. The patient may present with hematemesis, melena, hematochezia, or signs and symptoms of anemia caused by occult bleeding [2]. The patient in the case was only 36 years old, which is not the common age of stromal tumors, and he didn't have typical clinical symptoms such as gastrointestinal bleeding.

Numerous studies have reported the CT, and magnetic resonance imaging (MRI) features of primary GIST, including heterogeneous enhancement, exophytic growth, size of > 5cm, a necrotic or cystic center, mucosal ulceration, tumor vessels, and aneurysmal dilatation [18-20]. Anatomical and metabolic information in a single image can be obtained through a one-step PET/CT examination. Compared to traditional imaging techniques, PET/CT has a more obvious advantage. Fluorine-18-FDG PET/CT has been well established for staging, restaging, and monitoring treatment response of GIST [21-23]. However, to the best of our knowledge, few studies have reported the ^{18}F-FDG PET/CT findings of multiple primary EGIST. Due to the rarity of EGIST, much of the available radiological information is derived from small case series, which identify EGIST as large masses with solid and cystic components and intensely ^{18}F-FDG uptake [24]. However, the majority of the EGIST appeared as isolated mass, and multiple primary EGIST on ^{18}F-FDG PET/CT was not described in literature before. This will lead to a lack of understanding of the imaging diagnosis of this lesion. International guidelines have recommended the use of PET/CT for diagnosis and staging of lymphoma [25]. Most of the PET/CT findings of lymphoma were significantly increased glucose metabolism in enlarged lymph nodes or masses. It has also been reported that central necrosis also occurs when lymphoma tumors are relatively large [26, 27]. Therefore, it is hard to make a diagnosis between multiple EGIST and lymphoma when it happened to numerous masses in the abdominopelvic cavity with significant increased ^{18}F-FDG uptake.

Optimal management of EGIST requires carefully radiographic, exactly pathologic, systematic medical examination, and complete surgery. Surgery remains the standard treatment for primary resectable EGIST. Imatinib therapy before radical prostatectomy for patients with a diagnosis of EGIST may make surgical resection easier and reduce the risk of recurrence [24]. Multiple primary EGIST in the abdominal cavity is rather rare. It may be presented as multiple masses with significant ^{18}F-FDG uptake in the ^{18}F-FDG PET/CT scanning, and hope our case provides a differential diagnosis for clinicians in the following similar cases, such as lymphoma and metastatic malignancy, etc.

Ethics approval and consent to participate

Patients had provided informed consent for publication of the case. This study was performed in accordance with the Declaration of Helsinki.

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Bibliography

1. Miettinen MLasota J. Gastrointestinal stromal tumors-definition, clinical, histological, immunohistochemical, and molecular genetic features and differential diagnosis. *Virchows Arch* 2001; 438: 1-12.
2. von Mehren MJoensuu H. Gastrointestinal Stromal Tumors. J *Clin Oncol* 2018; 36: 136-43.
3. Sandrasegaran K, Rajesh A, Rydberg J et al. Gastrointestinal stromal tumors: clinical, radiologic, and pathologic features. *Am J Roentgenol* 2005; 184: 803-11.
4. Caramella T, Schmidt S, Chevallier P et al. MR features of gastrointestinal stromal tumors. *Clin Imaging* 2005; 29: 251-4.
5. Wei K, Pan B, Yang H et al. F-18 FDG PET, CT, and MRI for detecting the malignant potential in patients with gastrointestinal stromal tumors: A protocol for a network meta-analysis of diagnostic test accuracy. *Medicine (Baltimore)* 2018; 97: e0389.
6. Ulusan SKoc Z. Radiologic findings in malignant gastrointestinal stromal tumors. *Diagn Interv Radiol* 2009; 15: 121-6.
7. Joensuu H, Hohenberger P,Corless CL. Gastrointestinal stromal tumour. *Lancet* 2013; 382: 973-83.
8. Kosemehmetoglu K, Kaygusuz G, Fritchie K et al. Clinical and pathological characteristics of gastrointestinal stromal tumor (GIST) metastatic to bone. *Virchows Arch* 2017; 471: 77-90.
9. Xu L, Wen G, Ding Y et al. A lethal mesenteric gastrointestinal stromal tumor: a case report and review of the literature. *Int J Clin Exp Pathol* 2015; 8: 11715-21.
10. Zhu J, Yang Z, Tang G et al. Extragastrointestinal stromal tumors: Computed tomography and magnetic resonance imaging findings. *Oncol Lett* 2015; 9: 201-8.
11. Alabed YZ. Gastrointestinal Stromal Tumor of the Prostate: Staging and Evaluation of Response to Therapy With ^{18}F-FDG PET/CT. *Clin Nucl Med* 2018; 43: 52-4.
12. Zhang Y, Zhang A, Song L et al. Primary Extragastrointestinal Stromal Tumor on FDG PET/CT. *Clin Nucl Med* 2018; 43: 705-6.
13. Hanayneh W, Starr J, George TJ, Jr. et al. Extragastrointestinal stromal tumors of the pelvic cavity and the vagina: Two case reports and review of the literature. *Gynecol Oncol Rep* 2018; 25: 3-7.
14. Gupta N, Mittal S, Lal N et al. A rare case of primary mesenteric gastrointestinal stromal tumor with metastasis to the cervix uteri. *World J Surg Oncol* 2007; 5: 137.
15. Sawaki A. Rare gastrointestinal stromal tumors (GIST): omentum and retroperitoneum. *Transl Gastroenterol Hepatol* 2017; 2: 116.
16. Joensuu H, Vehtari A, Riihimaki J et al. Risk of recurrence of gastrointestinal stromal tumour after surgery: an analysis of pooled population-based cohorts. *Lancet Oncol* 2012; 13: 265-74.
17. Levy AD, Remotti HE, Thompson WM et al. Gastrointestinal stromal tumors: radiologic features with pathologic correlation. *Radiographics* 2003; 23: 283-304, 456; quiz 532.
18. Sandrasegaran K, Rajesh A, Rushing DA et al. Gastrointestinal stromal tumors: CT and MRI findings. *Eur Radiol* 2005; 15: 1407-14.
19. Kim HC, Lee JM, Choi SH et al. Imaging of gastrointestinal stromal tumors. *J Comput Assist Tomogr* 2004; 28: 596-604.
20. Horton KM, Juluru K, Montogomery E et al. Computed tomography imaging of gastrointestinal stromal tumors with pathology correlation. *J Comput Assist Tomogr* 2004; 28: 811-7.
21. Farag S, Geus-Oei LF, van der Graaf WT et al. Early Evaluation of Response Using ^{18}F-FDG PET Influences Management in Gastrointestinal Stromal Tumor Patients Treated with Neoadjuvant Imatinib. *J Nucl Med* 2018; 59: 194-6.
22. Van den Abbeele AD. The lessons of GIST--PET and PET/CT: a new paradigm for imaging. *Oncologist* 2008; 13 Suppl 2: 8-13.
23. Casali PG, Abecassis N, Aro HT et al. Gastrointestinal stromal tumours: ESMO-EURACAN Clinical Practice Guidelines for diagnosis, treatment and follow-up. *Ann Oncol* 2018; 29: iv267.
24. Liu YXu B. Primary Prostatic Extragastrointestinal Stromal Tumor on ^{18}F-FDG PET/CT. *Clin Nucl Med* 2016; 41: 224-6.
25. Barrington SF, Mikhaeel NG, Kostakoglu L et al. Role of imaging in the staging and response assessment of lymphoma: consensus of the International Conference on Malignant Lymphomas Imaging Working Group. *J Clin Oncol* 2014; 32: 3048-58.
26. Hopper KD, Diehl LF, Cole BA et al. The significance of necrotic mediastinal lymph nodes on CT in patients with newly diagnosed Hodgkin disease. *Am J Roentgenol* 1990; 155: 267-70.
27. Kahle XU, Montes de Jesus FM, Kwee TC et al. Relationship between semiquantitative ^{18}F-fluorodeoxyglucose positron emission tomography metrics and necrosis in classical Hodgkin lymphoma. *Sci Rep* 2019; 9: 11073.