18F-FDG PET/CT and MRI Imaging Features of Monomorphic Epitheliotropic Intestinal T-cell Lymphoma Involving the Liver and Lymph Nodes: A Rare Case Report

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Case report

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Abstract

Background: Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) is an uncommon, rapidly progressive, primary intestinal T-cell lymphoma. The most common site of occurrence is the small intestine. The prognosis of MEITL is extremely poor due to late diagnosis and lack of targeted therapy.

Case presentation: A case of MEITL involving the entire small bowel, part of colon, rectum, mesenteric lymph nodes and liver is herein reported. We are presenting the ^{18}F-FDG PET/CT features of MEITL, which showed all involved lesions with increased FDG activity. The MRI and pathological characteristics of MEITL were also described. Furthermore, some malignant diseases should be considered in the differential diagnosis.

Conclusions: Based on the lesions with high accumulation of FDG, our case shows the involved extent of MEITL, which is helpful for biopsy and treatment option decisions. We expect more and more physicians could know this disease and make an early diagnosis to improve the outcomes of MEITL.

Background

MEITL is an aggressive intestinal tumor of intraepithelial T-lymphocytes, accounting for <1% of all lymphomas[1]. It was formerly defined as type II enteropathy associated T-cell lymphoma. Since its distinctive characteristics and lack of association with celiac disease, MEITL is different from type I enteropathy associated T-cell lymphoma (EATL)[2]. It is a rare malignant lymphoma in western and the most common form of primary intestinal T-cell lymphoma in Asia[1]. Herein, we are presenting a case of a 48-year-old Chinses woman who initially suffered from abdominal pain and distension, diarrhea and urological symptoms and was found to have intestine wall thickening on the PET/CT and MRI which turned out to be MEITL.

Case Presentation

A 48-year-old woman was admitted to our hospital due to abdominal pain and distension for one month, diarrhea for three weeks, and urological symptoms including dysuria, frequency and urgency for one week. Watery stools were 3-5 times a day, without blood. She complained of associated fever with a maximum temperature of 39°C. She also reported with obvious weight loss of a month duration.

Laboratory findings on admission revealed the following: hemoglobin 97g/L (normal range, 115~150 g/L); white blood cells(WBC) 7.47×10^9/L (normal range, $3.5\sim9.5\times10^9$/L) with 83.5% neutrophils (normal range, 40%~75%) and lymphocytes 9.6% (normal range, 20%~50%); C-reactive protein 118mg/L (normal range, <8.20mg/L). Routine stool test showed yellow, mushy, occult blood ±. Urinalysis showed red blood cells of 199/hpf and WBC of 10/hpf (normal range, 0~2/hpf). Serum neuron specific enolase was 27.20ng/ml (normal range, <16.3ng/ml) and carbohydrate antigen 125 was 52.80U/ml (normal range, <35ng/ml). Patient tested negative for HIV.

Initially, the patient underwent an abdominal MRI (Fig 1, 2). It demonstrated that varied thickening wall of entire small bowel, part of colon, and rectum with iso or hypo-intensity on T_2WI, reduced diffusion on DWI, whereas low signal intensity on ADC map, and markedly homogeneous enhancement. In order to make a comprehensive assessment, a whole-body PET/CT (Fig 1, 2) was performed. The maximum intensity projection (MIP) showed the intestine in the left side of abdomen and pelvic cavity with diffuse increased FDG-activity, the spleen (SUVmax 4.8~5.3) and skeleton (SUVmax 7.6~8.8) were also with diffuse FDG-avid. The coronal CT and fused PET/CT revealed diffuse wall thickening of entire small bowel, part of colon and rectum with increased FDG-uptake (SUVmax 4.0~10.2), obviously in the proximal jejunum that showed a thick-walled prominent dilated loop with an air-fluid level. There were some swollen mesenteric lymph nodes with increased FDG-uptake (SUVmax 3.3~5.7). Multiple low-density lesions were FDG-avid (SUVmax 4.4~8.8) in the liver.

Given the high suspicion for malignancy, the patient underwent gastrointestinal endoscopes and histopathology. The results showed extensive ulcerations in the gut (Fig 3), and the result of biopsy pathology were suggestive of MEITL (Fig 3). The lymphoma cells are small to medium-sized, and monomorphic, expressing characteristically CD3, CD8, CD56 and so on, Ki-67 value of this patient was around 40%. Bone marrow biopsy demonstrated no evidence of lymphoma involvement.

MEITL is a primary intestinal T-cell lymphoma caused because of malignant proliferation of intraepithelial lymphocytes[2]. The most common site is small bowel, followed by large bowel, stomach, with frequent involvement of mesenteric and para-aortic or iliac lymph nodes. It can spread to lung (lesions are often accompanied with necrosis), liver, prostatic and central nervous system[2-5]. The SUVmax of all lesions in our case varied from 3.3 to 10.2, which was similar to Chan's report[1]. On PET/CT, MEITL needs to be differentiated from EATL and some aggressive B-cell lymphomas. EATL occurs as hypermetabolic lesions with high SUVmax, against a background of diffuse FDG accumulation of mild SUVmax evolving from refractory coeliac disease[5]. The SUVmax in B-cell lymphoma were generally obviously higher than those of MEITL[6]. Unfortunately, the prognosis of MEITL is extremely poor with a reported median survival of only seven months and one-year overall survival is only 36%[7], which is mostly due to late diagnosis and lack of targeted therapy. This patient died 3 months after she was diagnosed with MEITL.

Conclusions

Since the MEITL is a rare and aggressive disease with poor prognosis, and most radiologists are lack of knowledge about its imaging features, thus, we should pay particular attention to its radiologic features, consider the possibility of malignant lymphoma including MEITL and make an effort to diagnose this as soon as possible. Based on the lesions with high accumulation of FDG, the PET/CT can help to show the involved extent of MEITL, which is helpful for biopsy and treatment option decisions.

Abbreviations

MEITL: Monomorphic epitheliotropic intestinal T-cell lymphoma

EATL: type I enteropathy associated T-cell lymphoma

18F-FDG PET/CT: 18Fluorine-fluorodeoxyglucose Positron Emission Tomography/Computerized Tomography

SUVmax: the maximum of Standard Uptake Value

MIP: Maximum Intensity Projection

MRI: Magnetic Resonance Imaging

T_2WI: T_2 Weighed Imaging; DWI: Diffusion Weighted Imaging; ADC: Apparent Diffusion Coefficient

WBC: White Blood Cells

Declarations

Ethics approval and written informed consent

Written informed consent was obtained from this participant for publication of this case report and accompanying images.

Availability of data and materials

Not applicable.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

XY Z and SM L contributed to the conception and design of the work, and have drafted the work. XY Z was a major contributor in writing the article. QY and YL performed the PET/CT examination and the

collection of the data. SM L has substantively revised the manuscript. ZW L also has revised it and approved the submitted version. All authors read and approved the final manuscript.

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Figures

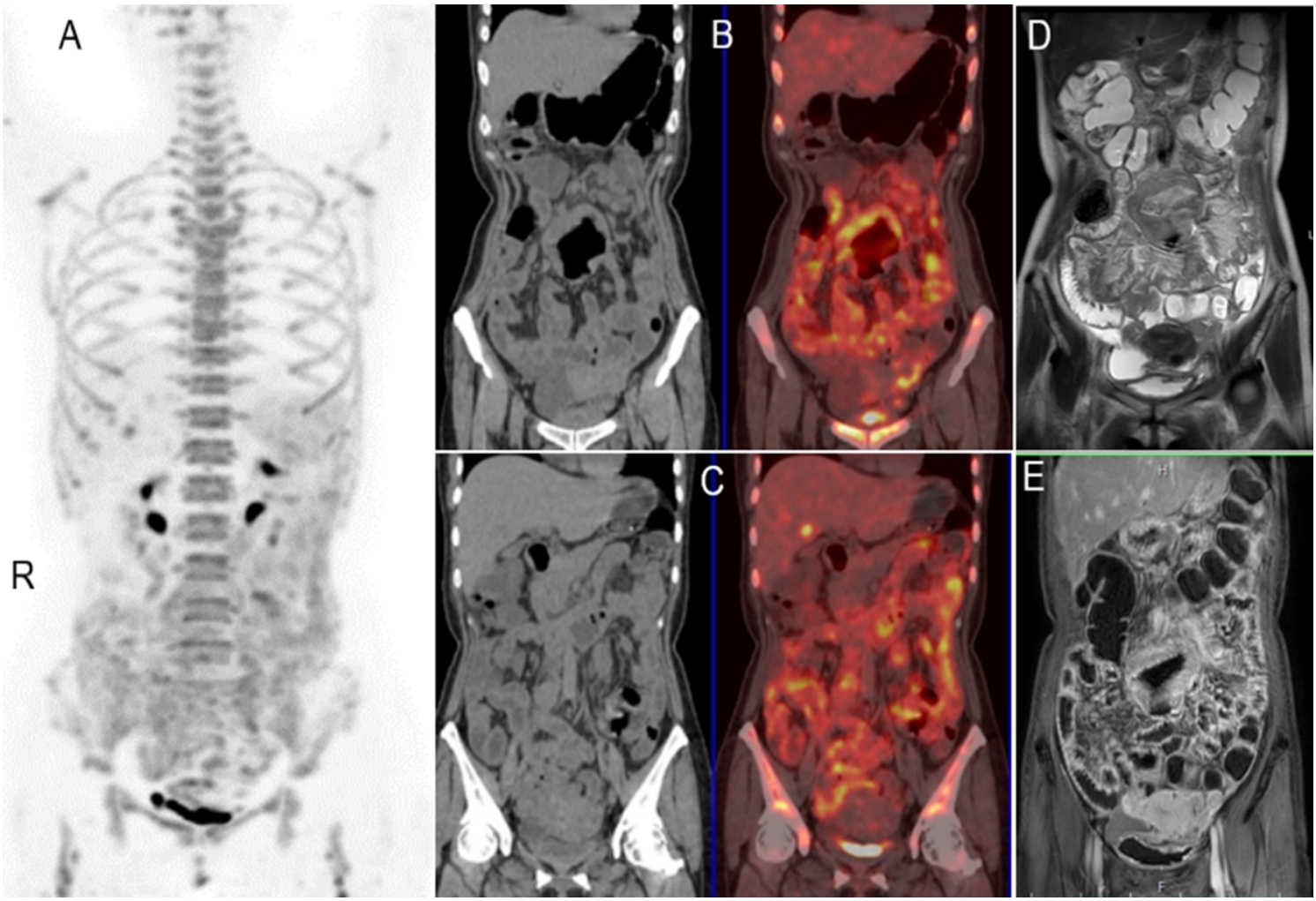

Figure 1

The MIP of PET/CT and the coronary images of PET/CT and MRI. The PET/CT (A, B, C) shows the intestine in the left side of abdomen and pelvic cavity with diffuse increased FDG-activity, the spleen and skeleton were also with diffuse FDG-avid. The coronal MRI demonstrated the thick-walled intestine with iso-low signal on T2WI (D), and obvious uniform enhancement (E).

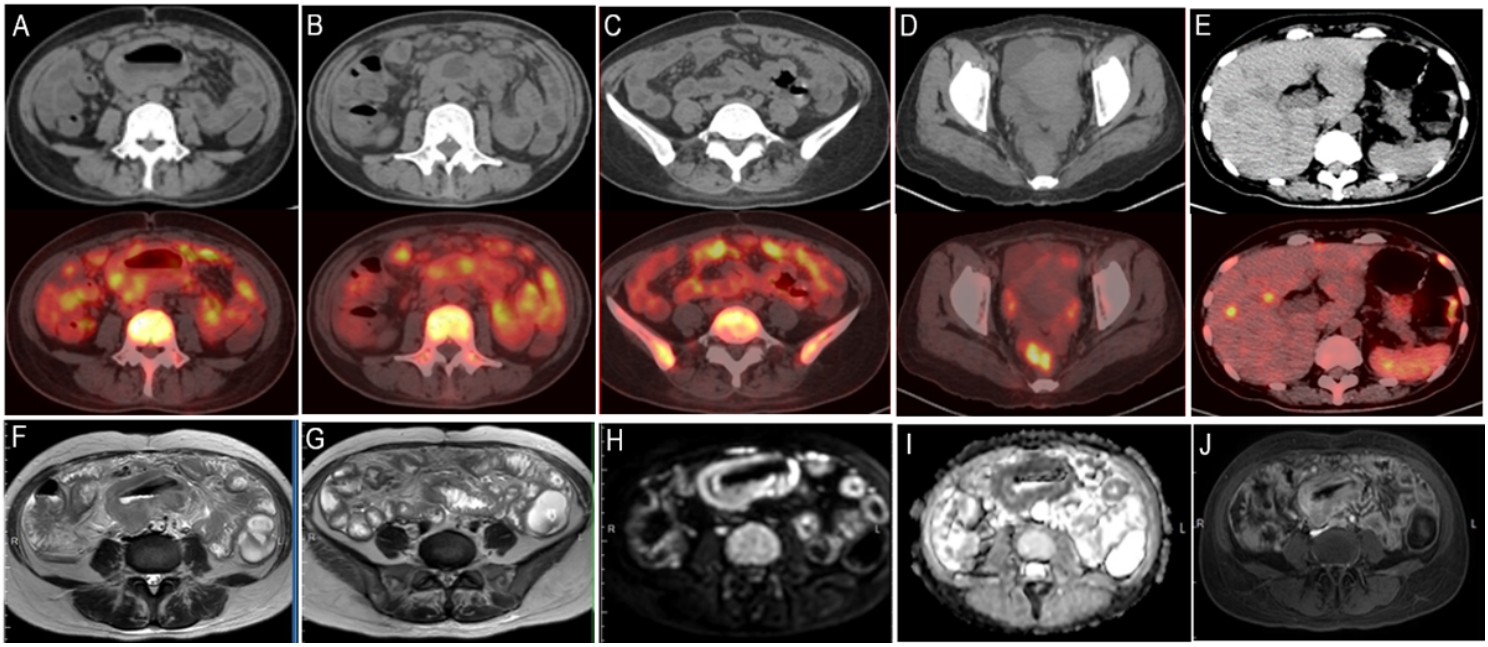

Figure 2

The trans-axial PET/CT and MRI images. The PET/CT (A~E) revealed diffuse, varied thickening wall of entire small bowel, part of colon, and rectum with increased FDG-activity, obviously in the proximal jejunum that showed a thick-walled prominent dilated loop with an air-fluid level. There were some swollen mesenteric lymph nodes with increased FDG-uptake. Multiple low-density lesions were FDG-avid in the liver. Abdomen MRI (F~J) demonstrated that the thickened intestinal wall with iso or hypo-intensity on T2WI, reduced diffusion on DWI, whereas low signal intensity on ADC map, and markedly homogeneous enhancement.

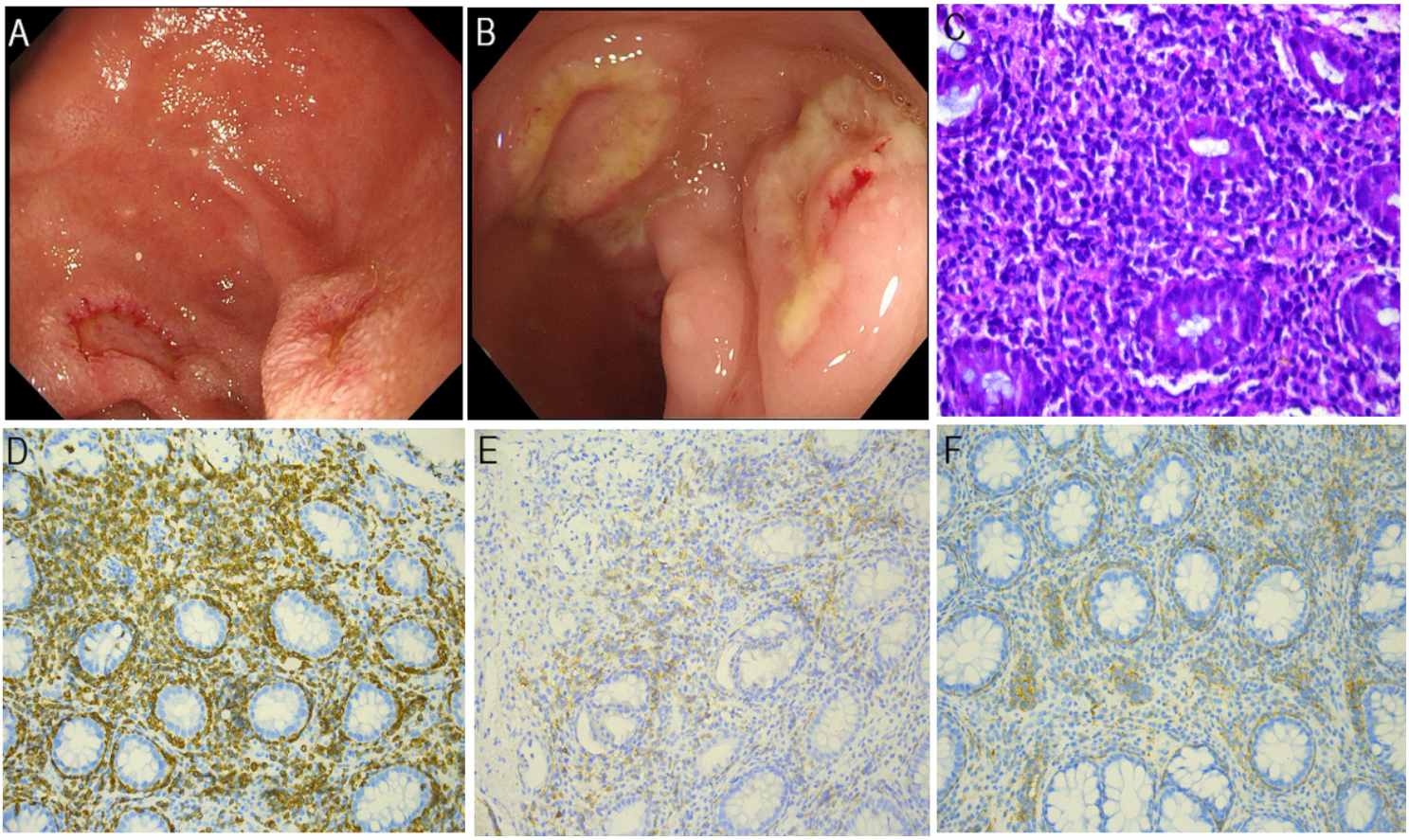

Figure 3

The results of gastrointestinal endoscopes and histopathology. Extensive ulcerations were showed in the gut (A, B). The lymphoma cells of MEITL (C) are small to medium-sized, and monomorphic (hematoxylin-eosin, original magnification×400), expressing characteristically CD3(D), CD8(E), CD56(F).