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CASE REPORT

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A case report of monomorphic epitheliotropic intestinal T-cell lymphoma with pulmonary manifestations as the first presentation

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Abstract

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) is defined as a type of lymphoma that occurs in the intestine, but may show extra-intestinal involvement, such as in the skin, brain, lungs, stomach, ovaries, and uterus, which have been previously reported. The disease has no specific clinical manifestations and is often diagnosed by pathological examination as a complication of intestinal perforation or obstruction. The difficulty of making a timely diagnosis is further compounded when the disease begins with non-gastrointestinal symptoms. In this paper, We report a case of MEITL with concurrent abdominal and pulmonary involvement on imaging, but only present with respiratory symptoms. The patient was diagnosed as Peripheral T cell lymphoma, Not otherwise specified (PTCL, *NOS*) initially based on lung biopsy. However, the diagnosis of MEITL was finally established due to complications of intestinal obstruction and perforation during treatment. Therefore, for lymphomas that occur at multisite outside lymph nodes, multiple-site biopsy should be performed to enhance the accuracy of the pathological diagnosis.

Keywords Monomorphic epitheliotropic intestinal T-cell lymphoma, Respiratory symptoms, Lung lesions, Intestinal lesions

Introduction

Monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) is a rare and highly aggressive intestinal-associated T-cell lymphoma, previously known as type II Enteropathy-associated T-cell lymphoma [1](type II EATL), which was classified by the 2016 revision of the World Health Organization (WHO) classification of lymphoid neoplasms as a separate clinical-pathological entity distinct from EATL [2], is not associated with celiac disease and is more common in Asia. Though the small intestine is the mainly infiltrated site and symptoms first appear, the concurrent or secondary involvement of the appendix, skin, brain, lung, stomach, ovaries, and uterus has been reported [3–8]. The prognosis for this disease is usually very poor, with a median survival time of only 7 months [9].

In this paper, we reported a case of MEITL, which presented as simultaneous lesions in the intestines and lungs on imaging, while clinically manifesting only respiratory symptoms.

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Case presentation

A 60-year-old male presented to the Department of Thoracic Surgery with a 1-month history of intermittent dry cough that was aggravated by cough and sputum production for 1 week. The patient has no systemic symptoms such as fever, night sweats, weight loss, or gastrointestinal symptoms such as abdominal pain, celiac disease, constipation and vomiting. The chest computed tomography (CT) scan revealed a space-occupying lesion in the basal segment of the lower lobe of the right lung, and an abdominal CT showed significant thickening of the small intestinal wall.(Figure 1A and B). The patient underwent a percutaneous biopsy of the pulmonary tumor. Microscopy examination identified a predominant populationr of medium-sized lymphoid cells exhibiting significant heterogeneity, with marked nuclear extrusion, deformation, and atypia, with no obvious characteristics of epithelial invasion.(Fig. 2A) Immunohistochemical analysis demonstrated the following profile: CD2 (-), CD7 (+), CD5(-), CD3 (+), CD56(+), TIA-1 (+), EBER(-) (Table 1). Based on these morphological and immunophenotypic findings, the patient was independently confirmed by two institutions as Peripheral T-cell lymphoma, Not otherwise specified (PTCL, *NOS*). The bone marrow staging by bone biopsy, flow cytometry, and cytogenetic analysis found no evidence of involvement. The patient was ultimately diagnosed with PTCL, *NOS*, classified as stage IV, group A, with a Prognostic Index for T-cell Lymphoma (PIT) score of 1, indicating low-to-intermediate risk.

The treatment with the CHOPE regime (cyclophosphamide, doxorubicin, vincristine, prednisone, and etoposide) was initiated. After two cycles of treatment, the patient's pulmonary lesions began to shrink, but it was complicated by acute intestinal obstruction, and surgical resection of the obstruction site was performed. The postoperative examination revealed monomorphic and medium-sized lymphoid cells with diffuse infiltration and no obvious characteristics of epithelial invasion.(Fig. 2C). Immunohistochemistry analysis demonstrated positive for CD7, CD3, CD8, CD56 and TIA-1, withfocal GranzymeB expression, while negative for CD5, CD2, CD4 and EBER(Table 1). These findings supported the diagnosis of monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL) confirmed by pathologists from both institutions. Considering that the treatment of the patient's lung lesions was effective, the patient continued to complete the CHOPE regimen for another four cycles and was evaluated as complete metabolic remission (CMR) by Positron Emission Tomography-Computed Tomography (PET-CT) 1 month after the end of treatment. Three months later, the patient was followed up with chest and abdominal CT scans showed local thickening and enhancement of the small intestine in the right pelvis without change of the lung (Fig. 1C and D). Subsequently, the intestinal biopsy confirmed the relapse of MEITL. Consequently, the patient was reassessed and administered the MIT-LSP + GDP regimen, which includes gemcitabine, cisplatin, dexamethasone, and mitoxantrone liposome. However, the patient underwent surgery for the third time due to gastrointestinal perforation following the treatment. The postoperative bowel pathology results were consistent with a diagnosis of MEITL. The patient was subsequently treated for 3 cycles with a single dose of mitoxantrone liposome every 4 weeks then stopped treatment. According to imaging assessment at the endpoint, the patient was still in CR status (lung lesions were still CR and intestinal lesions were postoperative changes) (Fig. 1E and F), but the patient gradually developed a loss of appetite and subsequent malnutrition and died due to multiple organ failure caused by cachexia. From diagnosis, the patient survived for 20 months.

Discussion

Although two distinct lesions in the lung and intestine were visible on the pre-treatment CT scans (Fig. 1A and B), the patient presented initially just respiratory symptoms. Based on the biopsy of the tumor in the lung, the pathological diagnosis was initially identified as PTCL, *NOS*. However, during the course of treatment, the patient successively developed complications of intestinal obstruction and intestinal perforation, and had to undergo surgery. Based on the postoperative specimen the patient was instead diagnosed with MEITL.

In addressing the discrepancies between pulmonary and intestinal specimen diagnoses, pathology experts compared the microscopic morphology of lung puncture samples and multiple intestinal biopsy specimens. The lung specimens obtained via fine-needle aspiration showed significant cellular deformation(Fig. 2A). However, detailed analysis revealed consistent tumor cell morphology in both samples: The lesion is predominantly composed of small to medium-sized, morphologically uniform, diffusely arranged atypical cells, lacking an inflammatory background. However, we did not observe distinct features of tumor cell aggregation towards the epithelium, which might be related to our specimen preparation.(Fig. 2). Furthermore, Despite the absence of CD4 and CD8 detection in the lung samples, the immunohistochemical analysis demonstrated an identical expression pattern in both the lung and intestinal tissues.(Table 1) Clinically, despite radiological evidence of intestinal wall thickening, the patient exhibited no gastrointestinal symptoms, necessitating differentiation from indolent T-cell lymphoproliferative disorder of the gastrointestinal tract(ITLPD-GI). ITLPD-GI is clinically indolent with minimal progression over time. Histopathological examination of the gastrointestinal mucosa revealed

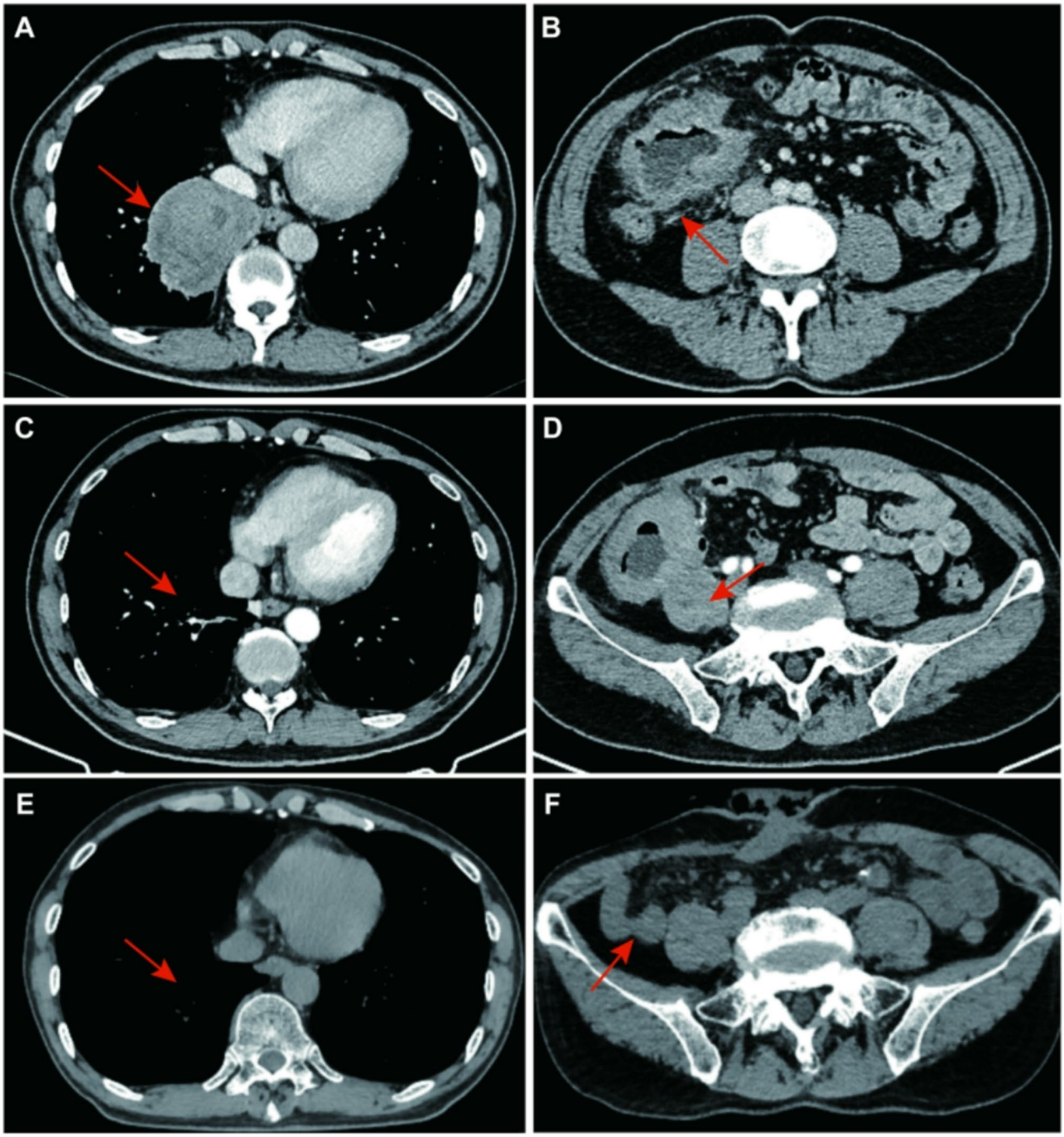

Fig. 1 Imaging evaluation of the patient. **A**: Chest computed tomography (CT) scan before treatment. A mass measuring approximately 79 mm x 81 mm x 69 mm is seen in the basal segment of the lower lobe of the right lung as indicated by the arrow. It was slightly ill-defined with irregular lobulated margins and localized protrusion into the posterior mediastinum. **B**: Abdominal CT scan before treatment. There is marked thickening of the bowel wall and dilatation of the bowel lumen at the point indicated by the arrow. The surrounding fat interstitium is blurred with multiple lymph node shadows. **C**: Chest CT scan after 6 treatment cycles. The large mass in the basal segment of the lower lobe of the right lung, indicated by the arrow, has subsided and only a few irregular debris shadows are visible. **D**: Abdominal CT scan after 6 treatment cycles. The arrowed area shows localized thickening and abnormal enhancement of the small intestine in the right pelvis, suggesting recurrence of the intestinal tumor. **E**: CT scan of the chest at the end of treatment. The large tumor in the basal segment of the lower lobe of the right lung at the point indicated by the arrow has completely disappeared to a state of complete remission. **F**: CT scan of the abdominal at the end of treatment. The area indicated by the arrow is a postoperative change after small bowel perforation fistula. It shows a slight thickening of the intestinal wall at the anastomosis site, with no obvious features of tumor involvement

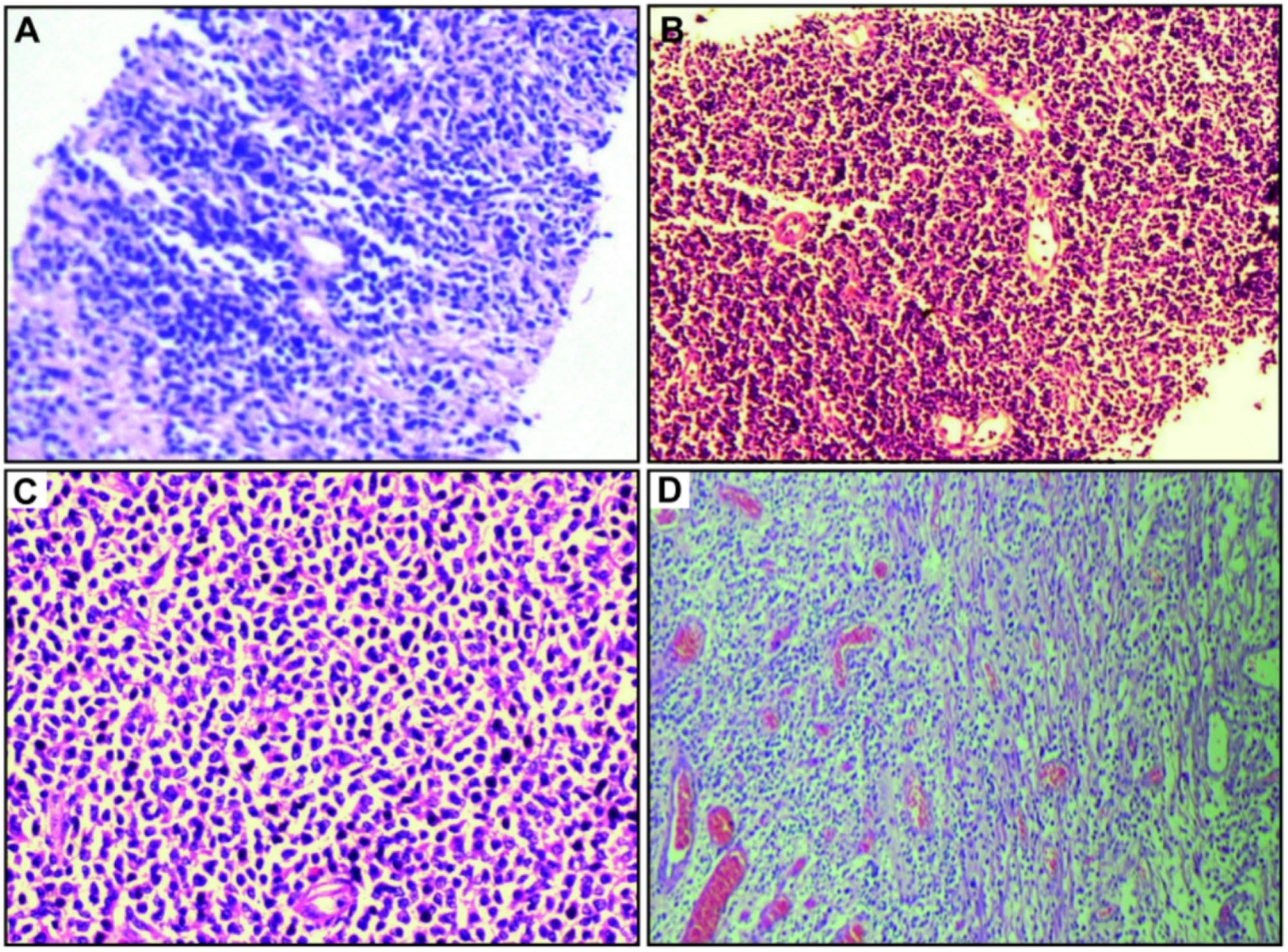

Fig. 2 Histopathology from different biopsy sites. **a** Lung. **b** Bowel mass. **c** Intestinal obstruction. **d** Intestinal perforation Upon microscopic examination, the specimen reveals small to medium-sized lymphoid cells that exhibit atypical features. These cells are distributed diffusely and display an infiltrative growth pattern, lacking an inflammatory background. **a**-**d**, H&E, (hematoxylin, and eosin, original magnification×200) Notes: **a**. The pulmonary specimens were obtained by fine-needle biopsy, some cells are moderately compressed and deformed. **d**. In the intestinal perforation specimens, hemorrhagic necrosis of the plasma membrane surface is seen

small lymphocyte infiltration with Immunohistochemical findings of CD3, CD5, CD7 and CD8 were typically positive, CD4 negative (occasionally positive), and CD56 and EBER negative, alongside a low Ki-67 proliferation index (less than 10%). These features facilitated the differentiation from ITLPD-GI. Based on these findings, after discussions between pathologists and clinicians, despite the more pronounced initial pulmonary symptoms and signs, considering the characteristics of MEITL originating from the intestine, the patient was diagnosed with MEITL with secondary pulmonary involvement.

MEITL is a rare form of T-cell lymphoma that originates in the intestines. In rare cases, it may spread to organs beyond the intestine. The uterus, ovaries, brain, epidermis, stomach, lungs, and adrenal glands have all been implicated in the literature [4–8, 10]. Gastrointestinal symptoms, such as diarrhea, melena, abdominal pain, or distention, remain the predominant initial symptoms, which may present independently or in association with intestinal obstruction or perforation. To date, there has been only one reported case of MEITL that initially presented with cutaneous symptoms and was ultimately diagnosed following the onset of intestinal perforation during subsequent chemotherapy [4]. Previous studies have reported MEITL secondary to pulmonary involvement, albeit with gastrointestinal symptoms as the initial presentation [6]. In contrast, our case demonstrates a unique presentation where the patient exhibited no gastrointestinal symptoms despite radiographic evidence of intestinal wall thickening at disease onset.

In the absence of gastrointestinal specimen, pathologists initially misdiagnosed the patient with PTCL, *NOS*. The patient subsequently, developed intestinal obstruction, recurrence, and perforation. A review of the intestinal specimens led to a diagnosis of MEITL. Discussions with pathologists regarding the diagnostic challenges

Table 1 Immunohistopathology from different biopsy sites

Histopathology	pulmonary	intestinal obstruction	bowel mass	intestinal perforation
CD2	-	-	-	-
CD3	+	+	++	+
CD4	N	-	N	-
CD5	-	-	-	-
CD7	+	+	++	+
CD8	N	+	N	+
CD10	N	+	N	N
CD20	-	-	-	-
CD21	N	N	N	-
CD23	N	-	N	N
CD30	-	N	-	N
CD43	+	N	++	N
CD56	+	+	++	+
P53	N	-	-	N
TIA-1	+	+	+	+
GranzymeB	N	+(few)	-	+(few)
KI-67	70%	50%	50%	10%
EBER	-	-	-	-

-, negative; +, positive; N, not done

included: (1) MEITL, a tumor originating from the intestines, is difficult to diagnose without direct symptoms or signs of gastrointestinal involvement, particularly when relying solely on symptoms, signs from secondary organs, or biopsy results. (2) PTCL-*NOS* is frequently characterized by abundant inflammatory cell infiltration and notable focal necrosis, while MEITL usually presents with an absence of inflammatory background. However, the lung tumor sampling via fine needle aspiration caused significant tumor cell compression and deformation, leading to the misinterpretation of some tumor cells as a large number of inflammatory cells by the pathologist. Furthermore, the sampling technique resulted in the microscopic observation of extensive necrosis, which further complicated the diagnostic process. Therefore, it is crucial for clinicians to provide as detailed patient information as possible to pathologists. Additionally, for such rare diseases, biopsies from multiple sites may be necessary when lymphoma affects multiple systems, and efforts should be made to excise the tumor as completely as possible.

MEITL is highly aggressive with a median survival of just 7 months [9]. The low incidence results in insufficient attention. Additionally, the disease typically presents with nonspecific symptoms, complicating early diagnosis. Notably, nearly 50% of patients are diagnosed following intestinal obstruction or perforation [11]. However, pertinent research has shown that patient outcomes can be significantly improved through early diagnosis. For instance, a patient was diagnosed at an early stage using video capsule endoscopy and double-balloon enteroscopy and survived for 21 months [12]. Another patient with MEITL, who presented with melena and was diagnosed at an early stage following repeated biopsies by colonoscopy, has a continuous CR for 5 years after 8 cycles of CHOP (cyclophosphamide, doxorubicin, vincristine, and prednisone) regimen [11]. The patient in our report was able to receive early treatment and ultimately survived for 20 months. Therefore, early diagnosis and therapy can improve prognosis. When patients have gastrointestinal symptoms that have failed conventional treatment and there is evidence of abdominal imaging abnormalities, they should be encouraged to undergo gastrointestinal endoscopy and timely biopsy to improve early diagnosis of the disease.

Conclusion

In conclusion, MEITL is a lymphoma that arises in the intestine. However, it may rarely infiltrate organs other than the intestine. Adding to the confusion, the disease may present with a primary clinical presentation that is secondary to symptoms associated with the affected organs, rather than being devoid of symptoms of intestinal distress. Consequently, for lymphoma with intestinal involvement, multiple-site biopsies should be performed whenever possible to enhance the accuracy of pathological diagnosis.

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Author contributions
Bai Dong collected patient data, and wrote and edited the manuscript; Yonglin Chen collected patient data and histopathology analysis; Binbin Ding collected patient data, Nana Li collected patient data, Zijian Li revised and reviewed the manuscript.

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Data availability
No datasets were generated or analysed during the current study.

Declarations

Ethics approval and consent to participate
This paper was approved by the ethics committee of the First Hospital of Lanzhou University (ethics approval number: LDYYLL-2024-278), and informed consent from the patient's family was obtained.

Competing interests
The authors declare no competing interests.

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