



# A Rare Case of Intestinal T-cell Lymphoma with Multiple Complications

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Intestinal T-cell lymphoma is a rare and aggressive form of non-Hodgkin lymphoma. Owing to its rarity and variable manifestations, intestinal T-cell lymphoma is usually diagnosed at an advanced stage. Therefore, it may be accompanied with serious complications at the time of diagnosis. Herein, I reports a case of intestinal T-cell lymphoma with multiple severe complications.

**Key Words:** Lymphoma, T-cell; Intestine, small; Complications

## INTRODUCTION

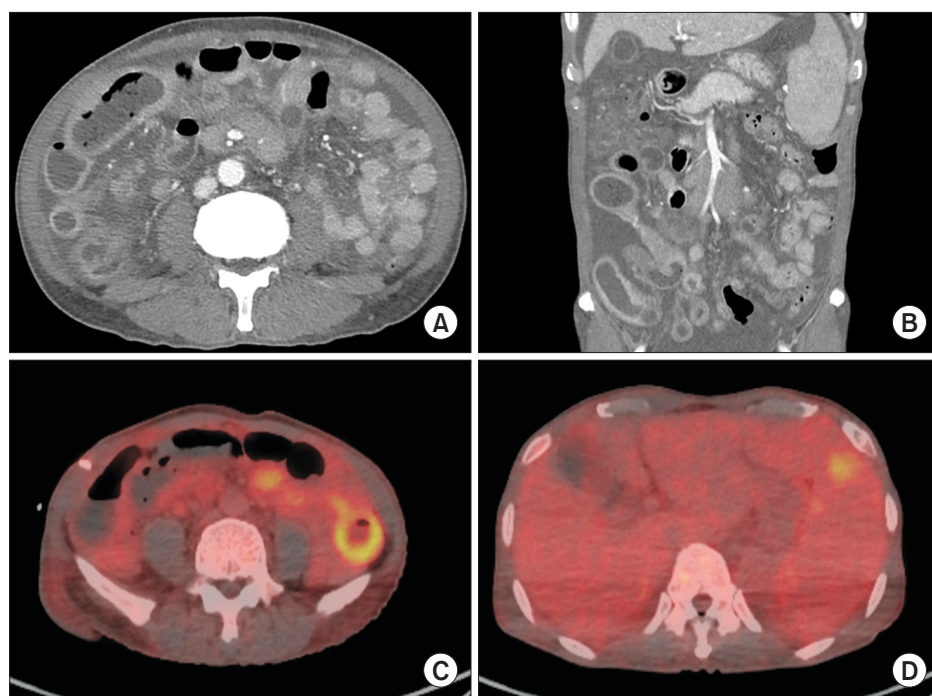
The gastrointestinal (GI) tract is a common site for extranodal lymphoma involvement, accounting for 5–10% of all GI cancers [1]. However, the majority of primary GI lymphomas are of the B-cell lineage, with T-cell lymphomas accounting for only 4–9% [2-4]. Intestinal T-cell lymphoma tends to have various aggressive clinical manifestations [5,6], leading to severe complications. Here, I present a rare case of intestinal T-cell lymphoma with multiple complications.

## CASE

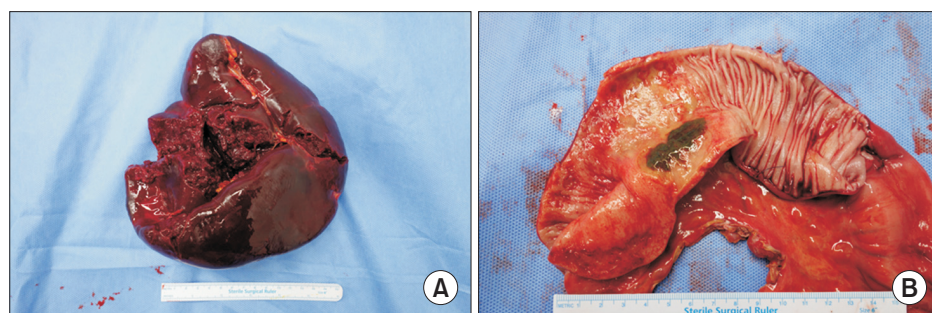
A 70-year-old male presented with diffuse abdominal pain and weight loss of 4 kg over 3 months. The patient had no relevant medical history except for hypertension and dyslipidemia, and no history of surgery or family history. His vital signs were stable, and on physical examination, his abdomen was found to be soft but moderately distended without definite tenderness. Esophagogastroduodenoscopy and colonoscopy performed at another hospital one month prior showed no significant abnormalities. The patient's laboratory data were as follows: white blood cell count, 18,860/mm<sup>3</sup>; hemoglobin, 15.9 g/dl; platelet count, 212,000/mm<sup>3</sup>. Abdominal

computed tomography (CT) revealed multifocal segmentally enhanced wall thickening of the small bowel and homogeneous enhancement of conglomerated lymph nodes (LNs) in the mesenteric root. An omental cake and a large amount of ascites were also identified (Fig. 1). Positron emission tomography showed diffuse/segmental wall thickening with moderate hypermetabolism in the ileum and several low-attenuated lesions with hypermetabolism in the enlarged spleen. Multiple hypermetabolic LNs were identified in the mesentery and retroperitoneum. For pathological diagnosis, laparoscopic partial omentectomy and ileal mesenteric LNs biopsy were performed. Hypovolemic shock developed on postoperative day 1, and a CT scan revealed spontaneous splenic rupture as the main cause; emergency splenectomy was performed to combat this complication (Fig. 2A). Three days after splenectomy, the patient developed hematochezia, leading to hypovolemic shock. CT tomography revealed small bowel bleeding in the ileum, for which emergency surgery was performed. The main small bowel lesion was identified 60 cm from the ileocecal valve and resected (Fig. 2B). Pathological examination revealed a segment of ileum with atrophic villi and transmural involvement of densely infiltrated lymphoma cells, which are relatively monomorphic small and round (Fig. 3). Immunohistochemical stain-

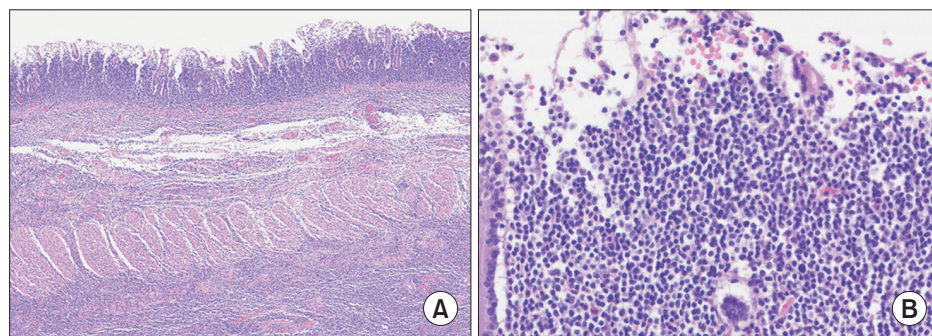




**Fig. 1.** An abdominal computed tomography scan shows multifocal segmentally enhanced wall thickening of the small bowel, omental cake, and a large amount of ascites. (A) Transverse view image. (B) Coronal view image. Positron emission tomography shows (C) moderate hypermetabolism in the ileum and (D) several low-attenuated lesions with hypermetabolism in the enlarged spleen.



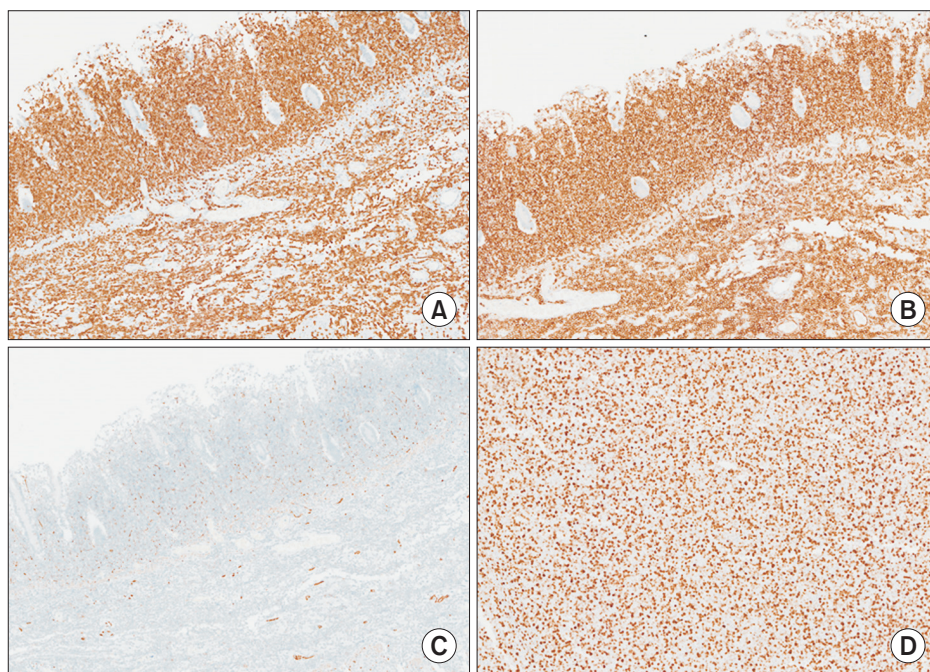
**Fig. 2.** Macroscopic finding of (A) spontaneously ruptured spleen and (B) main lesion of small bowel T-cell lymphoma leading to massive hematochezia.



**Fig. 3.** Microscopic findings of (A) a segment of ileum with transmural involvement of lymphoma cell (H&E, ×5), (B) atrophic villi and densely infiltrated lymphoma cells, which are relatively monomorphic small round. A few intraepithelial lymphoma cells are seen (H&E, ×400).

ing revealed that tumor cells are CD3+ and CD8+ (Fig. 4). Histological examination confirmed T-cell lymphoma as the cause of splenic rupture, and bone marrow examination further confirmed its presence. The final diagnosis was

intestinal T-cell lymphoma, suspected to be monomorphic epitheliotropic intestinal T-cell lymphoma (MEITL). The patient was referred to an oncologist and started on a regimen of cyclophosphamide, hydroxydaunorubicin, oncovin,



**Fig. 4.** Immunohistochemical stains. (A) CD3 positive, (B) CD8 positive, (C) CD56 negative, and (D) Ki-67 high proliferative activity ( $\times 100$ ).

and prednisolone (CHOP). However, five days after the first cycle of CHOP therapy with 50% dose reduction due to a recent surgery, the patient complained of severe abdominal pain, and a CT scan showed small-bowel perforation. Emergency small bowel resection was performed, and pathological examination revealed transmural involvement of intestinal T-cell lymphoma. The patient is currently receiving CHOP therapy.

## DISCUSSION

Intestinal T-cell lymphoma is a rare and aggressive form of non-Hodgkin lymphoma primarily affecting the GI tract [7]. It may involve different parts of the GI tract; however, the stomach is the most commonly affected site, while the small bowel is the second most common [8]. A recent World Health Organization classification system, revised in 2017, classified intestinal T-cell lymphoma into four types: enteropathy-associated T-cell lymphoma (EATL), MEITL, intestinal T-cell lymphoma not otherwise specified, and indolent T-cell lymphoproliferative disorder of the GI tract [9]. EATL is the most common subtype of intestinal T-cell lymphoma, more commonly observed in Europe and closely associated with celiac disease. In contrast, MEITL has no

apparent association with celiac disease, and its incidence is higher in Asian and Hispanic populations and among men and middle-aged individuals [10,11]. The small intestine, especially the jejunum, is the most common site of MEITL [12].

The clinical presentation of intestinal T-cell lymphoma varies considerably between studies; however, common symptoms include abdominal pain, bloating, diarrhea, weight loss, and fatigue [7,13]. These symptoms may mimic other GI conditions, leading to a delayed diagnosis. As EATL is associated with celiac disease, it is often preceded by symptoms such as diarrhea and malabsorption, which are not usually detected in MEITL [14,15]. In addition, owing to its aggressive nature, intestinal T-cell lymphoma can quickly spread to other organs, such as the liver, spleen, and lymph nodes. Because disease progression can lead to severe complications, including intestinal obstruction or perforation, it is often diagnosed incidentally through the occurrence of these complications [13]. In the current case, the patient presented with abdominal pain and subsequently experienced spontaneous splenic rupture, GI hemorrhage, and small bowel perforation.

Diagnosis of intestinal T-cell lymphoma requires a combination of clinical evaluations, imaging studies, and laboratory tests. Endoscopic procedures, such as upper GI endos-

copy and colonoscopy, are often performed to obtain biopsy samples from the affected intestinal area. These samples are then examined under a microscope to confirm the presence of abnormal T cells and to determine the specific lymphoma subtype. However, a previous multicenter prospective study reported that only 20% of intestinal T-cell lymphoma cases were diagnosed endoscopically [13]. In addition, endoscopically obtained tissues are often insufficient to accurately diagnose intestinal T-cell lymphoma. Moreover, its rarity also contributes to the difficulty of obtaining an accurate diagnosis. Consequently, patients are sometimes misdiagnosed with CD, intestinal tuberculosis, or cancer [6]. Immunophenotyping may help diagnose intestinal T-cell lymphoma and differentiate between disease subtypes. CD3+, CD4-, and CD5+ cells are typically identified in both EATL and MEITL. Additionally, MEITL cells can display CD8+, CD 56+, and CD 30- phenotypes [7,10].

The prognosis of intestinal T-cell lymphoma is generally poor, primarily because the disease is generally diagnosed at an advanced stage and displays aggressive behavior. Patients with MEITL often present with advanced-stage disease at the time of diagnosis, and the median overall survival of these patients has been reported as 14.8 months [10,16]. The 5-year survival rate for EATL has been estimated at 20% or less [13,17,18]. The treatment of intestinal T-cell lymphoma has proven challenging owing to its aggressive nature and the lack of standardized therapeutic approaches; currently, chemotherapy is the primary treatment for this condition. CHOP therapy has been widely adopted as an alternative treatment strategy; however, the response to treatment is generally poor [13,19,20]. Surgery is not recommended as a standard treatment, but is often performed for the diagnosis, treatment, and prevention of disease complications [6].

In conclusion, I reported a case of intestinal T-cell lymphoma with multiple complications. Intestinal T-cell lymphoma is often associated with severe complications; however, its rarity, variable presentation, and clinical course make diagnosis challenging. Clinical awareness and collaboration between gastroenterologists and oncologists are essential for timely diagnosis and appropriate management of the disease.

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## CONFLICTS OF INTEREST

No potential conflict of interest relevant to this article was reported.

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