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Extensive involvement of indolent T-cell lymphoma in a patient with ulcerative colitis

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Dear Editor,

A 53-year-old male presented with frequent stool, abdominal pain and hematochezia, and was diagnosed with ulcerative colitis in 2000. He was treated with Chinese herbal, tumor necrosis factor-α (TNF-α) inhibitor (infliximab) and lucocorticoids, but the efficacy was limited. Due to gastrointestinal bleeding, he underwent interventional embolization of the superior rectal artery and surgical resection of affected intestine successively. Computed tomography (CT) showed portal hypertension, ileocecal wall thickening enlarged multiple intra-abdominal lymph nodes, splenomegaly and hepatomegaly (Fig. 1A and B). Endoscopy showed multiple, segmental, intestinal large ulcers (Fig. 1C). In 2022, the patient underwent a splenectomy, liver and bone marrow biopsy due to giant spleen and severe pancytopenia.
Microscopy revealed diffuse infiltration of mildly atypical lymphoid cells in the intestinal sections (Fig. 1D). Immunohistochemistry was performed on bone marrow, spleen, liver and intestinal segment tissues. All tissues were positive for CD3, CD8, TIA-1 and TCR gene rearrangement, and negative for CD4, CD20, CD56 and GrB. Thus confirming the infiltration of tumor cells. The Ki-67 proliferation index was about 5% (Fig. 1E-G). Based on this, a diagnosis of gastrointestinal indolent T-cell lymphoproliferative disorder (GI T-LPD) was established. After surgery, the patient developed massive peritoneal effusion, breathlessness and a serious infection. Having been treated with mechanical ventilation and other supportive therapies, he was discharged after his condition improved two months later.

Discussion
GI T-LPD is a rare subtype of lymphoma. The symptoms are non-specific and may lead to misdiagnosis (1). Different from inflammatory bowel disease (IBD), nearly all GI T-LPD show a clonality in lymphoid proliferation with rearrangement of TCR genes (2). Until now, the molecular signatures underlying GI T-LPD remain largely unknown. A standard therapy for GI T-LPD has not yet been determined. Sometimes, steroid therapy, involved-field radiation therapy or resection of the lesion can control symptoms (3,4).

In this case, extensive lesions invaded the bone marrow, liver, spleen and intestine of the patient, which may be due to the long-term progression or the use of immunosuppressive agents and biologics. A well-conducted systematic review addressed the association between T-cell lymphoma and anti-TNF therapy for IBD (5). Anti-TNF therapy is known to cause lytic activation of Epstein-Barr virus (EBV). Coincidentally, the disease progressed with EBV activation in the present case, which is in line with previous mechanistic hypothesis. Nevertheless, additional pathogenic factors may exist in this case, and more studies are needed to investigate the effects of immunosuppressive agents and biologics on the progression of GI T-LPD.

References


Fig. 1. A. Computed tomography (CT) scan demonstrating portal hypertension and tortuous splenic vein. B. Intestinal wall edema and thickening; multiple enlarged lymph nodes in the abdomen, 1 to 2 cm in diameter. C. Endoscopy demonstrating multiple, segmental and big ulcerations. D. Microscopy revealed diffuse infiltration of lymphoid cells which showed mild atypia. E-G. Immunohistochemistry of spleen tissue, positive for CD3 (F) and TIA-1 (E); Ki-67 proliferation index was about 5% in the tumor cells (G).