Indolent NK-cell lymphoproliferative disorder of the gastrointestinal tract

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Caso clínico comentado

A 25-year-old female with a history of abdominal distension, abdominal pain, and intermittent melena for 8 months. Eight months prior to admission (February 2024), the patient experienced abdominal distension and pain of an episodic nature, accompanied by intermittent melena (1-2 times per day). The patient had been diagnosed with anxiety disorder over 4 years ago, with multiple visits to the neurology outpatient clinic and intermittent oral treatment with "Quetiapine Fumarate Tablets and Calcium Carbonate." During the course of the illness, the patient's mental state, diet, and sleep were fair, with normal bowel and urinary habits. She experienced a weight loss of 3 kg over six months (height: 145 cm, weight: 60 kg).

Colonoscopy performed at an external hospital indicated multiple colonic ulcers of undetermined nature. Small bowel CTE revealed: slight mucosal enhancement in the rectum and proximal ascending colon, with a suspected small ulcer in the ascending colon. No typical manifestations of ulcerative colitis or Crohn's disease were observed. A chest CT showed small nodules in both lungs, likely inflammatory, and a small amount of pericardial effusion. Pathological findings indicated: chronic inflammatory cell infiltration in the mucosa of the terminal ileum, transverse colon, descending colon, sigmoid colon, and rectum, with slightly disordered crypt architecture and no granuloma formation. TB-PCR was negative. During this period, the patient received traditional Chinese medicine (details unspecified), and her abdominal pain symptoms improved. For follow-up evaluation, she was admitted to our hospital.Gastroscopy indicated multiple Gastric ulcers and duodenal ulcers (Figura 1). Colonoscopy also indicated multiple colonic ulcers and terminal ileitis (Figura 2). Pathological showed lymphocytic infiltrate in the gastric and colonic mucosa (Figura 3). Immunohistochemistry showed positivity for CD3, CD4, CD7, CD56, TIA-1, and GranB, with Ki-67 (40%) and CK (epithelial+). Negative for CD20, CD5, CD8, CD21, CD34, CD123, and MPO (Figura 4). Gene rearrangement tests (TCRB, TCRD, TCRG, IGH, IGK, IGL) were negative, as were tests for EB virus, CMV, and TB. Capsule endoscopy suggests multiple ulcers in the small intestine (Figura 5). We conducted a multidisciplinary consultation and discussion. Based on immunohistochemical, gene rearrangement resultspathology, CT imaging, and endoscopic examination, we made a diagnosis of indolent NK-cell proliferative lesion of the gastrointestinal tract.

Figura 1. *Gastroscopy*

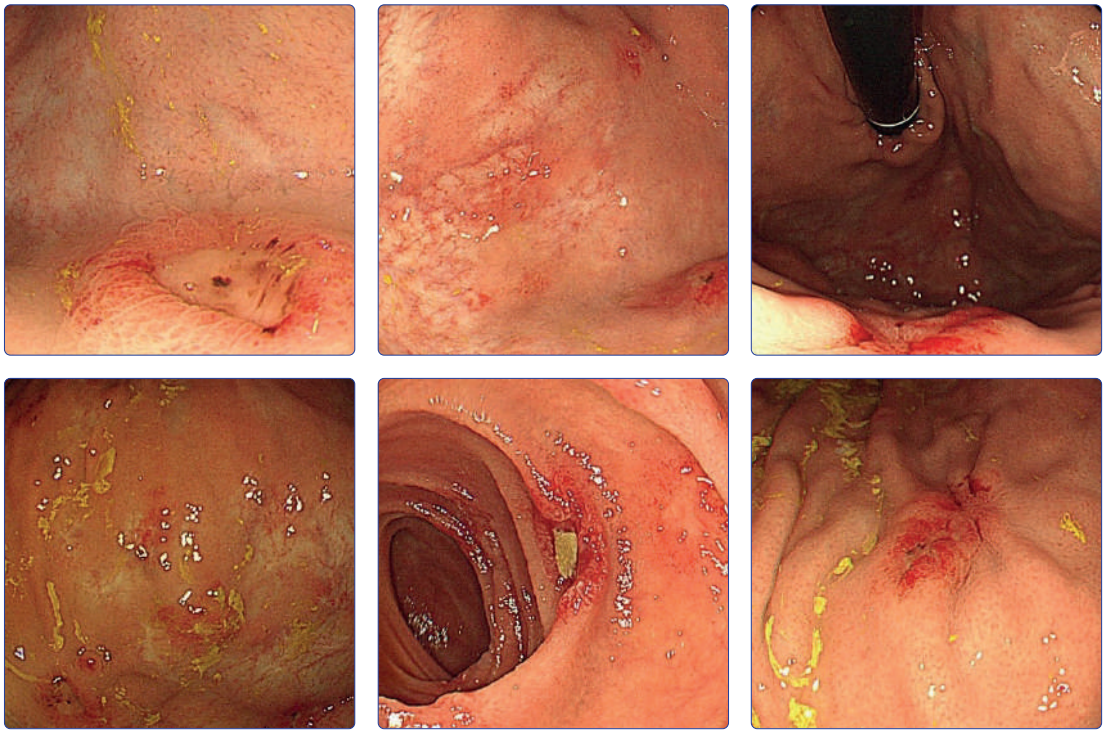


Figura 2. *Colonoscopy*

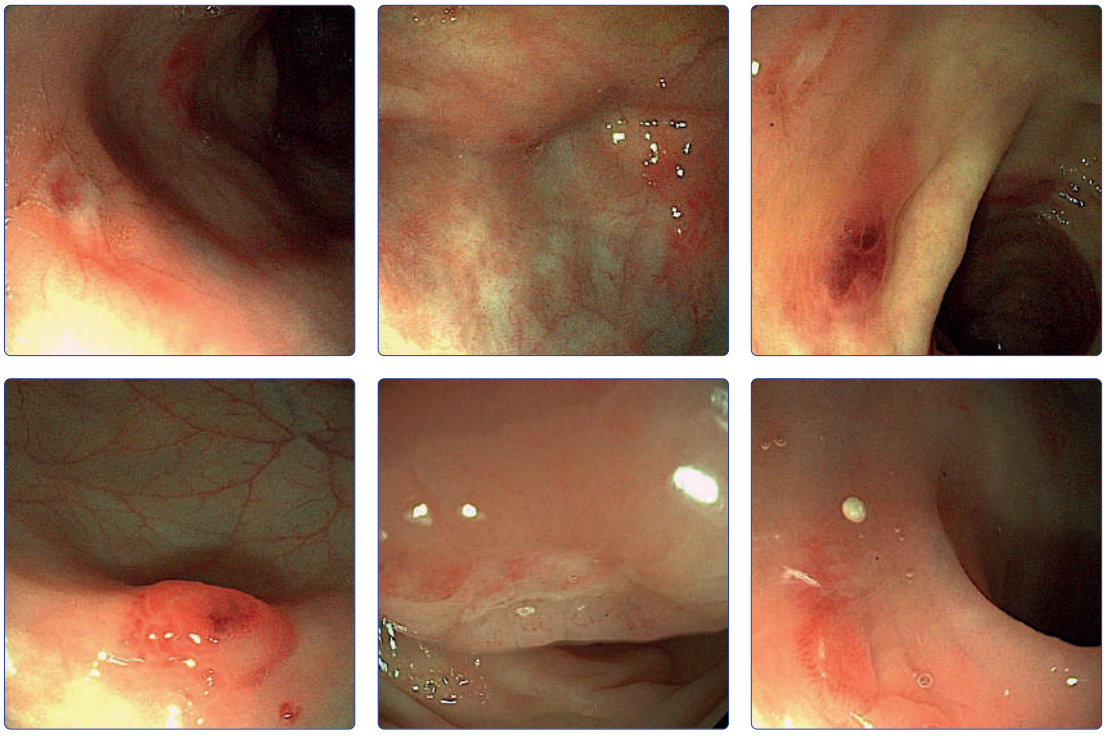


Figura 3. *Histopathology*

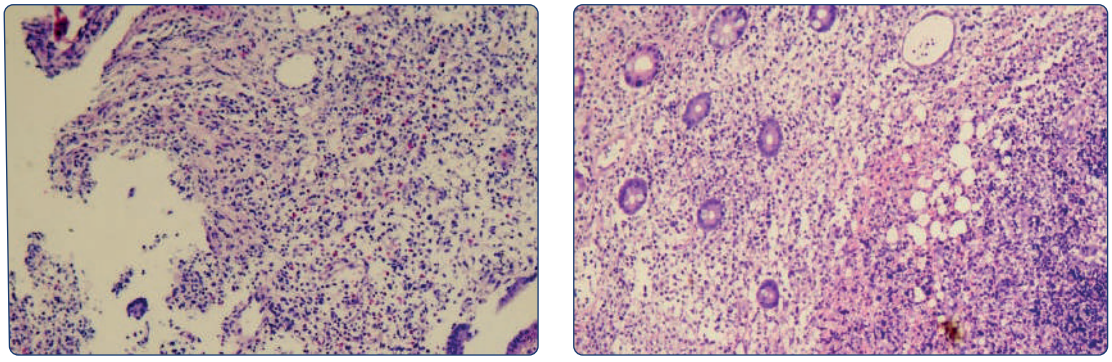


Figura 4. *Immunohistochemistry*

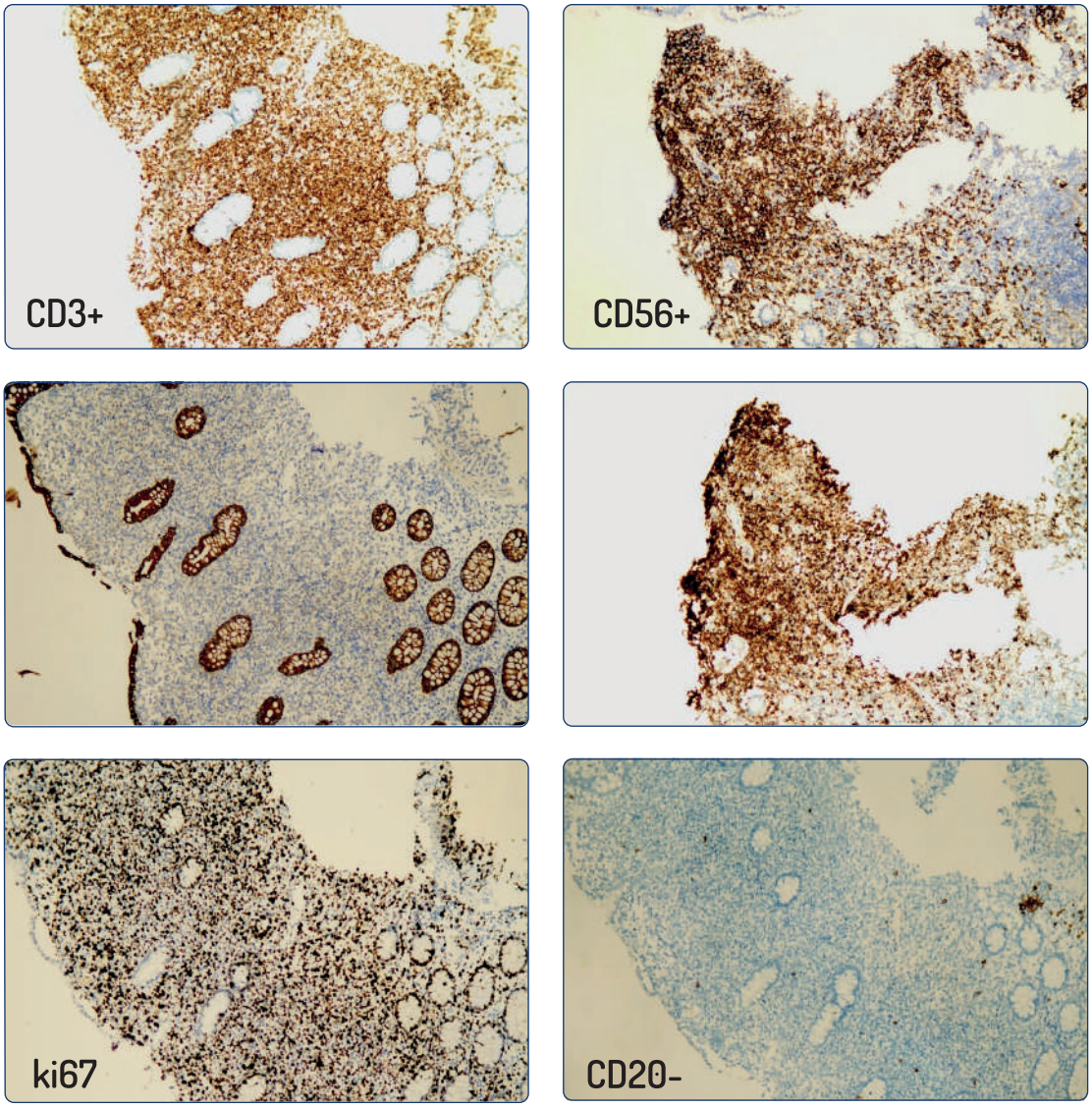
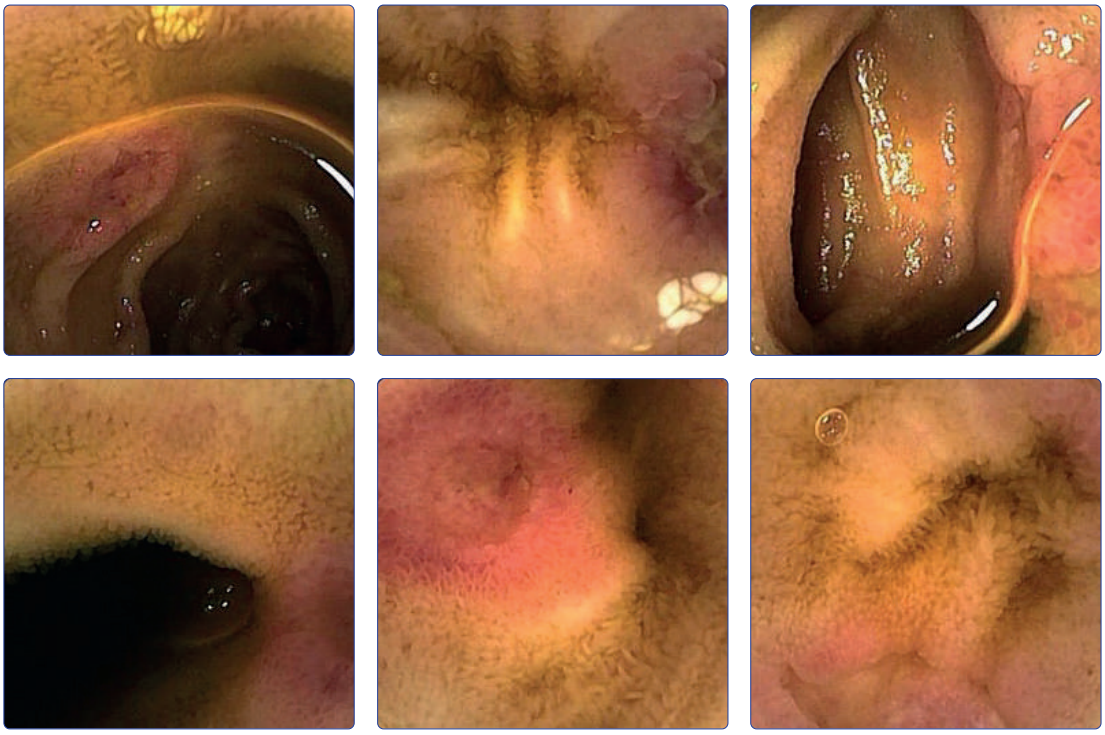


Figura 5. *Capsule Endoscopy*



Discussion

Indolent NK-cell lymphoproliferative disorder of the gastrointestinal tract is a lymphoproliferative disease primarily involving NK/T cells in the digestive system. It can affect the entire gastrointestinal tract, with the ileum and colon being the most commonly involved sites. Endoscopic findings include erosions, ulcers, and erythema, while isolated small intestinal lesions are extremely rare. Most cases undergo a phase of misdiagnosis (e.g., ulcerative colitis [UC], Crohn's disease [CD], intestinal tuberculosis [ITB]) [1, 2]. This disease has been classified in the 5th edition of the World Health Organization (WHO) Classification of Haematolymphoid Tumours [3, 4].

Its clinical characteristics include: Unknown etiology, possibly related to food allergies, Helicobacter pylori infection, or immune dysregulation. Non-specific gastrointestinal symptoms. Absence of hepatosplenomegaly or lymphadenopathy. Affected organs: stomach, duodenum, small intestine, colon, and occasionally the gallbladder. Endoscopic manifestations: shallow ulcers and polypoid changes. Slow progression and poor response to chemotherapy, [1, 2, 5].

Histopathological examination using hematoxylin-eosin (HE) staining revealed presents as medium-to-large lymphocytes expanding in the lamina propria, with round to irregular nuclei, mature chromatin, and variably prominent nucleoli. The cytoplasm is moderately abundant and pale, occasionally containing eosinophilic granules. While the lesions may be locally destructive, they lack angioinvasion or necrosis. Immunohistochemical profiling demonstrated positive expression of CD3, CD7, CD56, TIA-1, Granzyme B, and CD4, CD5, CD8 are negative,and. EBV-encoded RNA (EBER) is also negative.with a Ki-67 proliferation index of 10%–30%. Mutations such as JAK3 and K563_C565del may be observed[1.2.4.5].

Indolent NK-cell lymphoproliferative disorder of the gastrointestinal tract needs to be differentiated from inflammatory bowel disease, intestinal tuberculosis, , and Enteropathy-associated T-cell lymphoma. The clinical course of this disease is indolent and there is no special treatment method. Follow up is generally recommended (every 6-12 months).

Reference

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