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Case study

Indolent peripheral T-cell lymphoma involving the gastrointestinal tract

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Summary We describe an unusual case of indolent peripheral T-cell lymphoma with multifocal involvement of the gastrointestinal tract. The patient, a 42-year-old Asian man, has been followed up for more than 10 years without chemotherapy and multiple gastrointestinal biopsies showing similar findings. Histologically, the neoplasm expanded into the lamina propria and/or focally extended into the submucosa and was composed of small- to medium-sized lymphocytes with slightly irregular nuclear contours and clear cytoplasm and rare large lymphocytes. The tumor cells were positive for CD3, CD8, granzyme B, and TIA-1 (subset) and negative for CD5, CD56, and Epstein-Barr virus–encoded RNA. Molecular studies for T-cell receptor γ and/or β chain gene rearrangement demonstrated the same clone at different sites and times during the course of the disease. Rare cases of indolent peripheral T-cell lymphoma of the gastrointestinal tract have been previously described and need to be further characterized to avoid the use of aggressive chemotherapy. Published by Elsevier Inc.

1. Introduction

Peripheral T-cell lymphomas involving the gastrointestinal (GI) tract are rare, with enteropathy-associated T-cell lymphoma being the most frequent T-cell lymphoma arising in the small intestine [1,2]. Both type I and II enteropathy-associated T-cell lymphoma are associated with poor prognosis even after aggressive chemotherapy [3,4]. Although rare, other specific types of T-cell lymphoma that can present in the intestine include extranodal natural killer (NK)/T-cell lymphoma, nasal type, and γ/δ T-cell lymphoma [5]. Extranodal NK/T-cell lymphoma, nasal type, is characteristically positive for Epstein-Barr virus and has a poor prognosis [4,6]. γ/δ T-cell lymphomas with intestinal involvement [5] are clinically aggressive and usually positive for CD8, CD56, and cytotoxic proteins. Peripheral T-cell lymphoma, not otherwise specified, also can involve the GI tract.

Here, we report a case of peripheral T-cell lymphoma with multifocal involvement of the GI tract and an indolent clinical course characterized by persistent lymphoid infiltrate without evidence of large-cell transformation or disease progression. Rare cases of low-grade mucosal/intestinal T-cell proliferation have been previously described with similar clinicopathological features and may represent a unique entity that needs to be recognized to prevent the use of unnecessary or aggressive chemotherapy.

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2. Case presentation

2.1. Clinical history

A 42-year-old Asian man with history of peptic ulcer disease in 1987 presented in June of 1992 at another institution for upper endoscopic studies. At that time, a gastric biopsy was diagnosed as T-cell lymphoma. The patient was referred to our institution in March of 1993 for further evaluation and treatment. According to the clinical notes, at that time, the complete blood count showed white blood cell count 2.7 × $10^3/\mu$L with 71% neutrophils; hemoglobin, 12.3 g/dL; hematocrit, 36.2%; and platelet count 103 × $10^3/\mu$L. Physical examination showed splenomegaly with palpable tip of the spleen 2 to 3 cm beyond the left costal margin. Serologic studies were positive for hepatitis B surface antibody and hepatitis B core antibody, and they were negative for hepatitis B surface antigen, hepatitis C, and HIV 1 and 2 antibodies. Endoscopy of the upper gastrointestinal (GI) tract was performed and showed

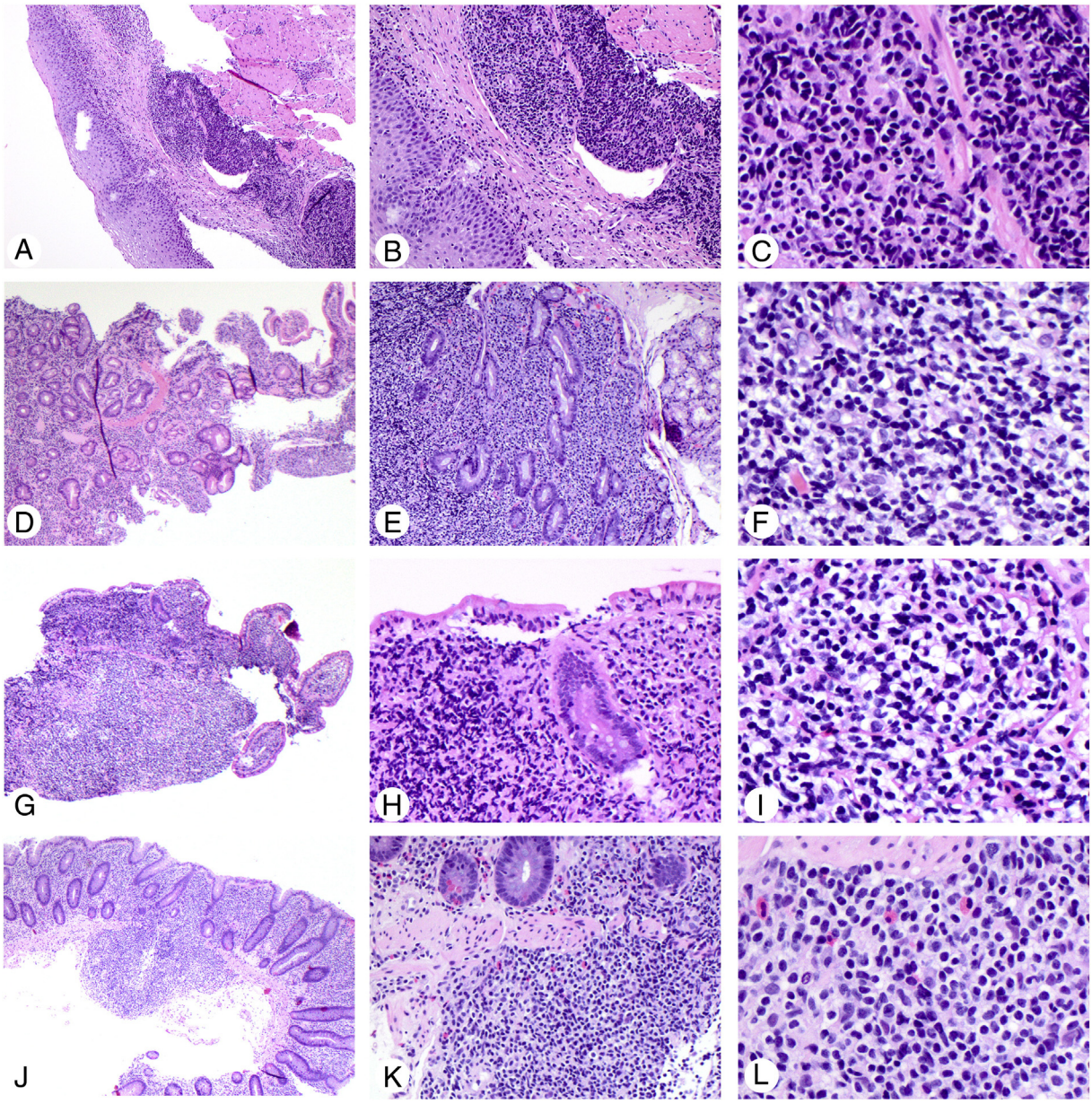

Fig. 1 Histologic features of endoscopic biopsy specimens from the GI tract. A-C, Esophageal biopsy specimen (2007) showed an atypical infiltrate of small lymphoid cells with irregular nuclei in the submucosa focally extending into the muscle. Duodenal biopsy specimens from 2008 (D) and 2012 (E and F) showed similar histologic findings including a lymphocytic infiltrate in the lamina propria without significant epitheliotropism. G-I, Biopsy specimens of the terminal ileum (2010). J-L, Colon biopsy specimens (2008) showed lymphocytic infiltrate in the lamina propria and focally in submucosa with preservation of colonic architecture and no acute inflammation.