

Primary small-bowel adult T-cell leukemia/lymphoma with gastric AL amyloidosis



Fig. 1 Primary small-bowel adult T-cell leukemia/lymphoma with gastric AL amyloidosis. Small bowel enteroclysis demonstrates a narrowing edematous lesion with multiple ulcerations in the jejunum.



Fig. 2 Capsule endoscopy shows multiple ulcers with enlarged Kerckring folds in the jejunum.



Fig. 3 Double-balloon endoscopy reveals enlarged Kerckring folds with annular and irregularly shaped shallow ulcers in the jejunum.

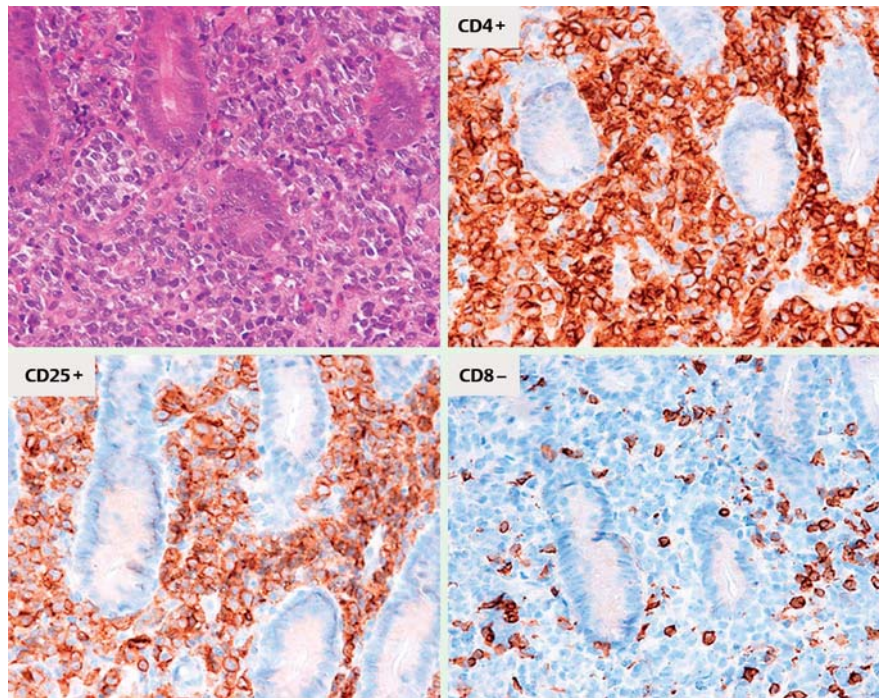


Fig. 4 Histology of a biopsy specimen from the jejunal ulcer showing diffuse infiltration of medium to large pleomorphic lymphoid cells with CD4+, CD25+, CD8- immunophenotype (magnification $\times 400$).

An 86-year-old man was admitted because of small-bowel wall thickening and mesenteric lymphadenopathy. Neither peripheral lymphadenopathy nor hepatosplenomegaly was noted. The leukocyte count was $7930/\text{mm}^3$, and no abnormal lymphocytes were found. The result of testing for serum antihuman T-lymphotropic virus type 1 (HTLV-1) antibody was positive. A bone marrow biopsy was normal without evidence of lymphoid infiltration. Small-bowel enteroclysis demonstrated a narrowing, edematous lesion with multiple ulcerations in the jejunum (Fig. 1). Capsule endoscopy (Fig. 2) and double-balloon endoscopy (Fig. 3) showed enlarged Kerckring folds with annular and irregularly shaped shallow ulcers. Biopsy specimens from the ulcers revealed diffuse infiltration of medium to large pleomorphic lymphoid cells with CD3+, CD4+, CD5+, CD25+, CCR4+, FoxP3+, CD8-, CD20-, CD56-, and TIA1- immunophenotype (Fig. 4). Biopsy specimens from the stomach, duodenum, ileum, and colon showed no lymphoma cells. Fluorodeoxyglucose F 18 positron emission tomography showed abnormal uptake in the jejunal mass, wide areas of mesentery, and

para-aortic lymph nodes. Esophagogastroduodenoscopy with chromoendoscopy revealed granular stomach mucosa and swollen folds in the corpus, with pan-atrophic gastritis (Fig. 5). Biopsy from the granular mucosa and folds showed massive amyloid deposits of homogeneous acidophilic substances in the submucosa and around the muscularis mucosae (Fig. 6), which were immunohistochemically positive for λ light chain. Amyloid deposits were not observed in any specimen taken from the duodenum, jejunum, ileum, and colon. Therefore, the patient's diagnosis was stage IIE (Lugano classification) small-bowel adult T-cell leukemia/lymphoma (ATLL) accompanied by gastric AL amyloidosis.

Although systemic ATLL often involves the small bowel [1–3], primary small-bowel ATLL is extremely rare [4,5]. Endoscopic findings of small-bowel involvement with ATLL have included multiple polypoid or reddish elevated lesions [2,3]. However, we believe that the enlarged Kerckring folds with annular or irregularly shaped shallow ulcers observed in our case may be characteristic in primary small-bowel ATLL.

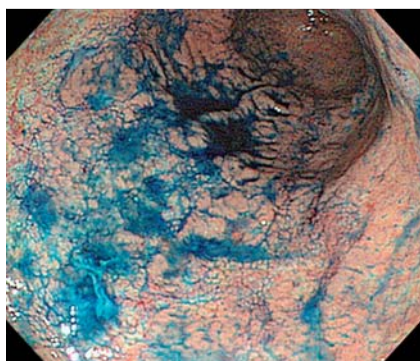


Fig. 5 Esophagogastroduodenoscopy reveals granular mucosa and swollen folds in the corpus, with pan-atrophic gastritis.

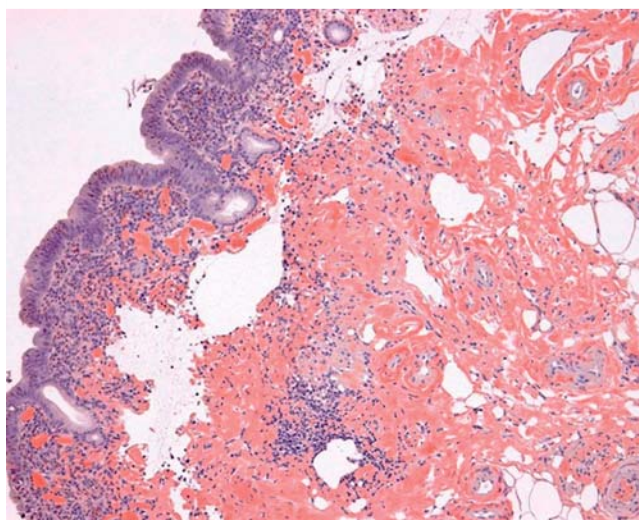


Fig. 6 Histology of a biopsy specimen from the granular gastric mucosa showing massive amyloid deposits of homogeneous acidophilic substances in the submucosa and around the muscularis mucosae (Dylon stain, magnification $\times 100$).

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Competing interests: None

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Bibliography

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