Endoscopic features of Cronkhite–Canada syndrome

Cronkhite–Canada syndrome (CCS) is a rare protein-losing enteropathy and profound malnutrition disease with high mortality [1–3]. CCS can be challenging to differentiate from other polyposis syndromes [4]. We describe the endoscopic features of CCS before and after treatment.

A 69-year-old woman presented with chronic diarrhea, body pigmentation, weight loss, and dysgeusia. Examination showed alopecia, glossitis, onycholysis, and palmar pigmentation. Her laboratory values showed abnormally low albumin (26 g/dL), hemoglobin (11 g/dL), and zinc (658 µg/L). Computed tomogram of the abdomen showed thickened folds and polyp-like protrusions in the stomach.

We performed an upper endoscopy, which showed normal esophagus. Upon entering the stomach, multiple large inflammatory polyps covering the antrum and body were seen (►Fig. 1). The duodenum showed mucosal edema, villous blunting, and atrophy. Similarly, colonoscopy revealed extensive large inflammatory polyps throughout the colon. The ileum appeared edematous with villous atrophy (►Fig. 2). Biopsies performed from the gastric polyps showed mild infiltration with inflammatory cells, submucosal edema, and tortuous hyperplastic foveolar glands (►Fig. 3). Based on the clinical features, characteristic endoscopic appearance, and histopathology findings, we diagnosed the patient to have CCS. We treated her using a tapering dose of prednisolone, azathioprine, and proton pump inhibitors. We provided oral nutritional supplements and corrected the micronutrient deficiency. After 3 months of treatment, her symptoms started to resolve. We repeated upper endoscopy and colonoscopy, which showed a regression of her gastric and colonic polyps (►Fig. 4, ►Fig. 5). The villous atrophy and blunting in the small bowel had reversed. We have reviewed our other similar cases, followed over an

►Fig. 1 Upper endoscopy at diagnosis: **a** multiple large inflammatory polyps in the gastric antrum; **b** edematous duodenal mucosa with villous atrophy.

►Fig. 2 Colonoscopy at diagnosis: **a** large inflammatory polyps carpeting the colon; **b** villous atrophy in the ileum.

►Fig. 3 Biopsy from the gastric polyp: **a** mild infiltration with inflammatory cells with submucosal edema; **b** tortuous foveolar gland hyperplasia.

► **Fig. 4** Upper endoscopy after 3 months of treatment: **a** decrease and disappearance of inflammatory gastric polyps; **b** reversal of villous blunting in the duodenum.

► **Fig. 5** Colonoscopy after 3 months of treatment: **a** disappearance of colonic inflammatory polyps; **b** improvement in villous blunting and edema.

► **Video 1** Endoscopic features of Cronkhite–Canada syndrome.

extended time (3 years), that were treated with long-term immunosuppressants (► **Video 1**). The patients have remained asymptomatic during the follow-up time. In conclusion, recognizing the distinct and specific endoscopic features of CCS

may allow it to be diagnosed and differentiated from other polyposis syndromes (► **Video 1**). Uniquely, in CCS, the multiple large inflammatory polyps may be reversed with treatment [3, 5].

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Competing interests

Dr. Roy Soetikno is a consultant for Olympus and Fujifilm.

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Bibliography

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