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
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].

References


Bibliography

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