A Case of Cronkhite–Canada Syndrome Involving the Entire Gastrointestinal Tract

We encountered a case of a 56-year-old woman with Cronkhite–Canada syndrome who presented with chronic diarrhea, alopecia, intermittent abdominal pain, hyperpigmentation, and nail dystrophy (Figure **1**). Her face, palms, and the back of her hands and soles of her feet were deeply pigmented with small dark brown spots. Her family history was noncontributory. Serum total protein was 5.4 g/dl, with 2.4 g/dl of albumin. Endoscopic examination of the stomach showed multiple sessile polyps, varying in size from 2 mm to 10 mm throughout the entire stomach and duodenum (Figure **2**). Small-bowel x-ray and colonoscopic examination revealed multiple polyps from the jejunum to the rectum. Histopathologic examination of these polyps revealed edema of the lamina propria, and mucosal erosion associated with evidence of chronic inflammation. Scintigraphy with technetium-99m-labeled human albumin demonstrated a protein-losing enteropathy. Hyperalimentation (2200kcal/d) was continued for 8 weeks. At 1 week later, the patient's diarrhea showed a decrease in severity, with improvement in appetite and weight gain. However, the diarrhea recurred after 2 weeks. After administration of prednisolone, 30 mg daily, and trimethoprim with sulfamethoxazole (Bactrim), clinical improvement was noted, with cessation of diarrhea, increased serum protein, disappearance of pigmentation, and regrowth of the scalp hair.

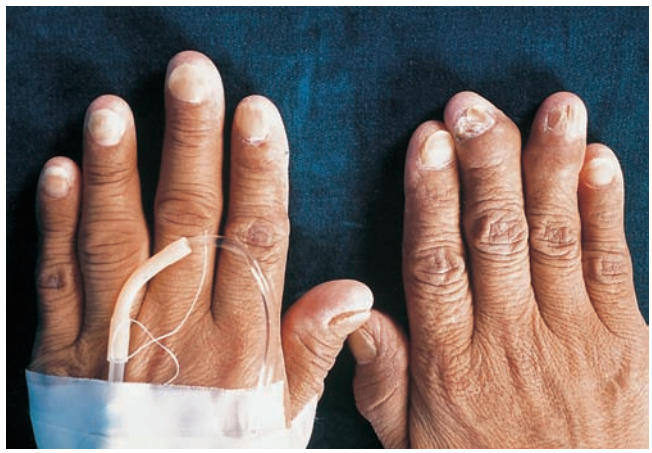

Figure 1 The patient's nails showed dystrophy such as splitting and partial separation from the nail bed

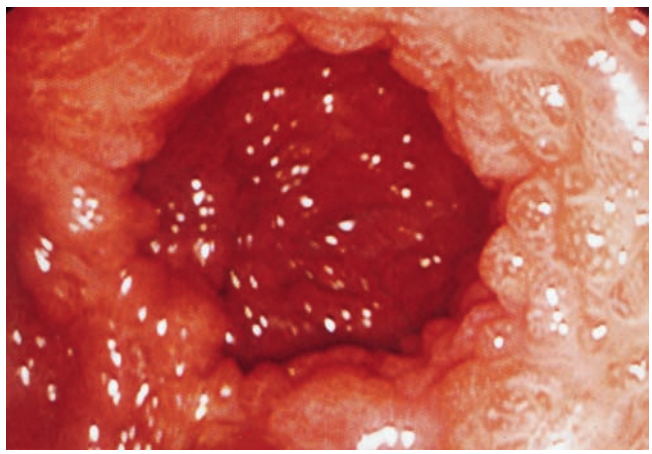

Figure 2 Endoscopic view of the stomach, showing multiple sessile polyps, varying from 2 mm to 10 mm in size

Cronkhite–Canada syndrome has a poor prognosis because of malnutrition resulting from altered absorption in the gastrointestinal tract [1]. The diarrhea and hypoproteinemia seem to arise as a result of protein loss into the gastrointestinal lumen. Many cases have had a fatal outcome [1, 2], with a reported 6-month survival rate of 40 %, but some cases of spontaneous remission have also been reported [3]. A partial recovery following administration of prednisolone suggests that it may be effective in preventing a leakage of plasma protein into the gastrointestinal tract. The accumulating evidence for remission in Cronkhite–Canada syndrome supports suggestions that it might have nutritional, infective, toxic, or other exogenous causes [1, 2]. Malnutrition and metabolic disturbances may be responsible for the observed ectodermal changes [4]. Although the possibility of a purely coincidental association between enteral nutrition and recovery cannot be excluded, the sequence of events suggests that nutritional support should be tried early in the course of this illness.

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