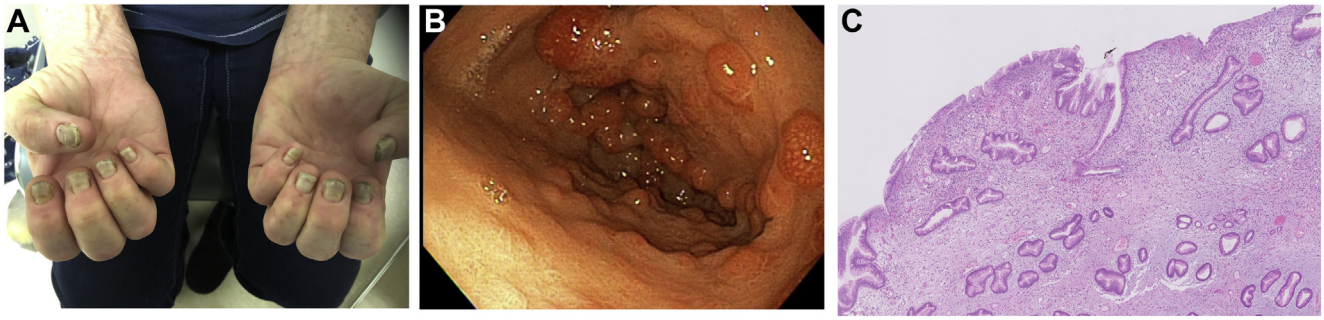
Nausea and Diarrhea with Fingernail and Hair Changes

Peter P. Stanich,[1] Giovanni Lujan,[2] and Amy Hosmer[3]

[1]Division of Gastroenterology, Hepatology and Nutrition, Ohio State University Wexner Medical Center, Columbus, Ohio; [2]Department of Pathology, Ohio State University Wexner Medical Center, Columbus, Ohio; and [3]Division of Gastroenterology and Liver Disease, Digestive Health Institute, University Hospitals Cleveland, Cleveland, Ohio



Question: A 58-year-old woman presented with 5 months of nausea with vomiting and diarrhea. Her symptoms were severe and she had noted a 25-pound weight loss over this time period. She had hypoalbuminemia noted on laboratory assessment. She had already seen a gastroenterologist and received upper endoscopy with reported inflammation in the antrum and duodenal bulb with pathology showing active gastritis with eosinophilic infiltrate as well as a colonoscopy showing only adenomatous polyps with unremarkable random colon biopsies. She received an empiric course of steroids, given the eosinophilic infiltrate, and had improvement in her symptoms with rapid return when they were stopped.

Given the lack of a diagnosis and her persistent severe symptoms, she was referred for further evaluation. On our examination, she was noticed to have fingernail deformities (Figure *A*) and she reported thinning of her hair since symptom onset. On upper endoscopy, she was noted to have multiple inflammatory-appearing friable gastric polyps in the antrum (Figure *B*), with the largest estimated to be 15 mm in size. Histologic assessment of the polyps showed hyperplastic epithelium with an inflamed and edematous lamina propria. These were reported as hyperplastic polyps and chronic inflammation (Figure *C*).

Based on the examination and endoscopic findings, what is the most likely diagnosis?

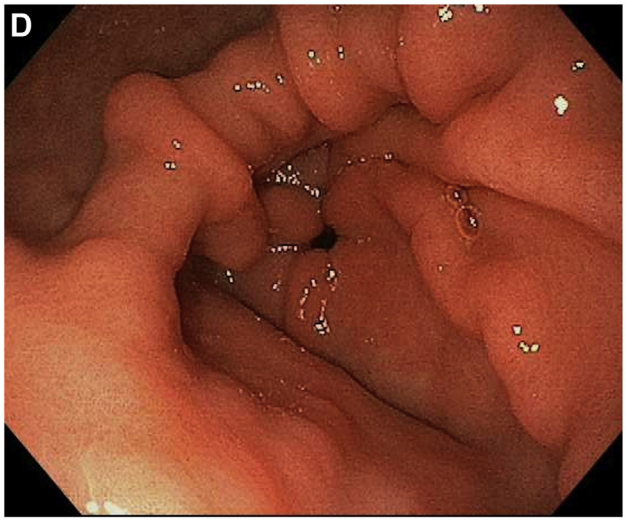
See the *Gastroenterology* website (www.gastrojournal.org) for more information on submitting your favorite image to Clinical Challenges and images in GI.

Correspondence
Address correspondence to: Dr Peter P. Stanich, 395 W 12th Ave, Columbus, OH 43210. e-mail: peter.stanich@osumc.edu.

Conflicts of interest
Dr Stanich receives research support from Emtora Biosciences, Janssen Pharmaceuticals, Pfizer, and the PTEN Research Foundation. Dr Lujan and Dr Hosmer have no conflicts of interest or pertinent disclosures.

0016-5085/$36.00
https://doi.org/10.1053/j.gastro.2020.10.048

Answer to Image: Cronkhite-Canada Syndrome



A diagnosis of Cronkhite-Canada syndrome was made based on the significant gastric polyposis in conjunction with her ectodermal findings (onychodystrophy and alopecia) and diarrhea. She was started on a steroid taper and transitioned to azathioprine. Her symptoms improved rapidly on this therapy and have not returned. At a repeated upper endoscopy after 1 year, complete resolution of gastric polyposis was found (Figure *D*), and at 2 years of therapy her nail changes had resolved (Figure *E*).

Cronkhite-Canada syndrome is a rare condition characterized by abdominal symptoms such as diarrhea and abdominal pain, gastrointestinal hamartomatous polyposis, and ectodermal changes such as alopecia and onychodystrophy.[1] The histologic features of the hamartomatous polyps are nonspecific and can overlap with juvenile and hyperplastic polyps, and as such it is imperative to place them in the context of the clinical history and examination.[2] Cronkhite-Canada syndrome is thought to be an autoimmune condition and responds to treatment with corticosteroids and other immunosuppressants, with azathioprine successfully used for long-term maintenance therapy.[1]

Keywords: Cronkhite-Canada Syndrome; Diarrhea; Hamartoma; Onychodystrophy.

References

1. Sweetser S, Ahlquist DA, Osborn NK, Sanderson SO, Smyrk TC, Chari ST, et al. Clinicopathologic features and treatment outcomes in Cronkhite-Canada syndrome: support for autoimmunity. Dig Dis Sci 2012;57:496–502.
2. Burke AP, Sobin LH; The pathology of Cronkhite-Canada polyps. A comparison to juvenile polyposis. Am J Surg Pathol 1989;13:940–946.