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Case Report

Panendoscopic characterization of Cronkhite–Canada syndrome

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Introduction

Cronkhite–Canada syndrome (CCS) is rare non-familial disorder of unknown etiology, characterized by gastrointestinal cystic polyposis and ectodermal manifestations. Less than 500 cases have been reported in the world literature so far, and most of these cases were reported from Japan.[1–3] Only a handful of cases of CCS have been reported in this part of the world.[3–5] Here, we describe the clinical features and panendoscopic findings of a patient with CCS.

Case report

A 68-year-old male patient presented with a 3-month history of hypogeusia, anorexia, alopecia, generalized hyperpigmentation, and 15-kg weight loss. Physical examination revealed generalized hyperpigmentation, onychodystrophy (Figs. 1 and

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2A) and partial alopecia. Routine blood investigations were normal. Blood tests for Addison's disease, thyroid disease and celiac disease were normal. Patient had negative result on antinuclear antibody (ANA) testing. Abdominal ultrasonography and chest radiograph were normal. CT-scan showed concentric thickening of gastric antrum, pylorus and duodenum (D1). Esophagogastroduodenoscopy (EGD) showed hypertrophy of gastric folds and polypoidal lesion in gastric antrum. EGD also showed partial atrophy and scalloping of duodenal folds. Gastric biopsies revealed edema, infiltration by lymphoplasmacytic cells and eosinophils and myxoid expansion of muscularis propria and submucosal cystic dilations. *H. Pylori* organism was seen in gastric biopsies. Patient was treated with 2-weeks of anti-*H. Pylori* therapy (clarithromycin, amoxicillin, and pantoprazole) followed by 6-weeks of PPI therapy. There was an improvement in symptoms such as dyspepsia, anorexia and hyperpigmentation. But, the patient developed watery diarrhea. Stool examination including assay for *Clostridium difficile* toxin was normal. Repeat EGD revealed partial improvement in endoscopy appearances. Eradication of *H. Pylori* was confirmed in repeat gastric biopsies. Single balloon enteroscopy further disclosed partial atrophy in the jejunal folds (Fig. 3). Ileocolonoscopy revealed multiple sessile polyps located throughout the colon (maximum size 1.5 cm; some with a strawberry-like appearance), nonspecific colitis, and occasional tiny ulcerations and mucosal congestion of ileum (Figs. 4 and 5). Duodenal biopsies showed complete villous atrophy, hyperplasia of crypt and normal intraepithelial lymphocyte cells. Edema, lymphoplasmacytic and esinophilic infiltration and myxoid expansion of muscularis propria were noted in biopsies from stomach, duodenum and colon (Fig. 6A). Colonic polyp biopsies revealed sessile polyps with variably sized cystically dilated glands suggestive of juvenile/hamartomatous polyp (Fig. 6B).

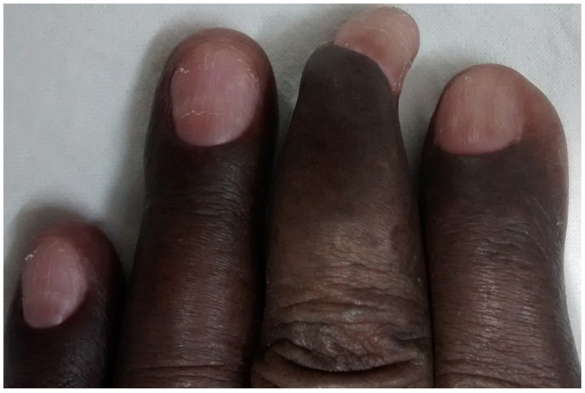

Fig. 1 – Onychodystrophy of fingernails.

Patient was diagnosed as CCS, and treated with systemic steroid therapy (oral prednisolone 40 mg daily, tapered down slowly during a period of 2-months to 20 mg), proton pump inhibitors (PPI), and nutritional support. Nine-months later, substantial symptomatic improvement noted in the patient's condition such as resolution of diarrhea, improvement of the skin hyperpigmentation, increase in appetite, and regrowth of his hair (Fig. 2B). However, minimal improvement in nail dystrophy was seen. Repeat EGD and colonoscopy showed significant improvement of endoscopic findings (Figs. 7 and 8).

Discussion

The Cronkhite–Canada syndrome was first described in 1955 by Cronkhite and Wilma Jeanne Canada.[6] The mean age of onset is estimated to be in the 5th–6th decade with a male: female ratio of 1.3–2.3:1. It is a disease with bad prognosis and a

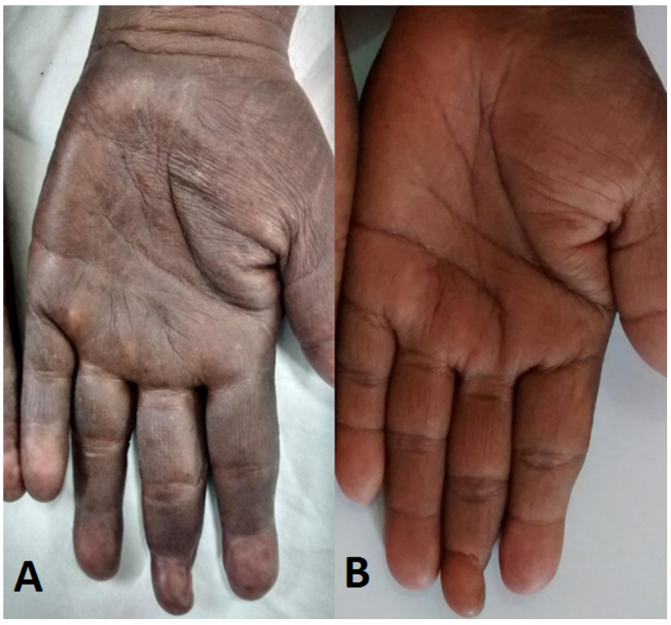

Fig. 2 – Hyperpigmentation before (A) and after treatment (B), note post-traumatic deformed middle finger.

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