CRONKHITE-CANADA SYNDROME - REPORT OF A CASE AND LITERATURE REVIEW

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ABSTRACT

This article reported a case of Cronkhite–Canada syndrome.The patient was 56 years old, presented with diarrhea, anorexia and weight loss. On physical examination, there were glossitis, hair loss, onychomadesis, palmar and sole hyperpigmentation. Laboratory revealed evidence of nutritional deficiencies. Radiological and panendoscopic studies showed gastrointestinal polyposis in the stomach, duodenum and colon. Biopsies of all polyps revealed hyperplastic polyps with retention cystic gland dilatation. After 6th month of proper neutritional support, his condition improved strikingly and ectodermal change disappeared. Colonic polyposis showed almost complete regression and partial regression of the gastric polyposis. This case is not reported for its rarity. Emphasis is made for early recognition with prompt and appropirate therapy to avoid its potentially high morbidity and mortality.

INTRODUCTION

The Cronkhite-Canada syndrome is a noninherited condition, sporadically occurring disorder affecting adults. It is characterized by a tetrad of cutaneous hyperpigmentation, alopecia, onychodystrophy, and intestinal polyposis.[1] This syndrome has a worldwide distribution with a fairly acute onset and a rapidly progressive course.

The following case is not reported for its rarity; emphasis is made on early recognition with a prompt and appropriate management to avoid fatality which may occur within a short time.

CASE REPORT

A 56-year-old diabetic man with residual cerebro-vascular accident was admitted to the medical ward, Ramathibodi Hospital on June 22th, 1998 because of hypogeusia and lack of desire for food. Two months prior to the admission, he had diarrhea lasted for 10 days, followed by 5-kg weight loss. His hair turned gray with excessive shedding. There were nails separation and hyperpigmented area over palms and soles. One year prior to the hospitalization following the development of left-sided numbness, examination with MRI and nerve conduction velocity revealed multiple ischemic foci and peripheral neuropathy. On examination, he had premature gray hair, baldness over frontal part. There were atrophic tongue papillae (Fig.1), onychomadesis (Fig.2). Rectal examination revealed nodular mucosa.

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Laboratory studies: Hemoglobin 13.3 g/dl, WBC 9,400 /ml with PMN 74% lymphocyte 21%, MCV 98 um^3 ; stool occult blood was positive. FBS 151 mg/dl, BUN 6 mg/dl, Cr 0.7mmol/L, sodium 137 mmol/L, potassium 5.12 mmol/L, chloride 105 mmol/L, CO2 26.3 mmol/L, calcium 7.5 mg/dl, phosphate 3.4 mg/dl, total protein 61.3 G/L, albumin 35.5 G/L, total bilirubin 0.2 mg/dl, direct bilirubin 0.1 mg/dl, alkaline phosphatase 66 u/L, Cholesterol 85 mg/dl, SGOT 28 u/L, SGPT 14 u/L; normal coagulogram, B12 level 100 pg/ml (300-900). Upper GI and small bowel series revealed polyposis in the stomach, duodenal bulb and 2nd part of duodenal loop (Fig.3). Barium enema revealed diffuse mucosal and submucosal conglomerated nodules involving the entire colon, appendix and rectum; large polypoid mass was seen at distal transverse colon and inferior part of rectum (Fig.4). Gastroscopy revealed multiple nodules and sessile polyps in the swollen mucosa of stomach and duodenal loop. Colonoscopy up to hepatic flexure revealed diffused sessile and pedunculated polyps in swollen mucosa (Fig.5). Biopsy of the polyps at the gastric antrum, duodenum and colon was performed. Microscopically, the polyps were lined by normal corresponding mucosa,the cores or stalks were composed of broadened lamina propria with slight degrees of smooth muscle cell proliferation in some polyps. A considerable amount of inflammatory infiltration consisting of lymphocytes, plasma cells and eosinophils was observed in the broadened lamina propria. Pathological diagnoses of all lesions were hyperplastic polyps. Both intramuscular and oral vitamin B12 were given.

Two weeks after the discharge, hypogeusia and palmar hyperpigmentation decreased. Three months later, he gained 4 kgs weight. Laboratory examinations : vitamin B12 level 495 pg/ml, serum folic acid 3.37 ng/ml (Normal 5-24), RBC folic acid 165 ng/ml (Normal 211-1113). Multivitamins and trace elements were added.

Six months after the discharge, he was asymptomatic; premature gray hair, glossitis, onychomadesis and palmar hyperpigmentation disappeared (Fig.6). He gained more weight. Laboratory tests were: total protein 63 G/L, albumin 36 G/L, calcium 8.8 mg/dl, cholesterol 135 mg/dl, B12 level 708 pg/ml, serum folic acid 10.04 ng/ml, RBC folic acid 264 ng/ml. Gastroscopic examination showed decrease in number and extent of gastric and duodenal polyposis; the mucosa was less swollen. Bacterial culture of jejunal aspiration grew no organism. Colonoscopic exmination detected only 5-6 small polyps (Fig.7). D-xylose test revealed 3.64 g (normal > 1.2 g) of 5- gram D-xylose obtained in the 5-hr urine specimen. Sudan stain of feces showed no fat globules.

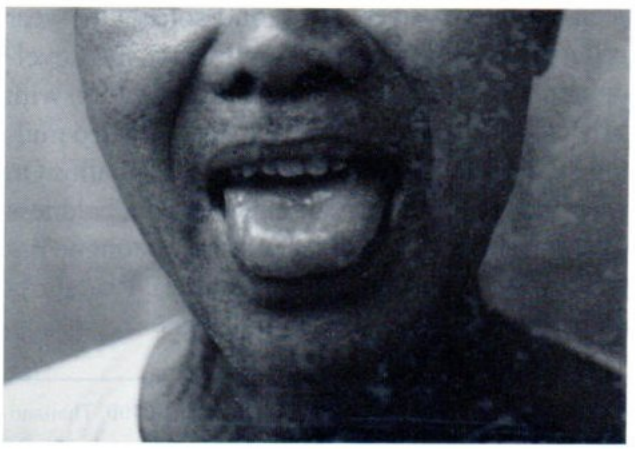

Fig. 1. Glossitis before treatment was shown in the patient with Cronkhite Canada syndrome.

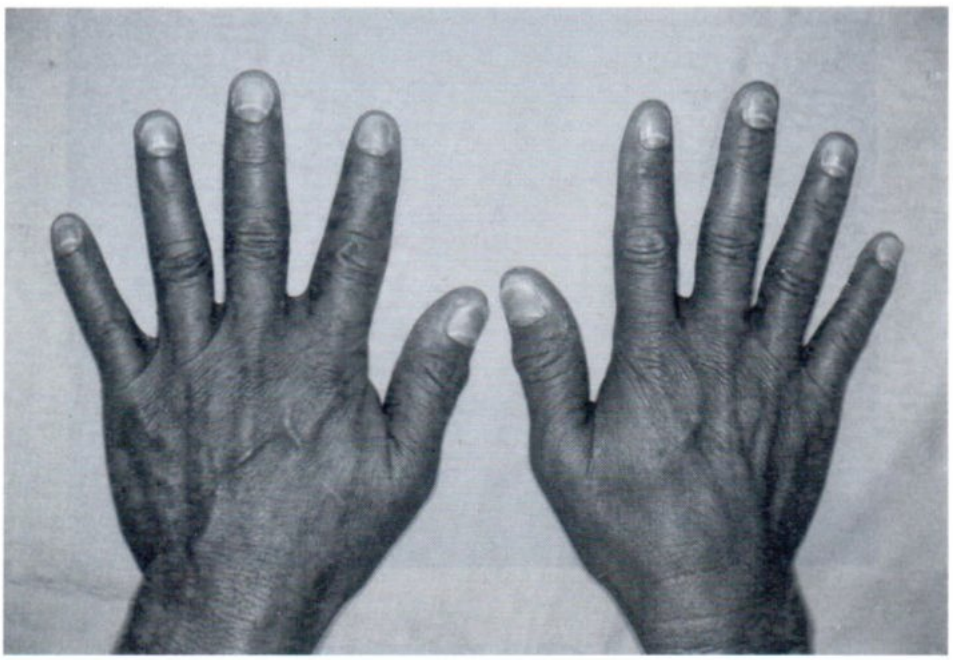

Fig. 2. Onychomadesis before treatment was seen.

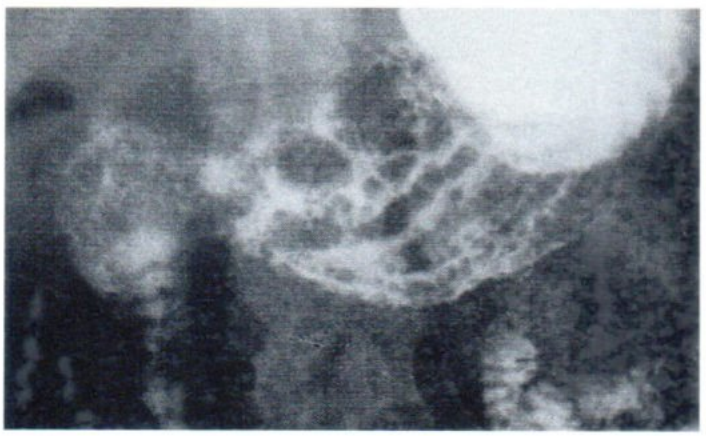

Fig. 3. UGI series demonstrated polyposis in the stomach, duodenal bulb and the 2nd part of the duodenal loop.

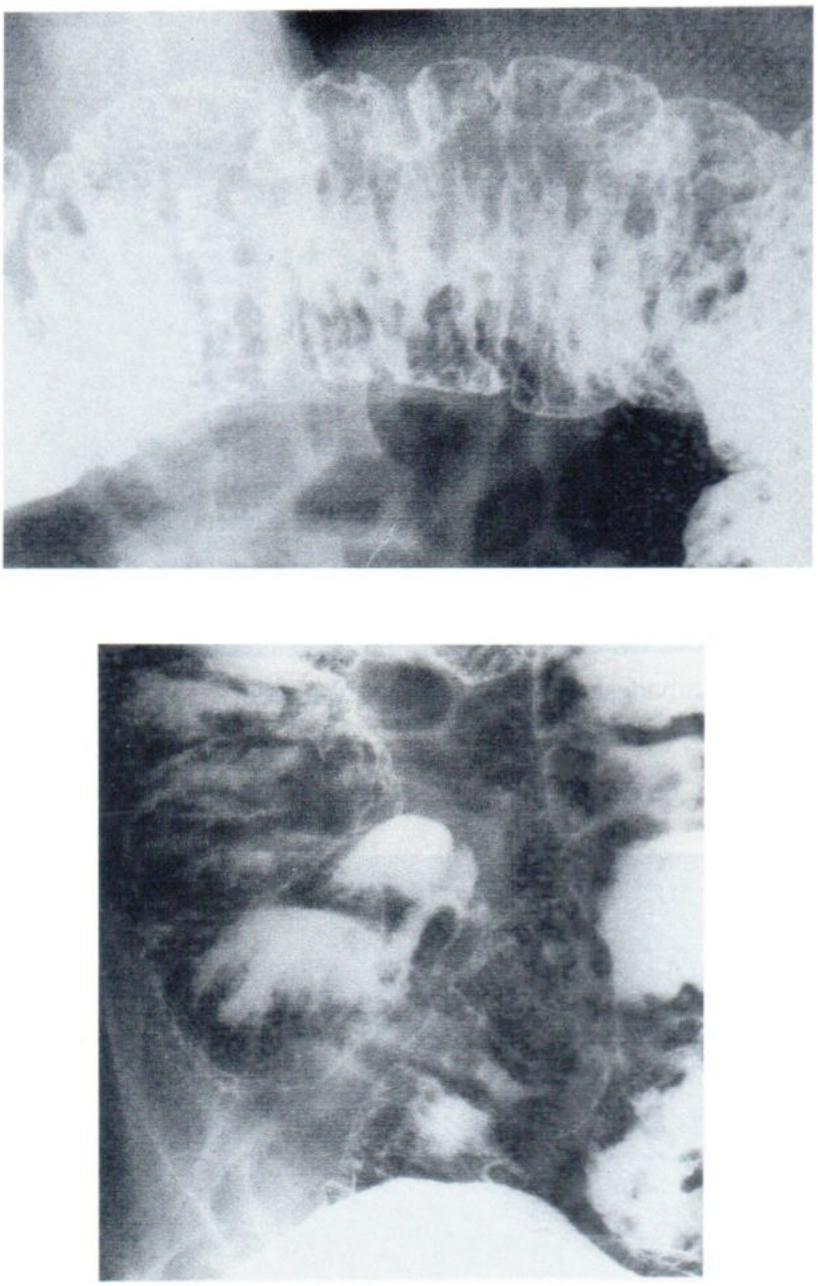

Fig. 4. Barium enema revealed numerous polyps in the entire colon.

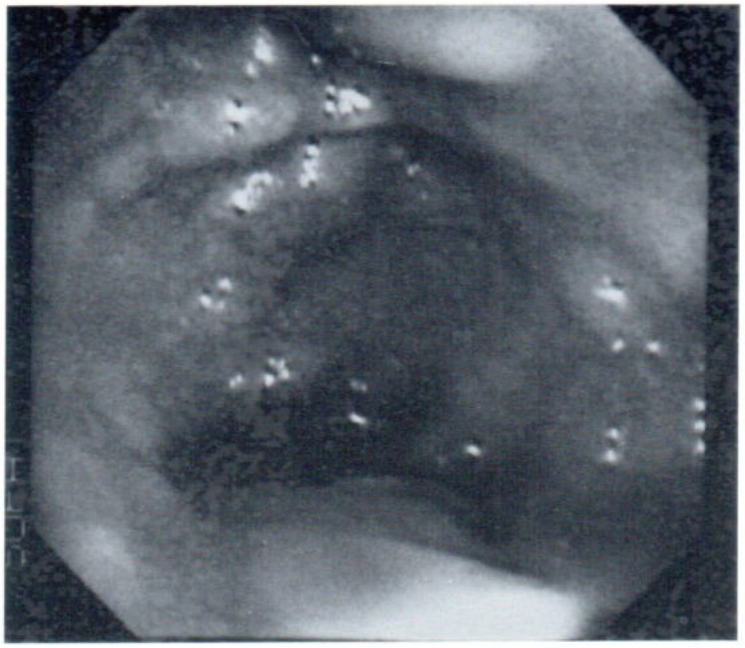

Fig. 5. Colonoscopy revealed diffuse sessile and pedunculated polyps in the swollen mucosa.

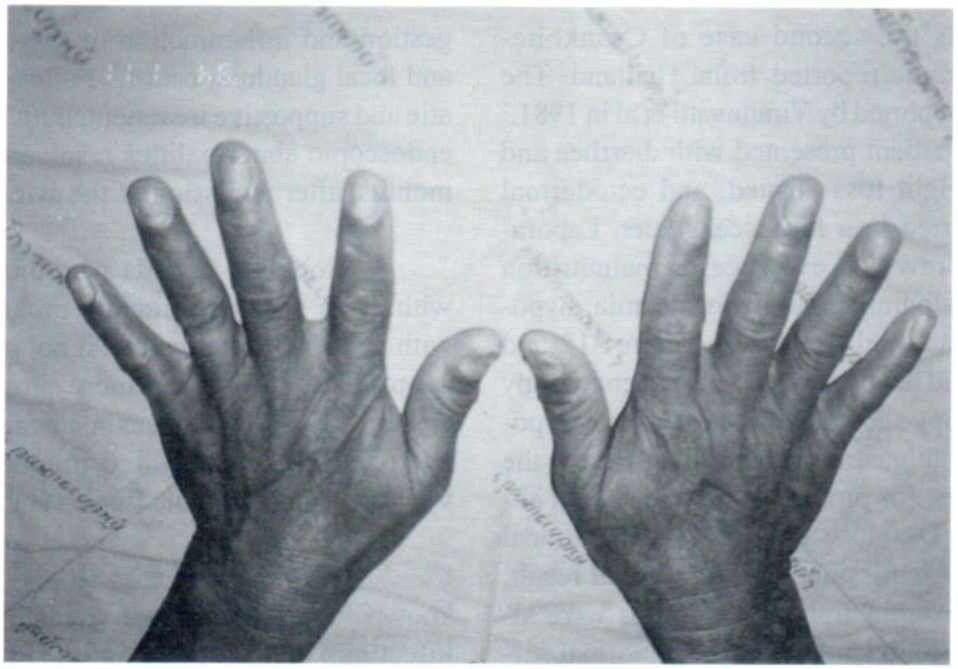

Fig. 6. Normal looking nailplates was shown in 6 months after nutritional support.

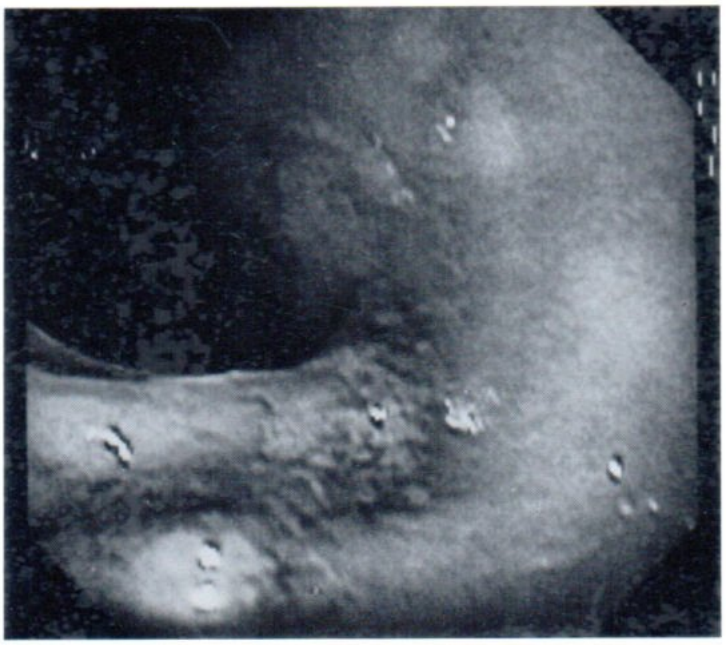

Fig. 7. Few colonic polyps were noted at colonoscope in 6 months after nutritional support.

DISCUSSION

This is the second case of Cronkhite-Canada syndrome reported from Thailand. The first case was reported by Viranuvatti et al in 1981.[5] Our reported patient presented with diarrhea and anorexia. Weight loss ensued, and ectodermal changes occurred in a few weeks later. Laboratory findings revealed evidence of malnutrition such as hypoalbuminemia, hypocalcemia, hypocholesterolemia; and decrease vitamin B12 and serum folic acid levels. Radiologic, panendoscopy and colonoscopy showed diffused sessile polyposis of the whole gastrointestinal tract except the esophagus. Test for proximal small intestinal absorption with D-xylose performed after clinical and laboratory remission, showed normal result. Histological examination of the polyps were almost identical to juvenile polyps, i.e. normal surface epithelium, proliferated tortuous gland with cystic dilatation and edematous chronically inflammed lamina propria.[6] However, in contrast to juvenile polyposis, the mucosa between polyps was also histologically abnormal with edema, congestion and inflammation of the lamina propria and focal glandular ectasia. Following symptomatic and supportive treatment, both laboratory and endoscopic abnormalities resolved within a few months, after remission of the symptoms.

Cronkhite-Canada syndrome may present with acute onset and rapidly progressive course with up to 50% fatality rate, if not recognized and appropriately treated, a more protracted course ensues with severe cachexia, anemia, congestive heart failure and impaired immunity, resulting variously in pneumonia, sepsis and shock.[1]

The cause of this illness remains unknown.[1,3] Various etiologies including nutritional, infectious and immunologic causes have been mentioned. Spontaneous remission of Cronkhite-Canada syndrome,[10] including the report by Viranuvatti et al,[5] have been reported in the literature. In our patient, only symptomatic and supportive treatment including nutritional supplemen-

tation were given and the patient appeared to be heading towards spontaneous remission.

The mainstay of therapy in this condition is proper nutritional support. Antibiotic may be useful if bacterial overgrowth is suspected.[1,3] Systemic corticosteroid and anabolic steroid have been reportedly helpful in isolated case. Surgery is hardly recommended except only when complications such as bleeding, intussusception take place. Malignancy could happen. (15% of the patients in one report).[1,3,8]

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