

Radiological features of Cronkhite-Canada syndrome

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ABSTRACT

This report describes the radiological features of a case of Cronkhite-Canada syndrome. Barium meal follow through with pneumocolon, used as a single radiological test, revealed diffuse polyposis involving the stomach and both the small and large intestines, as well as findings of gastric rugal thickening, whiskering, crypts and ulcerated polyps.

Keywords: barium meal follow through with pneumocolon, Cronkhite-Canada syndrome, polyposis

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INTRODUCTION

Cronkhite-Canada syndrome (CCS) is a rare non-hereditary gastrointestinal polyposis of unknown aetiology that is associated with ectodermal changes. This syndrome was first described in the year 1995.⁽¹⁾ To the best of our knowledge, only 400 cases of this syndrome have been reported, the majority of case reports being from Japan.⁽²⁾ Very few cases have been reported from the Indian subcontinent.^(3,4) The radiological features of this syndrome have previously been described by only a few authors.⁽⁵⁻⁷⁾ Barium meal follow through with pneumocolon is a single, simple and cost-effective test which adequately depicts the features of CCS.⁽⁸⁾ The present case is being reported due to its rarity and typical radiological features.

CASE REPORT

A 65-year-old Indian man presented with a history of significant weight loss, diarrhoea, pain in the abdomen and loss of taste. Physical examination revealed average health with hyperpigmentation of the skin and nail changes. The patient's serum albumin was 2.6 gm/dl. Complete blood count, liver function tests, blood urea nitrogen, serum creatinine, thyroid function tests and stool examination were normal.

Barium meal follow through examination with pneumocolon was performed, which showed a normal



Fig. 1 Radiograph of the stomach after double contrast barium study shows rugal thickening (white arrow), multiple duodenal and small bowel (lines) and ulcers (black arrow).

oesophagus. Double contrast films of the stomach and duodenum (Figs. 1–5) showed multiple polyps of up to 1.6 cm in size in the fundus and antrum of the stomach with mosaic pattern, whiskering, crypts and thickened rugal folds. Pooling of barium was also noted in the stomach, which was suggestive of ulcerated/ruptured polyps. Multiple polyps at the base of the duodenal cap, and tiny polyps in the second part of the duodenum and the proximal jejunum were also observed. All the polyps were sessile. Sessile polyps were also noted in the rest of the small intestine (Fig. 6). Delayed follow through film with pneumocolon showed polyps in the large intestine and terminal ileum. The size of the polyps in the large intestine was up to 2.5 cm. The “whiskering” sign was also noted in the transverse colon (Figs. 7–9). All the polyps were sessile. Gastroscopy and colonoscopy revealed a mosaic pattern in the stomach and multiple sessile polyps in the stomach, duodenum and large intestine. On biopsy, they were found to be hyperplastic in the stomach and of the juvenile type in the duodenum and colon.

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Fig. 2 Radiograph of the stomach after double contrast barium study shows whiskering (black arrow) and fundal polyp (white arrow).



Fig. 3 Radiograph of the stomach after double contrast barium study shows a mosaic pattern in the stomach (arrow) and multiple small as well as minute filling defects in the duodenum due to polyps.

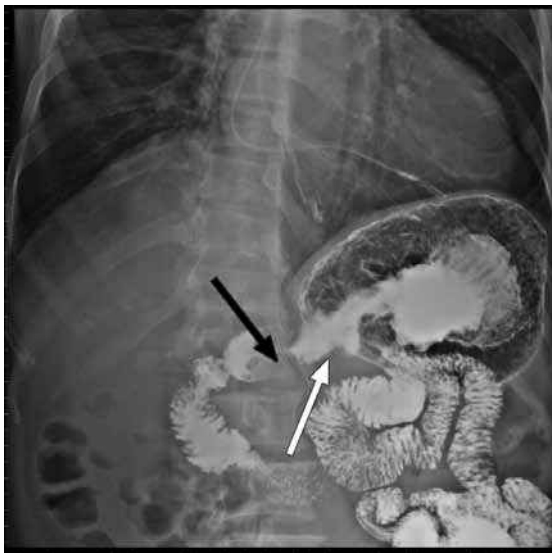


Fig. 4 Radiograph of the stomach after double contrast barium study shows whiskering (white arrow) and multiple polyps at the base of the duodenal cap (black arrow) and the second part of the duodenum.



Fig. 5 Radiograph of the stomach after double contrast barium study shows rugal thickening (white arrow) and polyps in the antrum and duodenal cap (black arrow).

DISCUSSION

CCS is a non-familial gastrointestinal polyposis syndrome. The first two cases of CCS were described by Cronkhite and Canada in the year 1995.⁽¹⁾ To the best of our knowledge, more than 400 cases have been reported so far in the literature, two-third of which are from Japan, with a male preponderance (male-female ratio is 2:1). The most common age of onset of this disease is 50–60 years. The disease has characteristic features of diffuse gastrointestinal polyposis and ectodermal changes (alopecia, hyperpigmentation and

onychodystrophy). Gastrointestinal polyposis leads to malabsorption and protein-losing enteropathy, resulting in clinical symptoms of abdominal pain, diarrhoea and weight loss.⁽²⁾

Goto published the largest series of 110 cases of CCS from Japan. The disease was staged into five categories based on the clinical presentation and subsequent clinical course, viz. diarrhoea (35%), hypogeusia (40.9%), xerostomia (6.4%), abdominal discomfort (9.1%) and alopecia (8.2%).⁽⁹⁾ Hypogeusia (taste disturbance) is related to zinc deficiency. The histological features of



Fig. 6 Radiograph of the small bowel shows multiple filling defects (arrows) due to polyps.



Fig. 7 Delayed radiograph shows multiple small polyps in the ileum (black arrow) and whiskering in the transverse colon (white arrow).

CCS include the universal finding of hamartomatous polyps of a juvenile (retention) type throughout the gastrointestinal tract except in the oesophagus, with chronically inflamed lamina propria, cystically dilated and tortuous glands filled with proteinaceous fluid or inspissated mucus. The polyps are almost always sessile, and the mucosa between them is abnormal.⁽²⁾ A hyperplastic type of polyps has also been described.⁽¹⁰⁾

The differential diagnoses of polyps include juvenile polyposis, hyperplastic polyposis, lipomatous polyposis, nodular lymphoid hyperplasia, inflammatory polyposis, lymphomatous polyposis, Peutz-Jeghers polyposis and Menetrier's disease. Confusion can also arise with villous atrophy, *Helicobacter pylori* gastritis, eosinophilic gastroenteritis and intestinal candidiasis.⁽²⁾ The aetiology of CCS is uncertain. In the Japanese population, mental and physical stress has been suggested as a cause of CCS.⁽¹¹⁾ Also, the unique involvement of two epithelial tissues in CCS suggests a loss of normal proliferative stimuli as a cause.⁽¹¹⁾ IgG4-related autoimmune disease has also been speculated as a cause of CCS.⁽¹²⁾

The prevalence of serrated adenomatous polyps in patients with CCS was noted in 40% of cases with microsatellite instability. Overexpression of the p53 protein was found in a CCS patient with serrated adenoma and cancer, thus proposing the possibility of a serrated adenoma-carcinoma sequence underlying some CCS gastrointestinal malignancies.⁽¹³⁾ The overall prevalence of gastrointestinal malignancy in CCS is about 10%.⁽²⁾

The association of CCS with hypothyroidism, erosive arthritis, triple carcinomas (oesophageal, gastric and lung cancer), peripheral neuropathy, colon cancer and adenomatous transformation has also been reported.^(3,14-17) Treatment includes nutritional support, antibiotics, corticosteroids and zinc supplement. The prognosis is poor, and remissions and recovery have been described in a few cases.

Kilcheski et al reported two cases of CCS and retrospectively reviewed 13 cases in the literature with regard to the radiographic abnormality on upper gastrointestinal barium series. They found gastric polyposis in all 13 patients, polyps and rugal thickening in seven, and polyps, rugal thickening and whiskering in three patients.⁽⁵⁾ Whiskering has been thought to occur due to the enlargement of the areae gastricae, with barium entrapment in the surrounding sulci gastricae. Maurer described a similar appearance of whiskering as spiculations.⁽⁶⁾

Dachman et al reported the radiological features observed in six cases of CCS and found that the stomach was diffusely diseased in five cases and focally, in one case. Small bowel polyps were observed in four cases. The patterns of involvement were classified as innumerable small polyps carpeting large areas, scattered varying size polyps and sparse involvement with few polyps. Concordance of pattern was observed between the stomach and colon in five of the six patients.⁽⁷⁾

In our case, we also found all the features described

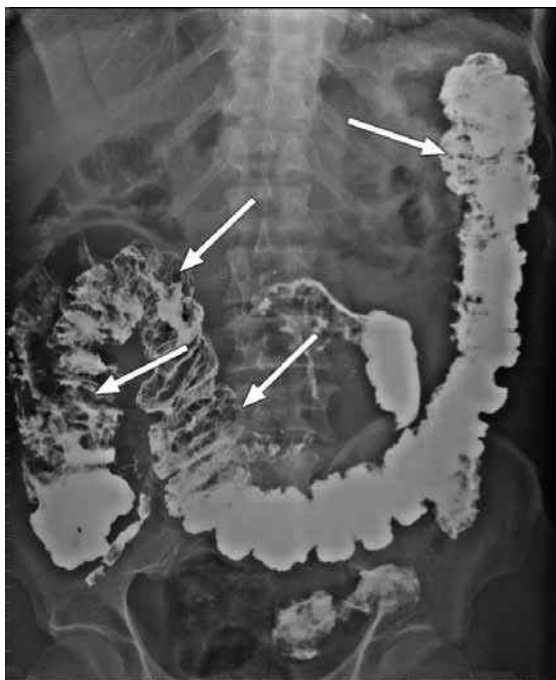


Fig. 8 Delayed radiograph of the colon with pneumocolon shows multiple polyps (arrows) scattered throughout the colon.

above, i.e. diffuse gastrointestinal polyposis of varying sizes, whiskering and rugal thickening. In addition, mosaic pattern, crypts and ruptured/ulcerated polyps in the stomach were noted in our case, which are new findings and have not previously been reported. The mosaic pattern was probably due to the thickened areae gastricae seen on double contrast film and is similar to the whiskering seen in the profile. Ulceration was likely due to the rupture of polyps caused by cystically dilated glands. To delineate all these lesions, a single test of barium meal follow through with pneumocolon was performed. This is a new, modified technique that was developed by Nijhawan et al.⁽⁸⁾ In this technique, air is insufflated per rectally in order to adequately distend the barium-filled colon, after the barium column of follow through examination has reached up to the descending colon and the distal ileum has emptied. This provides a good view of the colon and obviates the need for colonoscopy in many patients. It simultaneously detects lesions such as strictures, ulcers and polyps in the small and large intestines. In addition, the unsuspected tumours, polyps and ulcers in the large bowel may be detected even when the small bowel is normal.

In summary, we report a rare case of CCS and a detailed review of the literature, with particular reference to its radiological features. Additional radiological findings of mosaic pattern in the stomach and ulcerated polyps were detected, which have not been previously described. In our view, a typical radiological appearance

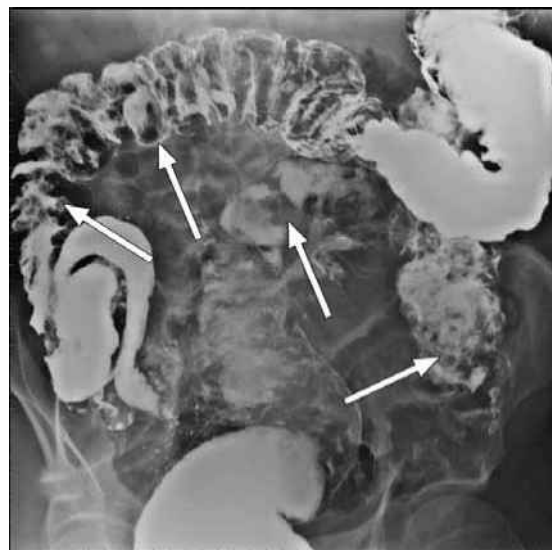


Fig. 9 Delayed radiograph of the colon with pneumocolon shows multiple polyps (arrows) throughout the colon and rectosigmoid.

of CCS on barium meal follow through with pneumocolon used as a single test, in conjunction with the characteristic clinical features, is sufficient to make a diagnosis of CCS. Endoscopy and biopsy may be undertaken for further characterisation of polyps and to rule out malignancy.

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