Cronkhite-Canada Syndrome - Case Report

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Abstract
Cronkhite-Canada syndrome (CCS) is a rare non-inherited syndrome characterized by diffuse polyposis of gastrointestinal tract, diarrhea, weight loss, abdominal pain, cutaneous hyperpigmentation, dystrophic changes of fingernails and alopecia.

We present a rare case of gastrointestinal polyposis where a 55 year male patient presented with abdominal pain, vomiting, Hematochezia, cutaneous hyperpigmentation, swelling of legs, endoscopy showing multiple sessile polyps in stomach and duodenum and colonoscopy showing multiple sessile polyps throughout colon. Histological study revealed inflammatory polyps. Based on clinical features, endoscopic and histopathologic findings a diagnosis of CCS was correctly made.

Keywords: Cronkhite-Canada syndrome (CCS), Onychodystrophy, Hematochezia hyperpigmentation, Steatorrhoea

Introduction
Cronkhite-Canada syndrome is a rare non inherited gastrointestinal polyposis syndrome with distinct cutaneous findings like hyperpigmentation and onychodystrophy. The etiology is probably autoimmune and diagnosis is based on history, physical examination, endoscopic findings of Gastrointestinal polypsis and histology. CCS was reported for the first time in 1955 by Leonard W. Cronkhite, jr. and Wilma J. Canada as a new distinct clinical entity in two female patients with generalized gastrointestinal polyps, cutaneous pigmentation, alopecia and onychodystrophy [1]. The disease is very rare, until 1999 over 500 cases have been reported worldwide [2]. The course may be rapidly progressive with a grave prognosis. Here we report a case of Cronkhite-Canada syndrome.

Case Report
55 year male presented to hospital with two months history of abdominal pain, hiccup, associated with vomiting, Hematochezia and occasional loose motion 4 times/day. These symptoms were not associated with Hematemesis or Melena. There was progressive hyperpigmentation of skin all over the body, which was more marked over hands (Figure 1). He also complained of swelling of feet during last two months, with occasional steatorrhoea. Patient had no history of fever, anorexia, jaundice. Patient was neither an alcoholic nor had tuberculosis, diabetes and inflammatory bowel disease, SLE, RA, Hypothyroidism. On examination patient was edematous and moderate pallor was present. Patient had diffuse hyperpigmentation of skin all over body, more marked

Figure 1: Hyperpigmentation of hands
over hands without jaundice, lymphadenopathy, alopecia or onychodystrophy.

On investigation: Patient had normal CBC, except patient’s Hb% was 8 gm%. Serum protein 4 gm%, Albumin 1.6 gm%, but other liver function tests were normal. Serum IgG4 was 500mg % (Normal 40-140mg%). Fasting blood sugar was 72mg%, Serum Na⁺ was 134meq/L, Serum K⁺ 4.8meq/L, Serum Creatinine 0.8 mg% and Serum cortisol (8 am) was 215.8nmol/L. His urine and stool examinations were normal with no evidence of blood in stool. Upper gastrointestinal endoscopy showed multiple sessile polyps of varying sizes (1-3 cm) present in stomach and duodenum (Figures 2 and 3). Gastric sample for RUT (Rapid Urease Test) was negative for Hpylori infection.

Colonoscopy showed multiple sessile polyps of sizes 1-5 cm throughout the colon (Figure 4).

Histopathology study of polyps showed inflammatory polyps with metaplastic changes but p55 immunoreactivity accumulation in the tissue was found to be negative. Ultrasonography of abdomen showed mild fatty change in liver with cholelithiasis.

Patient was treated with prednisolone 40 mg once daily, symptomatically improved within 10 days but subsequently lost to follow up.

Discussion

Despite being first described over 50 yrs ago, Cronkhite-Canada syndrome (CCS) has an obscure etiopathogenesis. Given increased Ig G₄ mononuclear cell staining in CCS polyps, an autoimmune mechanism may be involved [1]. CCS can develop in all ethnic groups and onset occurs at mean age of 60yrs. Abdominal pain, diarrhoea, dysgeusia are the most common initial symptoms, with dermatologic symptoms of hyperpigmentation, alopecia and onychodystrophy often occurring later [1]. Polyps in CCS patients can develop throughout GI tract (except in esophagus) and are non-neoplastic, mostly inflammatory [3,4].

Our diagnosis CCS was based on features of malabsorption in the setting of characteristic clinical, endoscopic and histological findings [5]. Although CCS often had characteristic features, the differential diagnosis includes a number of polyposis syndromes including FAP, Peutz-jegher’s syndrome, cowden disease and juvenile polypsis. Usually it is not difficult to distinguish CCS from these polyposis syndromes as each exhibits its own characteristic clinico pathologic features [6]. However the endoscopic and histologic features of CCS polyps and juvenile polyps overlap and may appear identical. A useful distinction between the two polyps is that the mucosa among CCS polyps is histologically abnormal revealing edema, congestion and inflammation of lamina propria [inflammatory polyp] as in our case, in contrast the mucosa among juvenile polyps is normal [7]. In addition IgG₄ plasma cell infiltration occurs in CCS polyps but not in juvenile polyps. The question of whether polyps in CCS patients possess malignant potential is controversial. One possibility is that the chronic generalized mucosal inflammation in CCS may increase neoplastic transformation similar to inflammation induced mutagenesis of idiopathic IBD, as seen in the largest single centre case series conducted in CCS patients to date, the incidence of colorectal neoplasia within the follow up period was high (Adenoma71%, cancer 14%) [3].

Given the rarity of CCS, there is no evidence based therapy. Numerous treatments have been attempted in CCS patients with varying degree of success. These treatments include corticosteroid, proton pump inhibitors, antibiotics, hyperalimentation, anabolic steroids, surgery, cromolyn sodium and combination of these [8].

Corticosteroid is the main stay of medical treatment for CCS. The steroid regimen is prednisolone 40 mg daily for 1 week with a 5 mg decrease every week until the patient is tapered off. In one study symptomatic response was seen within 3 months in 10 of 11 CCS patients treated with these regimens [3].

Our patient received prednisolone 40 mg once daily with PPI, within 10 days patient was symptomatically improved and discharged but subsequently he was lost to follow up.

Conclusion

We reported a patient diagnosed with rare gastrointestinal
polyposis syndrome of CCS based on combination of clinical (abdominal pain, diarrhea, Hematochezia and hyperpigmentation of skin), endoscopic (diffuse sessile polyps in stomach, duodenum, colon), histopathologic findings (inflammatory polyps with metaplasia). Immunosuppression with corticosteroid may lessen or eradicate manifestations of Cronkhite-Canada Syndrome.

References