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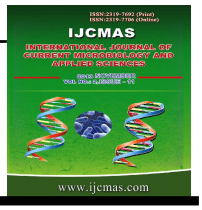

Original Research Article

Cronkhite - Canada syndrome, a rare noninherited gastrointestinal polyposis - a case report

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K e y w o r d s

GI polyposis; Cronkhite; Canada Syndrome.

A B S T R A C T

Cronkhite- Canada syndrome is one of the rare noninherited gastrointestinal polyposis causes intestinal and extraintestinal manifestations. Chronic diarrhoea is one of the major intestinal symptoms.

Introduction

Case report

A 48 year old male presented to medicine OPD on 21/1/2012 with history of passing loose stools since one and a half years duration which has increased in past one month and was associated with diffuse dull aching abdominal pain. Since one month he complains of passing 3-4 episodes of loose stools after 2-3 hours of each meal which was watery in consistency and not associated with blood or mucus in it, so he had to decrease his regular food intake. Patient also started noticing splitting of nails in both upper and lower limbs since one and a half years. On examination, patient was poorly built and moderately nourished. Alopecia, dystrophic nail changes of fingers and toes, hyperpigmentation of palms and soles were observed, oral cavity was normal except for loss of papillae and hyperpigmentation of lateral wall of buccal cavity (Fig 1,2,3) . No edema or lymphadenopthy were present. Systemic examination revealed no significant abnormality. Laboratory investigation was of normal range except for stool for occult blood was positive (table 1). Upper GI scopy showed more than 100 small polyps in stomach (fig.4), this raised our suspicion on the polyposis syndrome in patient and subjected to colonoscopy which revealed Multiple, Minute Polyps Scattered from Rectum till Caecum up to Ileocaecal valve(fig 5). Biopsy from polyps taken was suggestive of Hamartomatous Polyposis. Karyotyping

Table.1 Laboratory examinations

Haemoglobin	14.8 gm%
Total WBC Count	12900 cells/cumm
Eosinophils	5%
ESR	5mm
Total Protein	5.8 g/dl
Albumin	3.0 g/dl

Stool Examination - Occult blood present

Table.2 Laboratory examinations

USG – Thyroid	Well defined heterogeneous nodule in left lower part of left lobe measuring 7x5 mm – suggestive of Adenoumatous Nodule.
Upper GI Scopy	More than 100 small polyps seen in stomach.
Colonoscopy	Multiple, Minute Polyps Scattered from Rectum till Caecum up to Ileocaecal valve.
Histopathological Examination	Suggestive of Hamartomatous Polyposis.
Karyotyping	Normal Male Chromosomal pattern.

Figure.1 Lentigens and Alopecia

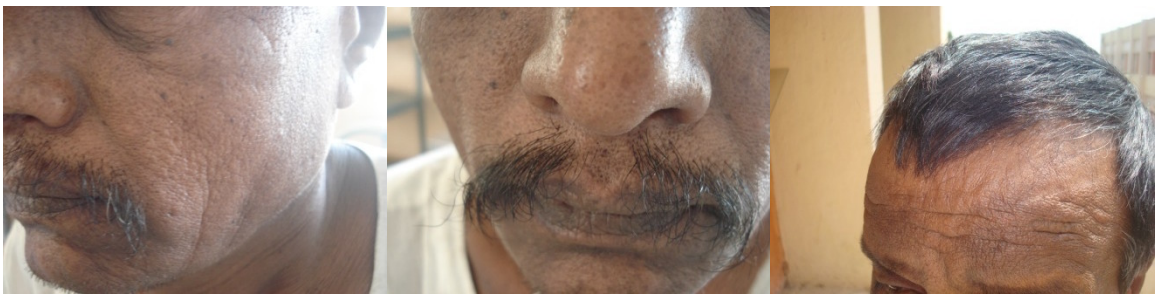


Figure.2 Lusterless Hair and Hyper pigmentation of Hands and Soles

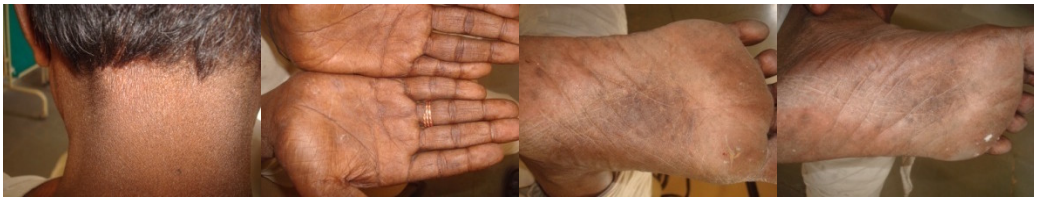


Figure.3 Dystrophic Nail Changes

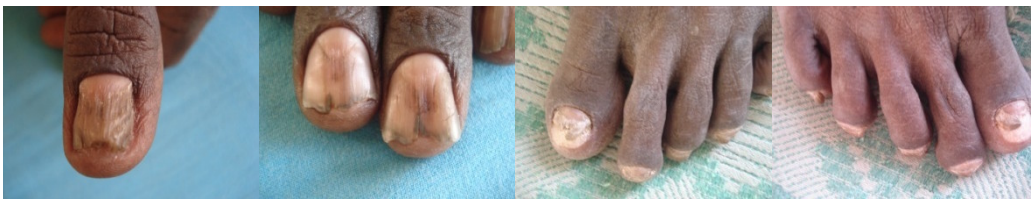
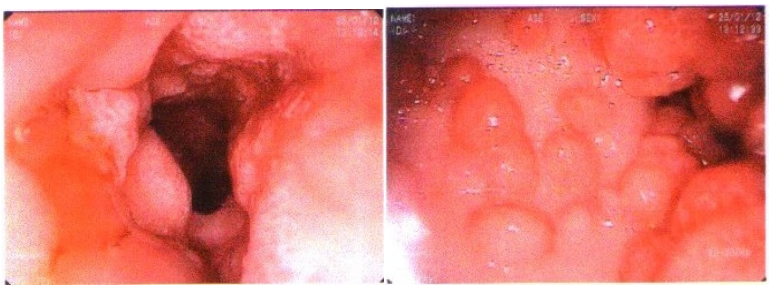


Figure.4 Upper GI Scopy – Stomach



Multiple, Small and large sessile polyps seen in stomach

Duodenum - Showing small polyps

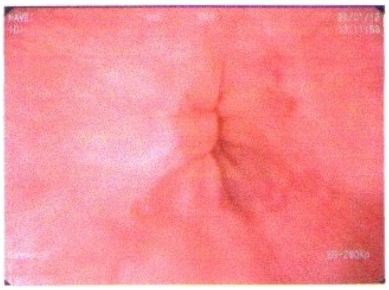
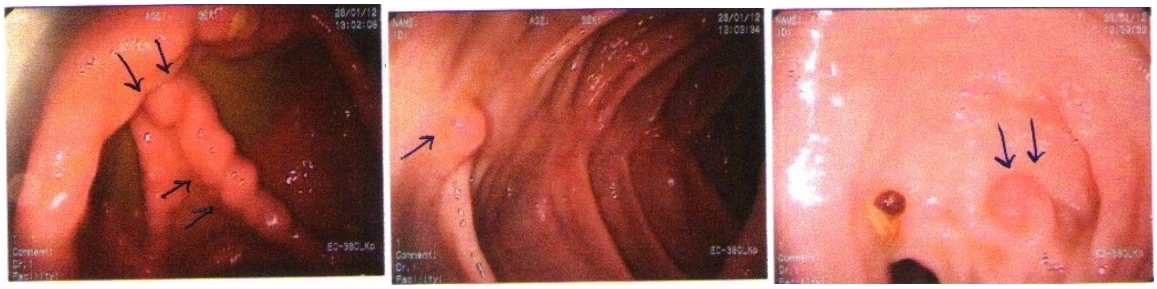
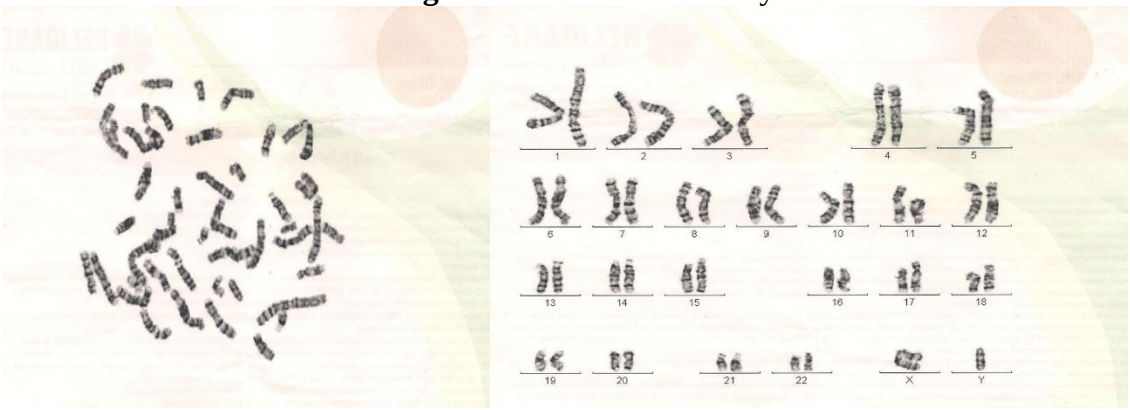


Figure.5 colonoscopy



Multiple, Minute polyps scattered from rectum till caecum

Figure.6 Chromosomal analysis



Normal male kyotype

was done to rule out genetic abnormality which showed normal male chromosomal pattern. His 18 year old son was evaluated clinically and found to have lentegens over the lips, so subjected to upper GI scopy to rule out familial inheritance which was normal. Hence, with detailed clinical history and clinical features and special investigations, we diagnosed the patient to have Cronkhite-Canada Syndrome. Initiated him on a high protein diet, proton pump inhibitors and empirical antibiotic therapy. Patient showed significant improvement in his abdominal complaints. Patient is being regularly followed-up for malignant transformation.

After six months patient was again subjected to endoscopy for malignant changes but revealed no significant changes from previous endoscopy and biopsy showed negative for malignancy .patient was symptomatically better.

Diarrhea is passage of abnormally liquid or unformed stool at an increased frequency, said to be chronic if persistent for more than 4 weeks in duration. Chronic diarrhea warrants evaluation to exclude underlying pathology, as most causes are non-infectious. In estimated two-third of the cases remains unclear after initial encounter and further testing is required. Stool collection and analysis can yield important objective data, Upper Endoscopy and Colonoscopy with biopsies to rule out structural or occult inflammatory diseases'1.

Cronkhite-Canada Syndrome (CCS) is a rare non-familial polyposis syndrome presenting as chronic diarrhea. CCS is characterised by epidermal manifestations like alopecia, dystrophic nail changes and hyperpigmentation. GI changes include generalized hamartomatous polyposis'2 .

The mucosal proliferation leads to fluid and electrolyte abnormalities, malnutrition, malabsorption and GI bleeding. These changes lead to clinical symptoms like diarrhea, abdominal pain and malnutrition. Diarrhea is multifactorial,the beneficial effects of antibiotics are attributed to small bowel overgrowth.'3 Steroids are most likely effective as anti-inflammatory agents'4. Polyps are believed to contribute to diarrhea'2. Data from most cases support the belief that ectodermal features are the result of malnutrition but many symptoms and signs appear or remit in a manner inconsistent with this theory'3. The aetiological factors leading to progression, spontaneous remission, or optimal treatment has not been established. Current understanding of the disease is based on small series of cases reported since it was first described by Cronkhite and Canada in 1955'2.

The prevalence of GI malignancy is about 10%. Most of the complications encountered are manifestations of polyposis. Some patients with CCS have been diagnosed after presenting initially with eosinophilic gasteroenteritis and intestinal candidiasis.

Chronic diarrhea associated with malena has to be evaluated thoroughly with upper GI scopy and Colonoscopy to rule out or confirm rare causes, such as Cronkhite-Canada syndrome for better management.

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