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Cronkhite Canada Syndrome: A Moroccan Case

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ABSTRACT

Cronkhite Canada Syndrome (CCS) is a rare, non-hereditary disease that combines chronic diarrhea, diffuse intestinal polyposis and onychodystrophy. We are reporting on the observation of the first case of Cronkhite Canada Syndrome in Morocco. The case has to do with a 33-year-old patient with chronic bloody diarrhea associated with alopecia and onychodystrophy within a context of general deterioration and significant weight loss. The biological assessments were characterized by the presence of microcytic anemia with hypo albuminemia, malabsorption syndrome and hydro electrolyte disorders. The colonoscopy had shown several hamartomatous polyps located at the recto sigmoid level. The histopathological examination showed hyperplastic inflammatory polyps with extensive propria oedematous lamina, and cystic glands. The diagnosis of CCS was made in the face of a host of clinical, biological, endoscopic and histological arguments. Our patient received oral corticosteroids as treatment, along with nutritional support and a treatment of electrolyte disorders. Cronkhite Canada syndrome is associated with high mortality and a risk of malignant transformation. Clinicians should considerand think about CCS for a patient with unexplained chronic diarrhea and dermatological manifestations (Alopecia, Onychodystrophy, Hyperpigmentation).

Introduction

First described in 1955 by Cronkhite and Canada, this syndrome includes the association of non-hereditary digestive polyposis, skin hyperpigmentation, onychodystrophy and alopecia [1].

It is a rare disease with about 500 cases that have been reported in the literature, that it is more likely to affect males in their fifties. CCS has a potential for cancer that affects 9 to 15% of cases reported in the literature [5,8]. Despite various medical and surgical treatments, no optimal treatment is yet known.

In this reported case, we describe the first case of CCS reported in Morocco with a literature review.

Clinical case

Mr. K.A. 33 years old, with no significant record of CCS in their family history, was hospitalized at the B Medical Clinic at Ibn Sina Hospital, in Rabat, in April 2017, for the treatment of chronic bloody diarrhea. The patient has been followed by a pediatrician ,since his childhood for a failure to thrive, with a normal etiological assessment.

In March 2006, at the age of 22, the patient was taken to the emergency room with severe colitis bleeding, requiring subtotal colectomy with ileoanal anastomosis. The anatomopathological examination of the surgical part showed, on the one hand, an aspect of acute ulcerative colitis with the presence of cryptococcosis parasitic agents, and, on the other hand, 3 hamartomatous polyps from Peutz-Jeghers. The progress was good until January 2013, at this time the patient was operated on at the emergency room for graft occlusion on flanges, a hail resection with terminal ileo-rectal lateral anastomosis was performed, the same histological aspect was found, namely Peutz-Jeghers polyps and intestinal cryptococcosis.

In April 2017, the patient was received by our department due to a series of bloody and mucus diarrhea (more than 9 stools per day). The clinical examination found the patient in bad general condition, pale, and dehydrated. Phanareal-cutaneo examination revealed alopecia and onychomycosis lesions. Biological examination revealed microcytic hypochrome anemia at 5.4g/dl, hypo albuminemia at 19 g/l, elevated CRP at 88.5 mg/l, hyponatremia at 117 mmol/l, hyperkalemia at 5.6 mmol/l. Thyroid tests, cortisol levels, as well as stool parasitology came back negative.

Gastroduodenal fibroscopy was normal. The colonoscopy showed the presence of several hamartomatous polyps of variable size and shape, bleeding easily at the contact with the rectum and the ileo-rectal anastomosis. The histological study had shown inflammatory and hyperplasic polyps.

With the presence of a combination of intestinal polyposis, onychomycosis, alopecia, alteration of the general condition of the patient, with endoscopic and anatomopathological data; the diagnosis of CCS was retained. The patient received nutritional support and treatments containing vitamins, combined with full-dose corticosteroid, antibiotics with anti-protozoals, pro-biotics and aminosalicyles. An improvement was noted. However, 7 days later the patient died in a severe sepsis.

Discussion

Cronkhite Canada Syndrome is a rare, unhealed disease without a family form, whose origin remains unclear, even if a possible autoimmune origin is mentioned.

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First described by Leonard Cronkhite and Wilma Canada in two patients[4]. Since then, more than 500 cases have been reported worldwide with a predominance among the Asian population, more likely to affect males in their fifties, triggering factors such as stress and fatigue have also been identified. The pathogenesis of CCS is still unclear, however in published cases, it may be associated with systemic lupus erythematosus, vitiligo, rheumatoid arthritis and hypothyroidism, suggesting an autoimmune factor [1,8].

Clinically, CCS is characterized by digestive and skin symptoms. It most often associates watery, hemorrhagic diarrhea, linked with abdominal pain, weight loss, anorexia, and asthenia. These disorders are secondary to malabsorption syndrome and exudative enteropathy, which are themselves secondary to changes in the digestive mucosa. Dermatological disorders are not specific to CCS, they are marked by order of frequency, onychodystrophy, alopecia, and hyperpigmentation. The etiology of these disorders seems to be deficient, and there is remission under protein and/or vitamin supplementation.

In biology, deficiencies are most often found. It is classic to encounter anemia, hydroelectrolytic disorders, and hypoprotidemia linked with hypo albuminemia.

Endoscopically, a diffuse digestive polyposis is found, characterized by juvenile, hamartomatous polyps affecting the entire digestive tract, except the oesophagus, and most often having an aspect of "strawberry" or "raspberry". The histological study shows an infiltration of inflammatory cells, as well as sub-mucosal edema in the lamina propria linked with cystic dilation of the mucous glands. The polyp-free mucosa is characterized by significant alteration of crypts and constant villositary atrophy.

The main differential diagnoses are, in the first place, digestive polyposes such as family adenomatous polyposis or Peutz-Jeghers syndrome, juvenile polyposis, Cowden's disease.

Treatment is not based on a standardized protocol. Indeed, at present, several treatments have been identified and used with various degrees of success. It is recognized that nutritional supplementation is a pillar of care, such as the corticosteroid treatment. The analysis of clinical-biological-endoscopic developments makes it possible to determine the effectiveness of treatments. Indeed, the use of corticosteroids seems to be the reason of the digestive disorders remission, both clinically and endoscopically. Skin disorders seem to be improved by nutritional protein and vitamin treatment, whether enteral or parenteral. The use of surgery and immunosuppressants seems to be reserved in case of failure of the above-mentioned treatments. Finally, other protocols were used, such as antibiotics and digestive anti-inflammatory drugs (sulfasalazine, salazosulfapyridine).

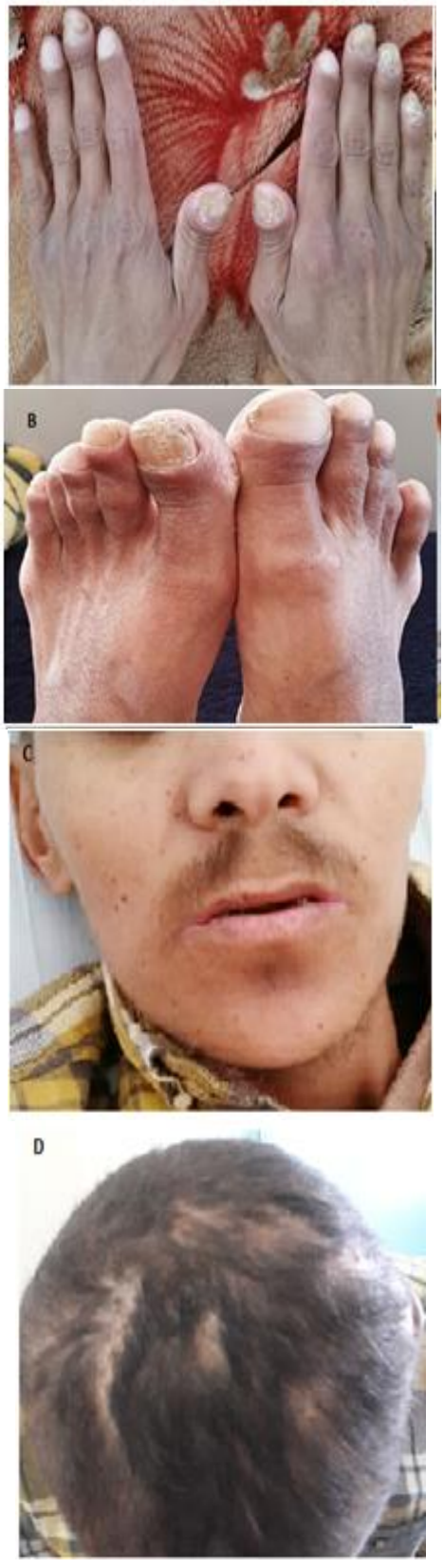
The prognosis is poor with an estimated mortality rate of 55% at 5 years. The development of the syndrome is characterized by many complications such as digestive hemorrhages that can be cataclysmic, malnutrition, infections and cancers. The frequency of neoplasia is more frequent in patients with CCS compared to the general population, and is estimated at about 18%. The etiology of this complication is still being debated today. Some hypothesize an adenome-adenocarcinoma sequence from the polyp, others suggest the possibility of de- novo cancer, and others assume an infectious origin.

In addition, it is important to provide regular endoscopic monitoring to remove any suspicious polyp and/or polyp larger than 1cm.

Conclusion

CCS is therefore a complex pathology, sometimes with diverse appearances, and whose diagnosis, which is not easy, requires clinical, biological, endoscopic and anatomopathological analysis. Delay in diagnosis is common. Nutritional support, antibiotics, corticosteroids, and surgery have all been used with varying degrees of success. Unfortunately, therapeutic trials were not possible due to the rarity of the syndrome.

Images



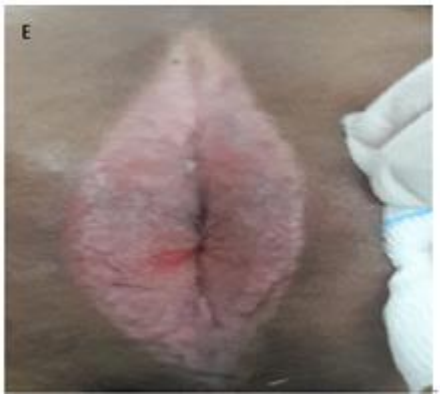


Fig1. physical signs of SCC: Onychodystrophy (A, B), Alopecia (D), hyperpigmentation (C), Anal mycosis (E)

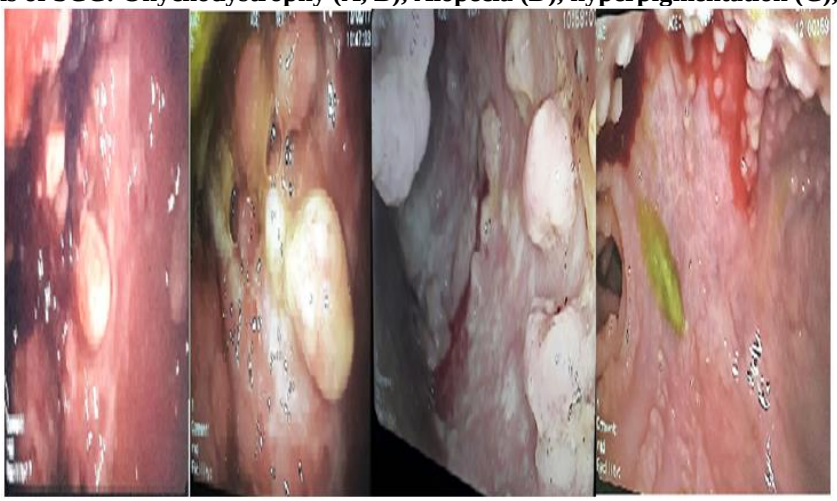


Fig.2. Endoscopic images showing several inflammatory polyps sitting at the rectosigmoid and ileum level

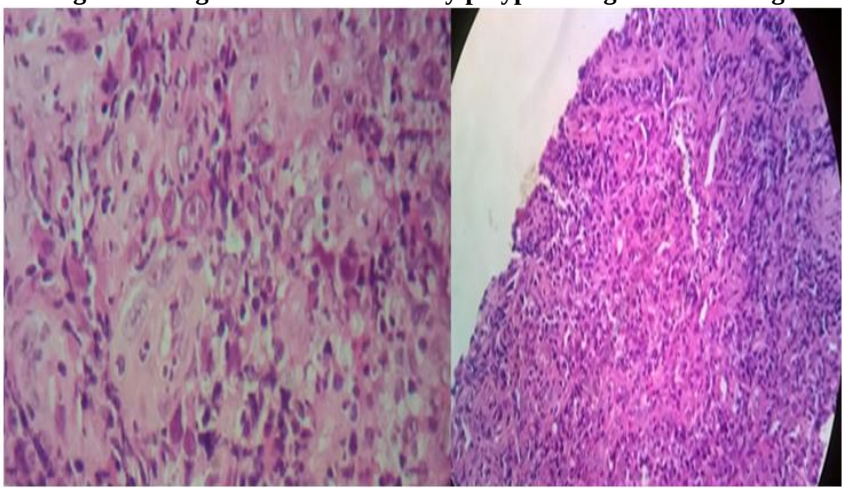


Fig.3.The anatomopathologic examination of recto-sigmoid biopsies showing hyperplastic inflammatory polyps

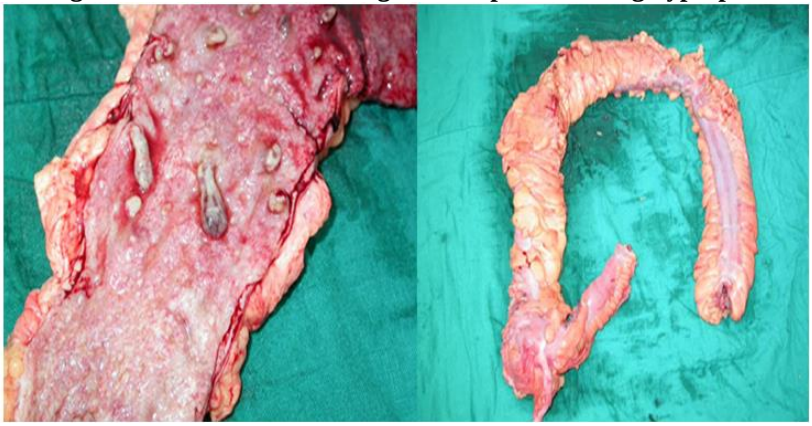


Fig.4. operative specimen of total subcutaneous colectomy; at the opening we note the presence of several polypoid formations along the colon which presents itself with a tubular aspect

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