



RADIOLOGIC PATHOLOGIC CORRELATION / *Gastrointestinal imaging*

## A rare gastric polyposis: Cronkhite-Canada syndrome

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### KEYWORDS

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### Case report

A 78-year-old man, without personal or family antecedents, consulted for acquired anorexia and profuse diarrhoea with eight to 10 bowel movements per day for the last 6 weeks, appearing suddenly without an epidemic or medicinal context and not relieved by symptomatic treatments. His general condition was preserved but the patient reported the loss of 6 kg. The clinical examination detected onychomadesis (detachment of the nails) (Fig. 1) and alopecia of the scalp. His abdomen was supple, without palpable mass or hepatosplenomegaly. The laboratory tests detected a hypoalbuminemia at 23 g/L and a macrocytosis at 102 fL. The blood count, the inflammatory (sedimentation rate, reactive protein C, electrophoresis of the plasma proteins and fibrinogen), immune and thyroid assessments were normal. The faecal culture and the parasitology examination of the stools proved to be negative. The gastroscopy found congestive antral gastritis with large very erythematous cerebriform folds, with an infiltrated appearance of the bulb and the duodenum (Fig. 2a and b). The colonoscopy revealed a great many raspberry-like, red non-ulcerated and non-hardened sessile polyps along the entire surface of the large intestine associated with a congestive mucosa without lesions suspected of malignancy (Fig. 2c and d).

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**Figure 1.** Onychodystrophy of the patient.

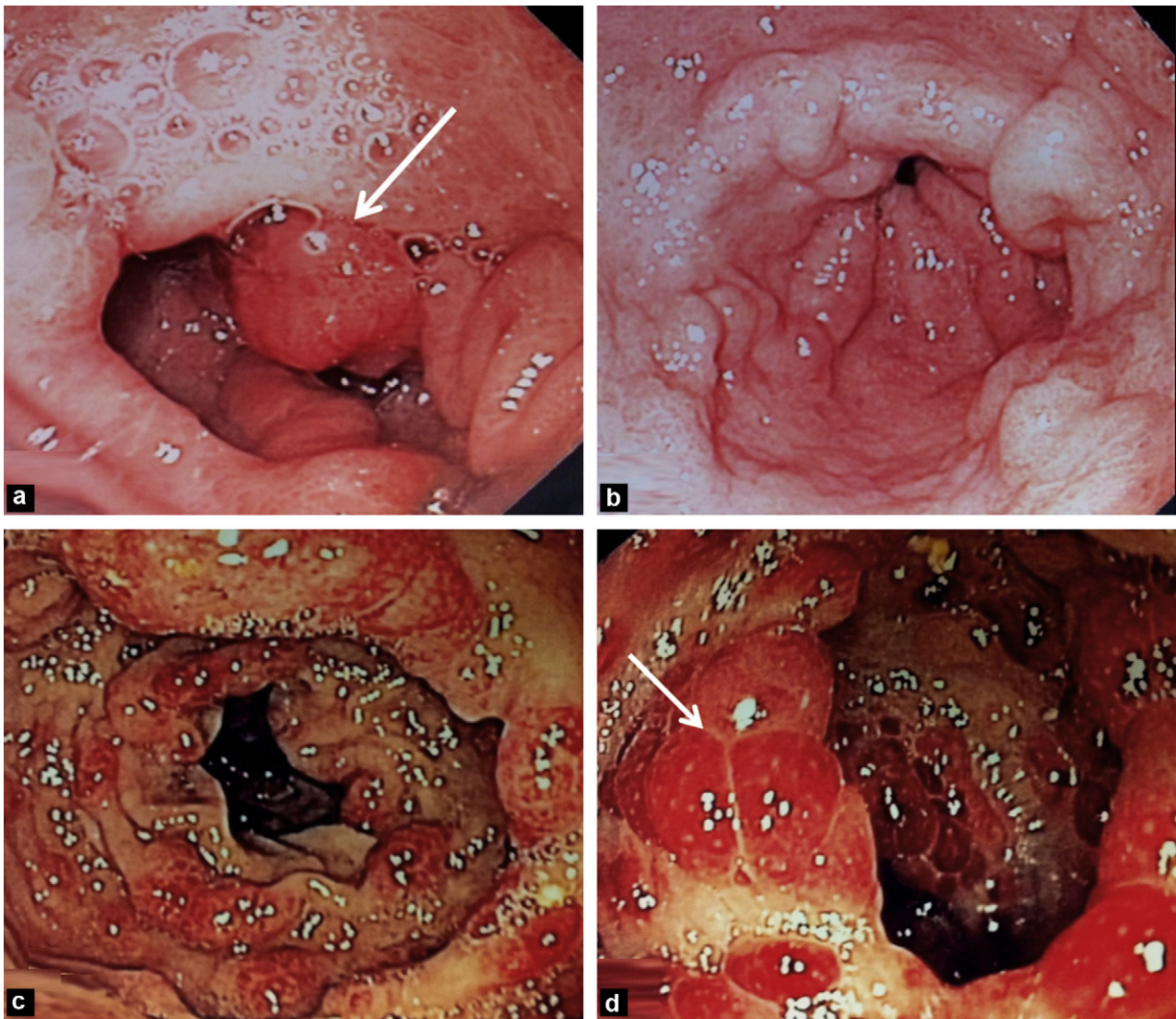
The gastric and sigmoid-colic biopsies reveal cystic and tortuous dilation of the glands of the gastric mucosa, coated with a well-differentiated and non-dysplastic single stratified epithelium (Fig. 3a–c). The glands are irregularly dispersed in an abundant, focally haemorrhagic chorion, consisting of a polymorphous inflammatory infiltrate of lymphoplasmocytary predominance (Fig. 3d). The patient also underwent a colon-MRI and CT enteroclysis that found multiple polyps along the entire gastric antrum and the large intestine, with more severe impairment of the sigmoid (Fig. 4a–d). A doubt remained as to the impairment of the small intestine by CT enteroclysis, although the endoscopic videocapsule carried out as a complement found a jejunum and ileum with a healthy mucosa, free of any polyps. The manifestations of the integuments (partial alopecia and onychotrophia), associated with gastric polyposis, support the diagnosis of Cronkhite-Canada syndrome raised during the anatomopathology examination. An initial treatment by intestinal anti-inflammatory consisting of 5-aminosalicylic acid derivatives (5-ASA) was found to be ineffective. A trial treatment with corticoids and nutritional measures was initiated and improved the clinical symptomatology in two

months. The patient was informed of the possibility of a colectomy in case of clinical aggravation or inefficacy of the drug treatments.

## Discussion

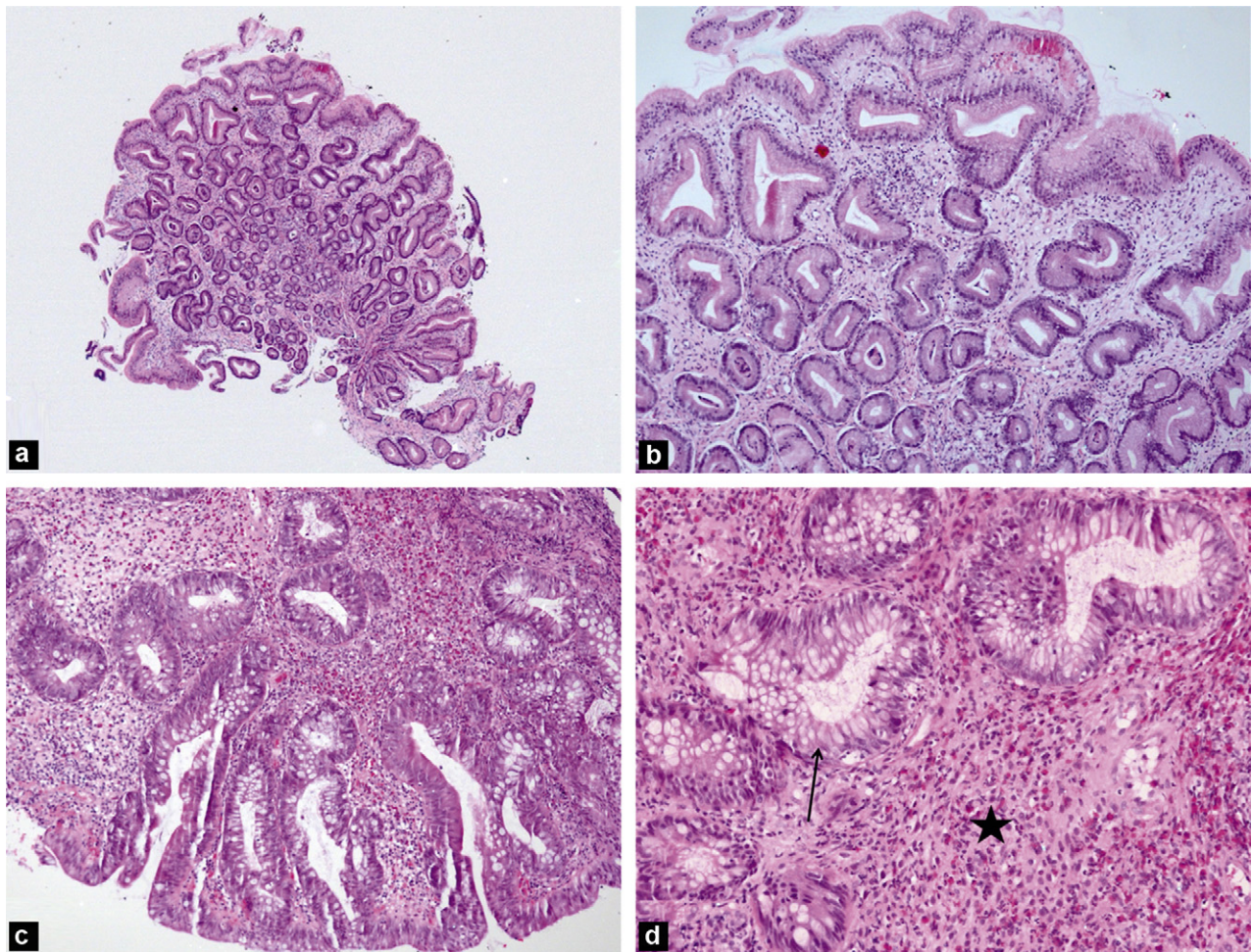
Cronkhite-Canada syndrome is a rare disorder of unknown pathogenesis. It was described in 1955 by Leonard Wolsey Cronkhite Jr, an internist and Wilma Jeanne Canada, a radiologist [1]. This syndrome associates non-hereditary gastrointestinal polyposis, onychotrophia, alopecia and skin hyperpigmentation. The incidence is estimated at 1/1,000,000 with 400 cases described in the world, 75% of which are in Japan [2]. The mean age of the diagnosis is 59 years [3]. The clinical picture is dominated by abundant diarrhoea, protein-losing enteropathy, frequently responsible for severe undernutrition and hydroelectric disorders that condition the prognosis of this disease. The 5-year rate of survival is 55%.

Polyposis is characterised by sessile, red, raspberry-like polyps of variable size that affect the entire digestive tube except for the oesophagus. Initially described as adenomateous polyps [1], in reality the lesions correspond to hamartomateous juvenile polyps. The histological examination shows a preserved surface epithelium, cystic proliferation and dilation of the glands of the mucous membrane enclosing mucus. The chorion is inflammatory, abundant and encloses a polymorphous cell infiltrate. Cases of cancerous degeneration have been reported although the origin is not certain since, classically, juvenile polyps do not degenerate [4,5]. The hypothesis of cancerisation of co-existent adenomateous polyps is most likely. Therefore, regular annual endoscopic monitoring is recommended [3]. Juvenile polyposis and the Peutz-Jeghers syndrome are the main differential diagnoses [6]. Given the rarity of this disorder, the treatment has not been clearly established. Dietetic measures and corticoid treatment are usually proposed. The duration varies according to the clinical response [3]. Surgical treatment by subtotal gastrectomy or colectomy is reserved for the complicated forms (gastric occlusion, cancer) or forms resistant to medical treatment due to its high morbidity.

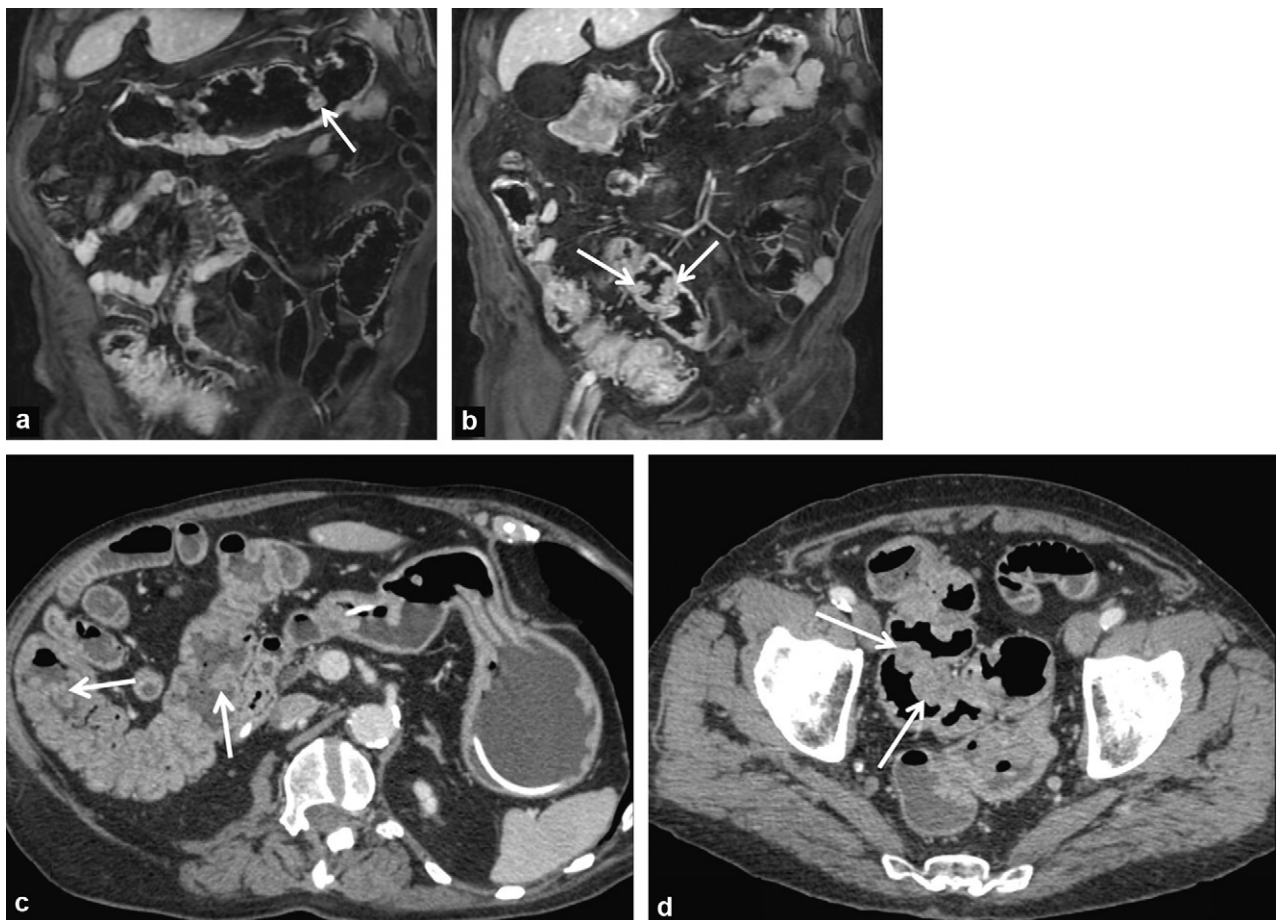


**Figure 2.** a: gastroscopy: polyp of the gastric antrum (white arrow); b: gastroscopy: voluminous cerebriform inflammatory folds of the gastric antrum; c: colonoscopy: multiples sessile polyps of the sigmoid colon; d: colonoscopy: raspberry-like, hamartomatous polyp of the sigmoid (arrow).





**Figure 3.** a: histological examination of a polyp of the gastric antrum: dilated and tortuous appearance of the glands of the gastric mucosa (HES  $\times 2.5$ ); b: detail of the glandular epithelium of the gastric antrum: ornate appearance without dysplasia (HES  $\times 10$ ); c: histological examination of a polyp of the sigmoid colon: cystic dilation of the glands and crypts of the colon mucosa (HES  $\times 10$ ); d: sigmoid colon: tortuous glands edged by a non-dysplastic epithelium (black arrow) dispersed in an abundant and inflammatory polymorphous chorion (black star) (HES  $\times 20$ ).



**Figure 4.** a: colon-MRI: polyp of the transverse colon (arrow); b: colon-MRI: sessile polyps of the sigmoid colon (arrows); c: CT enteroclysis: polypoid thickening of the right colic angle (arrows); d: CT enteroclysis: multiple sessile polyps of the sigmoid colon (arrows).

## Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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