The Cronkhite - Canada Syndrome: a Seldom Recognized Entity

The Cronkhite - Canada syndrome is a rare, benign, non-neoplastic, non-hereditary condition which consists of gastrointestinal polyps associated with alopecia, onychodystrophy, and hyperpigmentation of the skin. It usually presents with severe diarrhoea, weight loss, and other malabsorption symptoms. The age range is 31–86 years, with a mean age of onset of 60 years. Multiple polypoid lesions are found throughout the gastrointestinal tract, with the exception of the oesophagus (1,2), presenting a radiographic appearance of rounded filling defects. Microscopically, the polyps have a characteristic cystic appearance similar to that seen in juvenile polyps.

The hair loss (usually extensive) and nail changes (20% loss, with dystrophy in the remainder) are an intrinsic part of the condition, and are not secondary to malabsorption (1,2), but can precede or occur simultaneously with, or later than, the gastrointestinal symptoms. Hyperpigmentation of the skin most frequently occurs on the head, neck, arms and trunk, and on microscopy consists of accumulated melanin pigment without increased melanocytes (2). Malignancy has been seen in up to 16% of the 36 reported cases (3). In these cases, most polyps showed adenomatous areas with variable dysplasia (4). The diarrhoea may resolve spontaneously, or as a result of symptomatic or antibiotic treatment, or resection of the polyp-bearing intestine (1,2,3).

The aetiology of the condition is unknown. Various suggestions have been put forward, including an infectious cause (3), a lack of growth factors, or a failure to respond to them (2). The syndrome is associated with a poor prognosis (3), due to the effects of malabsorption and generalized debility. It is usually managed by intravenous nutrition, total parenteral nutrition, anabolic hormones, and antibiotics (3). In the 36 reported cases, only six survived for more than two years after the diagnosis (3), cachexia and infections being the most common causes of death (2).

The authors have seen two unreported cases (Figures 1–4). The first presented with a twelve-week history of diarrhoea, intermittent blood loss, and weight loss. A carcinoma was seen in the sigmoid colon, and this was incompletely resected initially. Following this, the patient experienced progressive total hair and nail loss. The diagnosis of Cronkhite - Canada syndrome was made during the post-mortem examination, when full clinical details were available, and the full features of the syndrome were observed. The cause of death was a massive pulmonary embolus following surgery for removal of the residual sigmoid carcinoma. The second case presented with the typical features of the syndrome, including skin pigmentation, hair loss, and onychodystrophy, as well as severe diarrhoea and weight loss over twelve months. The diagnosis was made during hospital admission. The patient died of severe bronchopneumonia with associated septicemia, after a prolonged illness with frequent infections.

![Figure 1:](image1.png) Onychodystrophy in the toes in Cronkhite - Canada syndrome. The toe nails show irregular transverse ridging in this case. Eighty percent of cases show onychodystrophy; the remaining 20% have total nail loss.

![Figure 2:](image2.png) Stomach, pylorus, and duodenum in Cronkhite - Canada syndrome. The mucosa is greatly thickened, and shows multiple polypoid nodules.
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