A 57-year-old man suffered from watery diarrhea, weight loss, and abdominal pain for 4 months. Noticeable signs of nail dystrophy (onychomadesis of all nails) were present (Fig. 1a) along with alopecia and cutaneous foci of hyperpigmentation (Fig. 1b). Upper-gastrointestinal endoscopy revealed a large number of strawberry-like polyps of different size in the stomach (Fig. 2).

Colonoscopy revealed polyposis of the whole colon, including the rectum. The majority of the polyps had a strawberry-like, adenomatous, and hyperplastic appearance (Fig. 3). Histologically, the majority of the polyps were juvenile-like with cystic dilatations of the glands and a benign mucinous epithelium. The glands were filled with a large amount of mucin (Fig. 4).

Some adenomatous polyps with low-grade dysplasia in the colon were also detected. A subsequent enteroscopy did not find polyps; however, edema and small indentation of the jejunum were present. Immunohistochemistry showed total alactasia and a strong positivity for tumor necrosis factor (TNF) in the macrophages and lymphocytes. Based on both the clinical and endoscopic picture, a diagnosis of Cronkhite-Canada syndrome was established.

Despite complex treatment, the clinical course was unfavourable. Since the clinical state of the patient did not improve, anti-TNF-α treatment was considered. TNF-α activity was examined in the small-intestinal mucosa, and the results showed a strong intracellular expression of TNF-α. Unfortunately, an experimental anti-TNF-α treatment could not be introduced, because of the rapid progression of the disease. The patient died 4 months after the diagnosis of Cronkhite-Canada syndrome was established.
Cronkhite-Canada syndrome is a rare non-hereditary polyposis syndrome of an unknown, possibly autoimmune etiology. The prognosis is poor and more than 50% of patients die within 2–4 years [1,2]. Cronkhite-Canada syndrome should be considered in all patients with gastrointestinal polyposis together with emergent alopecia and/or nail dystrophy [3].

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References

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Fig. 4 Histology. a Juvenile-like polyp with enlarged mucin glands. b Detailed view of mucinous glandular epithelium.

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