A fulminant course of Cronkhite-Canada syndrome

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. 1 a**) along with alopecia and cutaneous foci of hyperpigmentation (**Fig. 1 b**).

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.

Video 1

Endoscopy of the stomach.

Video 2

Endoscopy of the colon.

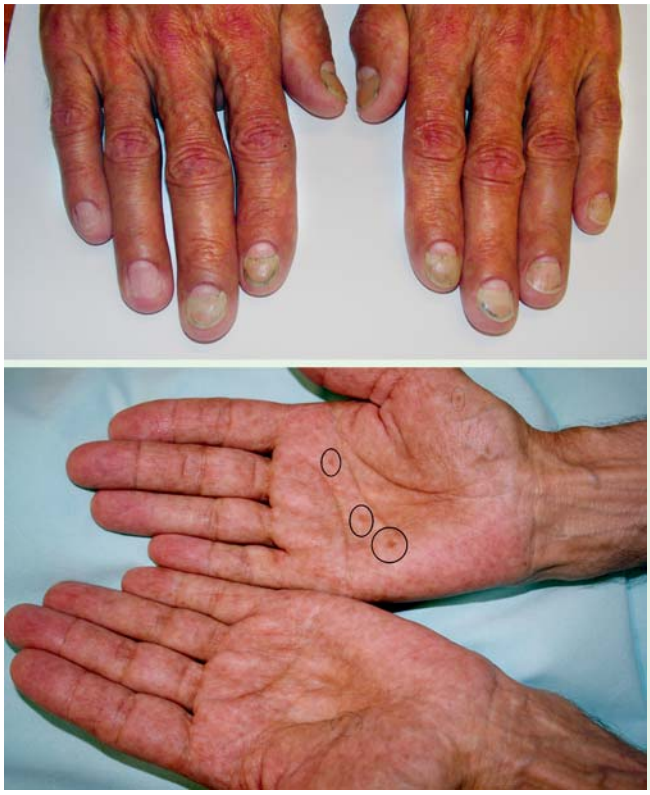

Fig. 1 Main physical findings. **a** All 20 nails with onychomadesis about 3 mm from the lunula. Several nail plates have been lost. **b** Hands with several small foci of skin hyperpigmentation.

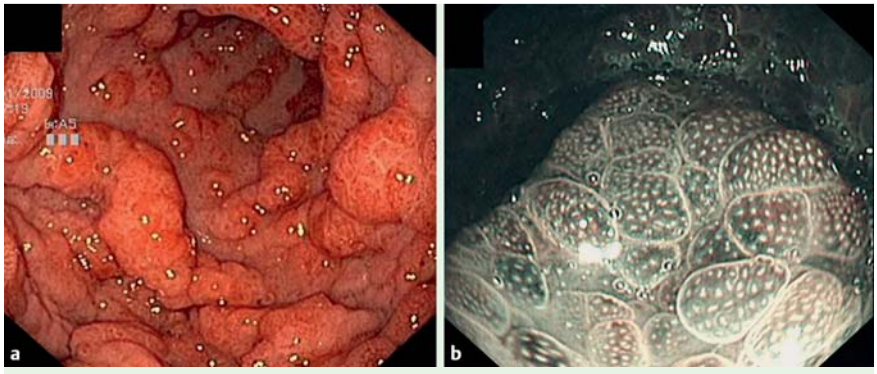

Fig. 2 Endoscopic findings in the stomach. **a** Polyps of gastric body and antrum. **b** Detailed view of gastric polyp using zoom and narrow-band imaging mode; note the strawberry-like appearance.

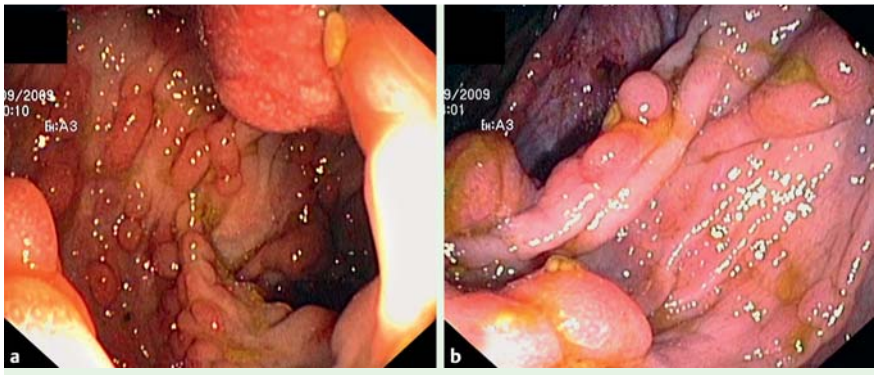

Fig. 3 **a, b** Endoscopic view of colonic polyps.

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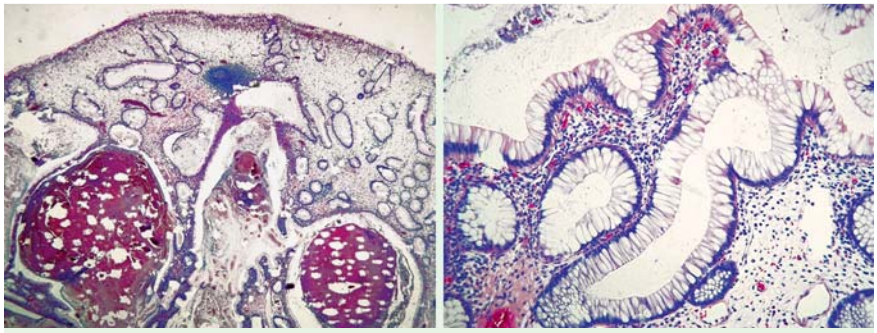

Fig. 4 Histology. **a** Juvenile-like polyp with enlarged mucin glands. **b** Detailed view of mucinous glandular epithelium.

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].

Competing interests: None

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Bibliography

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