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Strange cutaneous abnormalities and polyposis in an Asiatic man

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Polyposis

1. Case description

A 56-year-old-man, born in Laos, was hospitalized for weight loss to 9 kg in 3 months, asthenia, pruritus and a skin and nails changed color. He had no significant past medical history. He described a decreased appetite, bowel discomfort and diarrhea. Clinical examination discovered alopecia (Fig. 1A) with total loss of hair (pubis, armpits, etc.), melanodermia and severe onychopathy.

Blood test revealed microcytosis without anemia, no inflammatory syndrome, malnutrition syndrome (hypoproteinemia 50 g/l, hypokalemia 2.9 mmol/l) with a niacin and ascorbic acid deficiencies. There was neither steatorrhea nor creatorrhoea. CT scan was normal. Gastroscopy revealed a gastro-bulbar inflammation with edematous, thickened and hyperemic mucosa mimicking pseudo-polyps. A colonoscopy discovered a pseudo-polypoid proctitis, with mucosa edematous and hyperemic (Fig. 1B). Biopsies found elongated and hyperplasic crypts, edematous chorion, enlarged villosities, a pleiomorphic infiltrate chorion with lymphocytes, plasma cells, numerous polynuclear and eosinophilic cells, without *Trophyrema whipplei* (Fig. 1C). Immune staining for IgG4 was negative. A magnetic resonance enterography confirmed diffuse gastric, small bowel and colonic polyposis (Fig. 1D). Coeliac disease blood test, PCR *Trophyrema whipplei* on blood, saliva and stool, anti-gastric parietal cell antibodies were negative.

2. What's your diagnosis?

Cronkhite-Canada Syndrome (CCS)

3. Discussion

The association of melanodermia, alopecia, onychotrophia with a generalized gastrointestinal polyposis in an Asiatic patient conducts to the diagnosis of CCS.

CCS, first described in 1955 [1] is a rare protein-losing enteropathy, classically characterized by ectodermal changes and gastrointestinal polyposis respecting oesophagus. Five hundred cases were described in the literature, mostly in Japan [2] and China. Male-to-female ratio is 1.84, without any hereditary previous history, and median age is 63.5. Treatment is dominated by enteral nutrition, corrections of vitamins deficiencies and systemic corticotherapy. In refractory cases, azathioprine, ciclosporin and anti TNF-α antibodies were reported. Prognosis is poor with misdiagnosis and later on with intestinal malignancy.

Skin changes may proceed from vitaminic deficiencies like pellagra and scorbut.

A polymorph inflammatory infiltrate rich in eosinophils in chorion, sometimes IgG4 plasma cells in pseudo polyp identified [3] and a favorable response to immuno modulators are arguments for an immune dysfunction of CCS.

The patient was treated with a parenteral nutrition associated to vitamins B1, B3, B6, C for six weeks. Corticotherapy 0.8 mg/kg/day was introduced for four months associated to Azathioprine 2 mg/kg/day. The patient normalized his weight, nails and hair pigmentation at three months and normalized completely gastroscopic and colonic examination after one year of treatment.

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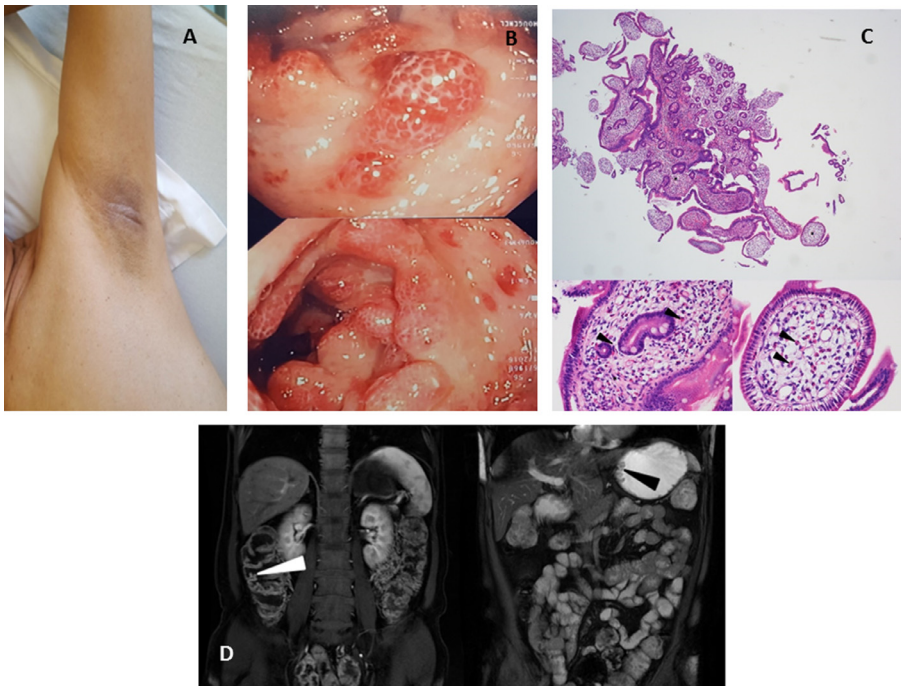

Fig. 1. A Total alopecia on the scalp B: Colonic inflammation with edematous, thickened and hyperemic mucosa mimicking pseudo-polyps on colonoscopy views C: Elongated and hyperplasic crypts, with an edematous chorion, enlarged villosities, a pleiomorphic infiltrate chorion with lymphocytes, plasma cells, numerous polynuclear and eosinophilic cells D: Diffuse gastric, small bowel and colonic polyposis.

Declaration of Competing Interest

The authors have no conflict of interest to declare.

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