Cronkhite-Canada syndrome

It is a rare serious disease. The etiology is unclear and it is thought to be an immune disorder. The disease was described in 1955 by internist L. W. Cronkhite and radiologist W. J. Canada. The disease occurs more in old age, and men are more often affected. Gastrointestinal polyposis is a characteristic feature, there are other gastrointestinal and skin symptoms, and colon and stomach tumors may appear later.

Clinical picture

The clinical picture in the disease is a typical combination of diarrhoea, weight loss, abdominal pain, skin hyperpigmentation, nail dystrophy and diffuse hair loss (hair, eyebrows, axillary hair, ..). Nausea, vomiting, dysgeusia, hypogeusia, xerostomia and atrophic language changes may also occur.

Despite the fact that only a few hundred cases have been described, the disease is divided into five groups according to the leading symptoms (Goto, 1995):

- **type 1** – the clinical picture is dominated by diarrhoea,
- **type 2** – the clinical picture is dominated by dysgeusia,
- **type 3** – the clinical picture is dominated by abnormal sensations in the mouth and thirst,
- **type 4** – the clinical picture is dominated by any abdominal manifestations with the exception of diarrhoea,
- **type 5** – the clinical picture is dominated by alopecia.

Serious complications can occur as the disease progresses. Gastrointestinal bleeding, intestinal intussusception and rectal prolapse are relatively common. In particular, anemia can follow, and infectious complications are no exception. Diffuse mucosal damage often leads to malnutrition. A number of other concomitant complications have been reported, but due to the rarity of the disease, it is not possible to determine whether it is an accidental coincidence.

Stomach and colon malignancies are a relatively common complications of the disease. Colon tumors do not arise on the basis of adenomas.

Pathology

nd swelling of the duodenal mucosa is also noticeable.

Histopathologically, the image corresponds to juvenile polyps or hamartomatous polyps. In addition, mild inflammatory infiltration with a more pronounced eosinophil count, marked submucosal edema, foveolar epithelial hyperplasia and cystic dilatation of the mucus glands are detected. Polyps often show adenomatous changes. Detection of conventional adenomas and serate lesions has also been described.

The epithelium shows damage even outside the polyps, the most significant changes are in the small intestine. These are mainly crypt architectural disorders, edema and mixed inflammatory infiltration, in which lymphocytes, plasma cells and eosinophils predominate.

Infiltrating plasma cells, even infiltrations outside polyps, show IgG4 staining for immunochemical staining. Therefore, some authors speculate that Cronkhite-Canada syndrome may be one of the manifestations of IgG4-associated disease.

Therapy and prognosis

Due to the unknown cause and pathogenesis, neither causal nor pathogenetic therapy is possible. The basis is symptomatic therapy, ie sufficient nutritional support, parenteral nutrition is preferred. Corticosteroids and antibiotics are given based on presumed immune disorders. Attempts to administer antihistamines and disodium cromoglycate have also been published, inspired by demonstrable eosinophil infiltration and, above all, the capture of degranulating eosinophils and mast cells in mucous membranes. Attempts to administer the more potent immunosuppressants azathioprine and tacrolimus have also been published. Other medications tested include the administration of proton pump inhibitors and the eradication of Helicobacter pylori, mesalazine and anti-TNFα.

The disease poses a significant risk to the development of malignancy of the stomach and colon, so regular endoscopic examinations are part of the disease management.

However, due to the rarity of the disease, the results were published for at most small series of patients. Because the disease can behave aggressively with rapid patient death, but spontaneous regression is not uncommon, the results are difficult to interpret. Therefore, there is currently no therapy that can be claimed to affect the course of the disease.

The prognosis of the disease is poor, five-year survival is less than 50%. On the other hand, spontaneous disease regression was observed in 5–10% of cases (regardless of the therapy chosen).

Links

Related articles in czech

- IgG4 asociovaná nemoc
- Polypy tlustého střeva

Literature

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- KOPÁČOVÁ, M., O. URBAN and J. CYRANY, et al. Cronkhite-Canada syndrome: review of the literature. *Gastroenterol Res Pract* [online] . 2013, vol. 2013, p., Also available from < https://www.hindawi.com/journals/grp/2013/856873/ >. ISSN 1687-6121.

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