Apoptotic colopathy as a manifestation of Good's syndrome

Authors


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44-year-old patient with 4 years of evolution of respiratory infections, fever, weight loss of 30 kg and chronic diarrhea with inconclusive colonoscopy studies managed as IBD (inflammatory bowel disease). History of thymectomy 4 years ago. On physical
examination with severe protein-calorie malnutrition, skin lesions compatible with herpes simplex infection and lower limb edema. Paraclinics with negative HIV, pancytopenia and hypoalbuminemia are requested; Chest tomography with budding tree pattern and bronchiectasis, but Sars-CoV2 negative. A colonoscopy was performed with the presence of ulcers in the sigmoid colon with an infectious aspect vs IBD, the biopsies revealed acute colitis with cryptic apoptosis without evidence of chronicity compatible with cytomegalivirus colitis. In the gastrointestinal panel, Ciclosporidium spp was documented and management with ganciclovir and nitazoxanide was started with initial improvement but persistence of clinical deterioration. For what is considered another type of immunodeficiency with the presence of hypogammaglobulinemia, reaching the diagnosis of Good's syndrome due to the association with thymoma and recurrent infections, treatment with Gamma globulin is started and later neurological deterioration is triggered, with a refractory supraconvulsive state and finally death.

Good's syndrome is defined as the presence of thymoma and acquired hypogammaglobulinemia with alteration in humoral and cellular immunity (1) predisposing to recurrent and opportunistic infections, as was the case in this patient. Chronic diarrhea is frequent in these patients secondary mainly to opportunistic infections that can be confused with other diseases such as IBD, for which the pathology is essential to reach an accurate diagnosis, in this case describing an apoptotic colopathy with the presence of CMV that rules out IBD. Apoptotic colopathy refers to inflammation with apoptosis as a predominant histological feature that can be due to multiple causes such as graft-versus-host disease, drugs, infections (particularly cytomegalovirus and adenovirus), immune disorders and others; which require a specific treatment so it is important to clearly identify them.

References:


Figure 1:

A. EH 40X, glandular structures are observed with marked increase in cryptic apoptosis, increased lymphoplasmacytic infiltrate and neutrophils in the lamina propria.

B. Immunohistochemical studies for CMV showing 3 viral inclusions.

C. Mucosa with erythema, erosion and superficial ulcer with fibrin background.

D. Punch ulcer in the sigmoid colon producing mild stenosis.