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CLINICAL CASE

Acute neurologic disorder in Crohn's disease: A rare life-threatening complication

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Abstract Clinicians should consider and approach inflammatory bowel diseases as a multisystemic disease. Though neurologic complications related to inflammatory bowel diseases are not rare, they are frequently underdiagnosed when compared with other organ complications.

We report on a 40-year-old patient with severe Crohn's disease and an acquired demyelinating polyneurophathy, malnourished, on biological therapy, who was admitted in our institution with an opportunistic infection (esophageal candidiasis). After successful treatment of infectious complication, he maintained unexplained dysphagia and gastric stasis requiring parenteral nutrition. Some weeks later he presented with ophthalmoplegia and cognitive impairment. A clinical diagnosis of Wernicke encephalopathy was suspected despite multivitamin infusion in standard doses. After high doses of intravenous thiamine, dysphagia and gastroparesis improved substantially.

Wernicke encephalopathy is unusual in inflammatory bowel diseases patients and dysphagia is a very rare symptom of thiamine deficiency.

PALAVRAS-CHAVE
Doença inflamatória intestinal;
Doença de Crohn;
Défice de tiamina;
Encefalopatia de Wernicke;
Perturbações da deglutição

Distúrbio neurológico agudo na doença de Crohn: uma complicação grave e rara

Resumo A doença inflamatória intestinal deve ser considerada e abordada clinicamente como uma patologia multissistémica. Apesar das complicações neurológicas relacionadas com a doença inflamatória intestinal não serem raras, são frequentemente subdiagnosticadas quando comparadas com as complicações que afetam outros órgãos.

Reportamos o caso clínico de um doente de 40 anos com doença de Crohn severa e uma polineuropatia desmielinizante adquirida, subnutrido, medicado com terapêutica biológica, admitido na nossa instituição por uma infeção oportunista (candidíase esofágica). Após o tratamento eficaz da complicação infeciosa manteve disfagia e estase gástrica não explicadas, necessitando de nutrição parentérica. Algumas semanas mais tarde foram objetivadas

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oftalmoplegia e alterações cognitivas. Suspeitou-se, pelo quadro clínico, de encefalopatia de Wernicke, apesar da administração parentérica de suplemento multivitamínico em doses *standard*. Foi administrada, empiricamente, tiamina por via endovenosa em doses elevadas, com melhoria clínica significativa, nomeadamente, da disfagia e gastroparésia.

A encefalopatia de Wernicke é uma complicação infrequente em doentes com doença inflamatória intestinal e a disfagia relacionada com défice de tiamina é um sintoma muito raro.

Introduction

Inflammatory bowel disease (IBD), Crohn's disease (CD) and Ulcerative colitis (UC) should be approached as multisystemic diseases. Extraintestinal manifestations in IBD are widely recognized, sometimes precede intestinal symptoms or have a more severe behavior than gastrointestinal involvement. On the other hand, complications secondary to medications can involve virtually any organ or system.

Neurologic complications are not infrequent but are less recognized when compared to other organ complications. Different mechanisms are believed to be involved in the pathogenesis of central and peripheral nervous system disorders, which may present separately or in combination. Neurologic manifestations in patients with IBD can be ascribed to several pathophysiological mechanisms, one being malabsorption and nutritional deficiencies (particularly vitamin B1, B12, D, E, folic acid and nicotinamide).[1,2] In addition, unspecified neuronal influence of enteric disease onto the nervous system (and vice versa) can hypothetically play a role, based on contemporary theories considering the existence of a brain-gut axis, as well as from studies on functional neuroimaging.[3,4]

Case report

The patient was diagnosed with CD in 2001 at the age of 31, after surgery for intestinal obstruction at another institution, resulting in ileocaecal resection with ileocolonic anastomosis. In 2004 he was evaluated for the first time in our institution. At the initial observation, he complained of intermittent diarrhea and weigh loss. He had a body mass index (BMI) of 19.53 kg/m^2 and was medicated with steroids for a long time (steroid-dependent). After further evaluation with blood tests, endoscopic and imaging studies he began treatment with azathioprine. The following year, the disease maintained a high level of activity (abdominal pain, diarrhea and weigh loss), and anti-tumor necrosis factor (TNF) α therapy was initiated (infliximab 5 mg/kg). In 2007, during clinical remission, he was diagnosed with esophageal candidiasis. At that time azathioprine was discontinued. In 2009, he had a clinical relapse and infliximab dosage was adjusted to 10 mg/kg every 8 weeks. In February 2010, disease was still active, the patient continued to lose weight (BMI 13.47) and a biological switch to adalimumab was attempted.

In October 2010 the patient complained for the first time of progressive paraesthesias in both feet and hands and muscular weakness in upper and lower limbs. He could not specify the time of onset of the symptoms (several years) but mentioned an aggravation in the previous month. He was evaluated in the Neurology department and an acquired demyelinating polyneurophathy was diagnosed. Chronic inflammatory demyelinating polyneurophathy related to anti-TNFα therapy was suspected but, because those symptoms had been present for several years, a causal relationship was difficult to establish. We decided to stop anti-TNFα therapy and steroids were started, without clinical improvement.

Short afterwards, in November 2010, he presented with dysphagia. Endoscopic evaluation revealed lesions suggestive of severe esophageal candidiasis. Chest radiography also revealed an infiltrate in the left lung suggesting pneumonia. He began antibiotics, anti-fungic and enteral nutrition (nasogastric feeding tube).

After two weeks, upper endoscopy was repeated and no esophageal lesions were observed. The nasogastric feeding tube was removed; however, the patient maintained complaints of dysphagia and began vomiting.

In December parenteral nutrition was prescribed, adjusted to caloric requirements with multivitamin infusion and trace elements supplementation. Concomitantly, enteral nutrition (nasoenteric feeding tube) was also initiated to stimulate gut protection and function.

Three weeks later, he presented dyspnea and chest radiography revealed pneumonia in the right lung with pleural effusion. Empirical antibiotic therapy was restarted and a right thoracocentesis was performed. The following day, chest radiography revealed a right pneumothorax and a thoracic drain was placed.

One week later, respiratory complications were resolved but esophageal and gastric dysfunctions were still present. The patient was severely malnourished (BMI: 10.93 kg/m^2) with muscular atrophy and complained of visual impairment. On neurologic evaluation, diplopia and limitation of eye movements were detected.

Three days later he presented unusual behavior and disorientation. A cranial computed tomography scan was obtained and acute vascular lesions were excluded. Wernicke encephalopathy (WE) was suspected based in clinical evidence, despite multivitamin supplementation in parenteral nutrition. Laboratory tests to assess thiamine levels and Magnetic Resonance Imaging (MRI) were not promptly available. Empiric treatment with high doses of intravenous thiamine (200 mg 3 times daily) was administrated due to the low incidence of adverse effects of the treatment.