Hypogammaglobulinemic sprue manifested as chronic intestinal failure: An uncommon but effective indication for home parenteral nutrition

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Dear Editor,

Intestinal failure (IF) includes all clinical settings in which the gut function is not sufficient for maintaining a healthy nutritional status and hydroelectrolytic balance without intravenous supplementation (1). This concept includes transient forms (IF type I) that usually occur after abdominal surgery and subacute forms (IF type 2) that may develop in patients with complex enterocutaneous fistula (1). However, the term IF is more often used for describing chronic and permanent conditions included in IF type III, which are mostly observed after multiple intestinal resections resulting in short bowel syndrome. Intestinal dysmotility, mechanical obstruction, and extensive small bowel mucosal disease are rare causes of type III, chronic IF (1,2). Home parenteral nutrition (HPN) is an effective approach for maintaining fluid balance and preventing or treating malnutrition (3).

A 54-year-old woman was referred to our artificial nutrition team because of severe malnutrition and dehydration. Eight years before, she had presented with follicular lymphoma that was treated with rituximab, and after 4 years, she developed diarrhea with significant weight loss. Although severe villous atrophy was confirmed in duodenal biopsies, celiac disease, parasite infection, and lymphoma relapse were excluded after an extensive diagnostic work-up. Hypogammaglobulinemic sprue associated with previous rituximab therapy was assumed considering the striking low levels of all serum immunoglobulin subtypes. Despite several therapeutic approaches, including cyclic antibiotic courses, gammaglobulin supplementation, and corticosteroid and anti-TNFα therapy (infliximab), diarrhea persisted; there was a progressive deterioration in her nutritional status. In addition, she became dependent on regular intravenous supplementation because of recurrent dehydration episodes with low serum potassium, magnesium, and phosphorus levels. On admission to our ward, she presented a body mass index (BMI) of 16.1 kg/m^2, anemia (hemoglobin levels, 9.4g/dlL with depleted iron and folate stores, and low serum protein (total proteins, 3.9g/dL; albumin, 2.7g/dL) levels. After admission, a right subclavian venous catheter was placed and parenteral nutrition was initiated. Although prior electrolyte replacement have been performed and nutritional support initiated using a hypocaloric regimen, refeeding syndrome developed during the 1st week, manifested by hypophosphatemia, hypomagnesemia, and hypokalemia; this was corrected without further complications. A tunneled catheter was placed, and the patient was discharged under HPN. She was maintained under close follow-up by the artificial nutrition outpatient clinic, being assessed every 2 weeks. Initially, HPN was performed daily using 35 Kcal/kg/day infused overnight, supplemented with magnesium and phosphate. Oral intake of gluten- and lactose-free diet was re-introduced to maintain intestinal tropism. Mild diarrhea was initially observed, but it was easily controlled with loperamide. After 3 months, marked improvement in her nutritional status was achieved; the patient recovered 12 kg (BMI: 20.7 kg/m^2), maintained normal fluid and electrolyte balance, and normalized serum protein levels and hematological parameters. HPN was reduced to five weekly administrations, giving the patient two nights "free" of HPN every week. No evidence of liver-, bone-, and catheter-related complications has been detected until now.

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Hypogammaglobulinemia may be a late consequence of rituximab therapy, predisposing patients to recurrent bacterial infections, chronic diarrhea, and malabsorption syndrome due to bacterial overgrowth and small bowel villi atrophy. Actually, these features are the same of common variable immunodeficiency (CVID), a primary immunodeficiency characterized by reduced immunoglobulin serum levels with absent or impaired antibody production (4,7).

Chronic severe malabsorption syndromes are a subtype of IF type III and an established indication for HPN (1). Only one previous report has described the effectiveness of HPN in a single patient with CVID and refractory chronic diarrhea (6). The present case report describes a unique and unreported indication for HPN with a short-term favorable outcome. It highlights the necessity to consider the use of HPN in severe malabsorption disorders and the need to be proficient in managing nutritional issues in everyday gastroenterology practice.

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