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Whipple's Disease in Western Amazonia

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Abstract

The present article reports a case of Whipple's disease as a rare cause of intestinal malabsorption syndrome, in a young individual with G6PD deficiency and hypothyroidism, without previous follow-up. The male patient initially complained of diarrhea and long-standing weight loss. The complementary examination findings were consistent with Whipple's Disease. Antibiotic therapy (Ceftriaxone) was initiated with subsequent clinical improvement. Although rare, this disease should be considered as a differential diagnosis in patients with intestinal malabsorption, especially considering the systemic impact of the disease and the possible control with appropriate antibiotic therapy.

Keywords: Malabsorption Syndrome • Intestinal Malabsorption • Whipple's Disease

Introduction

Whipple's disease (WD) consists of a rare, multisystemic infectious process that is usually characterized by polyarthralgia and an intestinal malabsorption syndrome with weight loss, diarrhea, and abdominal pain. [1] The etiologic agent responsible for this clinical entity is the gram-positive bacillus, Tropheryma whipplei (TW) [2]. The annual incidence is a conflicting figure among researchers, but is believed to average 30 cases per year, with a predilection for white males of European ancestry, suggesting a genetic predisposition. WD represents a diagnostic challenge because it is a multisystemic disease of an infectious nature, and it develops with varied clinical manifestations common to other pathologies. The small intestine is almost always involved, producing malabsorption, lymphangiectasis, and chronic diarrhea, but there are also records of involvement of extraintestinal tissues [3].

Case Presentation

A 22-year-old male patient, brown, from São Felipe D'Oeste, Rondônia, Brazil, with watery diarrhea for 7 years, associated with malnutrition grade III. The diarrheal episodes were up to 6 per day, regardless of food intake or fasting, with stools that were often liquid, without mucus or blood. He presented progressive weight loss, generalized asthenia, and a previous episode of hematochezia. In the last 4 years, he started suffering from polydipsia and polyuria, with daily diuresis of up to 6l. He denied fever, arthralgia and skin manifestations.

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On physical examination, his weight was 40kg and body mass index was 13.6 kg/m². He was bedridden due to difficulty in moving due to his consumptive state, cachectic, dehydrated, with grade-1 muscle strength, and carpopedal spasm.

He presented as previous comorbidities G6PD deficiency and hypothyroidism, without treatment. He reported several visits to the emergency room during periods of worsening diarrheal episodes and was referred for outpatient investigation, but there was loss to follow-up, delaying diagnosis. He denied smoking, alcoholism, and use of illicit drugs.

He was admitted for diagnostic investigation of diarrhea, presenting with deficiency anemia due to iron and cobalamin deficiency and Nephrogenic Diabetes Insipidus. Parenteral nutritional therapy was started, during which the patient evolved with refeeding syndrome, requiring correction of hydroelectrolytic disturbances and reduction of nutritional supplementation, with a more gradual evolution.

Laboratory tests indicates Anti-transglutaminase IgA >0.1U/ml; Gliadin IgA 0.5; IgA endomysium – not reagent (NR); Xylose test 9mg/dL; Lactose Absorption Test – no alternations; Serum G6PD dosage 1.2; TSH 7.8; Free T4 0.72; FAN – NR; C-ANCA – NR; Anti Saccharomyces cerevisiae IgA and IgG – NR; Anti-HIV 1 and 2 – NR.

Esophagogastroduodenoscopy (EGD) showed esophageal moniliasis (Figure 1), moderate enanthematous pangastritis, scalloping of the duodenal mucosal (Figure 2), reduction in Kerckring folds, and histopathological evaluation revealed expansion of the lamina propria by eosinophilic histiocytes positive for Periodic acid–Schiff (PAS) with diastasis. On ileocolonoscopy, sparse erosions were seen in mucosa of the ileum, colon and rectum of mild to moderate grade. Histopathological examination revealed mild nonspecific chronic ileitis with lymphoid hyperplasia, and moderate nonspecific rectocolitis. Abdominal ultrasonography showed fluid within the intestinal loops, accompanied by increased peristalsis and generalized aortic adenopathy.

The meeting of all these findings was consistent with WD, and treatment with intravenous ceftriaxone at a dose of 2 g per day for 15 days was initiated, evolving with clinical improvement, being counter-referred to another hospital treatment unit in the period, losing evolutionary follow-up afterwards.

Discussion

The spectrum of clinical findings due to TW infection is broad. Classic WD

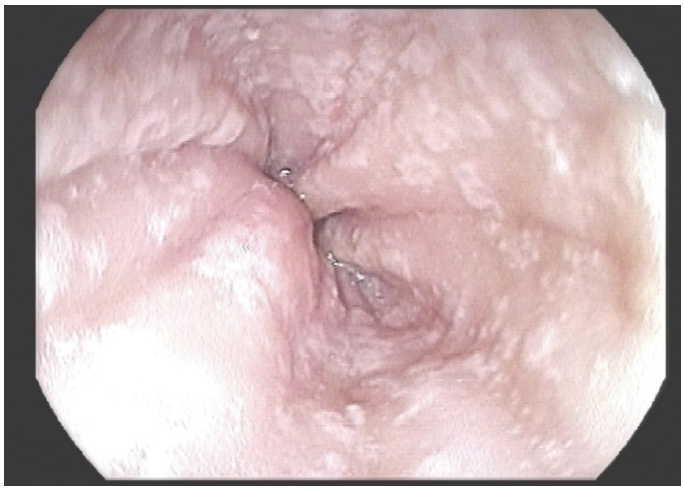

Figure 1. Esophageal mucosa.

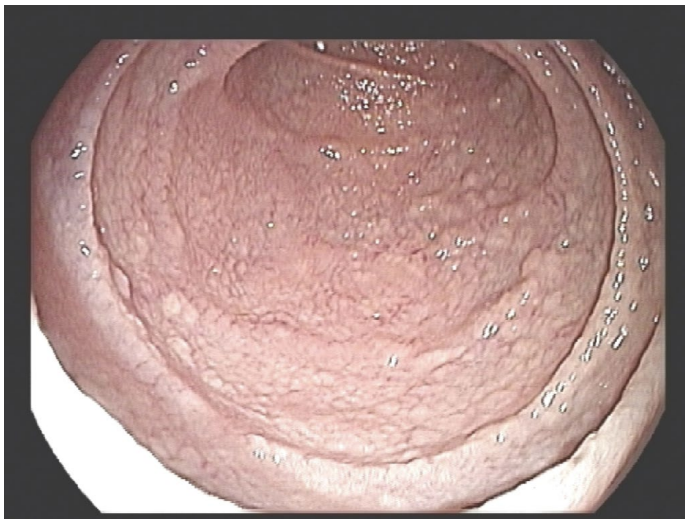

Figure 2. Duodenal mucosa

comprises a multisystemic process characterized by joint symptoms, chronic diarrhea, intestinal malabsorption, and weight loss. Isolated involvement of other organs, primarily the central nervous system and heart valves, can also occur in the absence of classic findings [4]. The patient reported in this case did not present with these other changes.

The age predilection of WD is not yet universally established, although some studies suggest that most patients affected by WD are male, with a mean age of 49 years [5]. In contract, the patient in question is 22 years old.

The diagnosis of WD begins with clinical suspicion, based on the presence of classic signs and symptoms, passing through the exclusion of more common etiologies. Interestingly, the accurate diagnosis in individuals with absence of intestinal manifestations is more challenging [6]. For patients where significant manifestation comprises the gastrointestinal tract, the starting point of the investigation is the performance of the upper digestive endoscopy with biopsies of the small intestine for the test of TW, i.e., histology with PAS, PCR and immunohistochemistry [7,8].

Radiological and endoscopic changes aid the investigation and may suggest the diagnosis of WD. On EGD, mucosal fold thickening and confluent whitish exudates alternating with erosions and regions of mucosal reliability are often found [8,9]. Abdominal ultrasound or abdominal CT usually shows enlargement of the lymph nodes of the mesentery. Because it courses with lymphadenomegaly, it is necessary to exclude some differential diagnoses, such as tuberculosis and lymphoma, diseases that may course clinically similar to WD [10,11].

For patients with chronic WD (i.e., classic chronic or localized infection), a suggested option consists of an initial phase for 14 days with an intravenous antibiotic that is active against TW and able to penetrate the blood-brain barrier, and may be with ceftriaxone (2 g IV daily) or penicillin (2 MU IV every 4 hours), followed by 12 months of oral maintenance therapy with sulfamethoxazole-trimethoprim (160/800 mg) [10].

Clinical failure is suggested by a positive PCR in the sample of patients who do not respond clinically to therapy or have recurrence of symptoms after initial improvement. Relapses are frequent and occur on average 4 years after diagnosis, with the central nervous system being one of the preferred sites. For this reason, clinical follow-up of patients with WD should be maintained for at least 10 years, with clinical and laboratory monitoring, aiming to normalize intestinal absorption parameters and regression of histopathological changes [11,12].

Conclusion

Like other rare diseases, WD is often diagnosed late. Thus, physicians must consider it among differential diagnoses when patients present with clinical intestinal malabsorption, especially due to its high potential for morbidity and mortality. Furthermore, WD is treatable once the antimicrobial therapy is established. WD patients must be clinically monitored for years, even after the end of the antibiotic regimen, to evaluate the response to treatment and the possibility of relapse.

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