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Case Report

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Whipple's Disease in Older Adults

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Abstract

Background: *Whipple's disease is a rare disease that has been described as a multisystem infectious disorder caused by Thropheryma whipplei, and characterized by joint problems, weight loss, diarrhea, and other symptoms that can clinically overlap with rheumatic diseases.*

Case presentation: *We present the case of an 85-year-old man with long story of poliartralgias treated with corticosteroids, admitted to the hospital with severe symptomatic anemia. A gastroscopy study was made as part of the study. which showed intestinal edema and PAS postivioe as a sing of WD.*

Conclusions: *WD should be considered in all patients with joint symptoms, chronic diarrhea and weight loss, particularly in males of European ancestry, and those with rheumatoid factor negative migratory poliarthritis that does not respond to immunosuppressive therapy [1].*

Keywords: Whipple`s Disease, Arthralgias, Weight Loss, Diarrhea

List of Abbreviations

WD: Whipple Disease

1. Background

Chronic pain is highly prevalent among older adults where it is associated with significant disability, suffering and greater costs to health care systems. The most likely etiology is osteoarthritis, which has grown exponentially with the increase in the geriatric population. However, the differential diagnosis includes conditions which should not be missed. In that regard, it has been described a multisystem infectious disorder called Whipple's Disease (WD), caused by Thopheryma whipplei, a gram-positive bacillus related to Actinomycetes and characterized by four cardinal manifestation: joint pain, diarrhea, abdominal pain and weight loss [2]. These symptoms can clinically overlap with rheumatic diseases, potentially resulting in misdiagnosis.

WD has been considered a rare disease. In Spain, there is very limited data until 1970. Since then, and up to the end of 2001, 72 cases were reported, with an average of two or three a year [3]. The most frequent symptoms reported by these patients were joint problems, particularly oligoarticular arthralgias (56%). The most common symptoms at diagnosis were weight loss (80%), diarrhea (63%), skin symptoms (32%), abdominal pain (27%) and joint symptoms (20%). It is important to mention that joint symptoms can precede others by many years, so not all symptoms may be manifest at the time of presentation.

Although the pathogenesis of WD remains obscure, many investigators have postulated a host immune deficiency as a predisposition to the disease, but also an immune down regulation induced by the bacterium, supported by the lack of immunologic response and widespread invasion of the bacillus through the body, including intestinal epithelium, endothelium, brain, liver, heart, lung, kidney, colon, synovium, bone marrow, macrophages and skin [4-7]. All of these sites show a remarkable lack of inflammatory response to the bacillus. There have been reports of WD being unmasked or accelerated by immunosuppressive therapy, typically given for presumed rheumatic disease, in some cases with severe complications, such us disseminated infection [8].

2. Case Presentation

We present the case of an 85-year-old man admitted to the hos-

pital complaining of breathless in the last week. His medical history included atrial fibrillation anticoagulated; a conventional tubular adenoma with hight-grade epithelial dysplasia in the ascending colon treated with mucosectomy, and partial gastrectomy for ulcus. The patient also complained of chronic polyartritis of the large joints and mialgias, treated for presumed rheumatic disease with corticosteroids for many years. Gradual loss of weight and strength with episodes of intermittent diarrhea were also present. No other cardiovascular disease or risk factors such as hypertension or diabetes.

Lab tests were performed and showed severe microcytic anemia and increase of inflammatory markers. The X ray revealed bilateral pleural effusion, that improved with parenteral furosemide. No evidence of SARS-CoV-2 infection was found. A Gastroscopy was performed, which showed a Bill Roth type I anastomosis with erythematous mucosa and edema, and a lesion suggestive of lymphangiectasia in the jejunal loop, with no signs of bleeding. In regards to suspected infection, the patient was treated with Amoxicillin-Clavulanic with progressive improvement. Finally, endoscopic biopsies showed in the small intestine mucosa abundant foamy macrophages in the lamina propria, and PAS staining with strong granular positivity for macrophages, considered the classic finding that makes the diagnosis of WD [2,9]. Results of PCR testing also confirmed WD.

3. Descriptive Legend for Figures

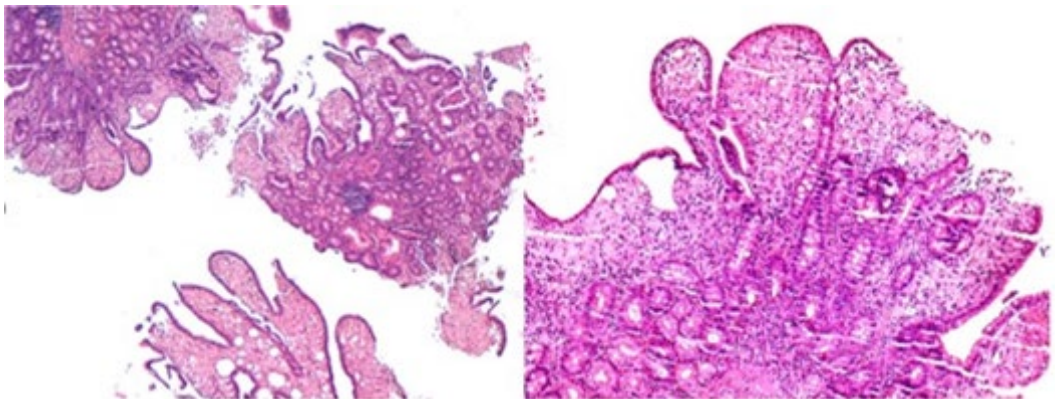

Figure 1: (He 4x and 10x). Small bowel biopsy with abnormal appearing mucosa. Duodenal villi occupied by foamy macrophages in the lamina propia and villous atrophy

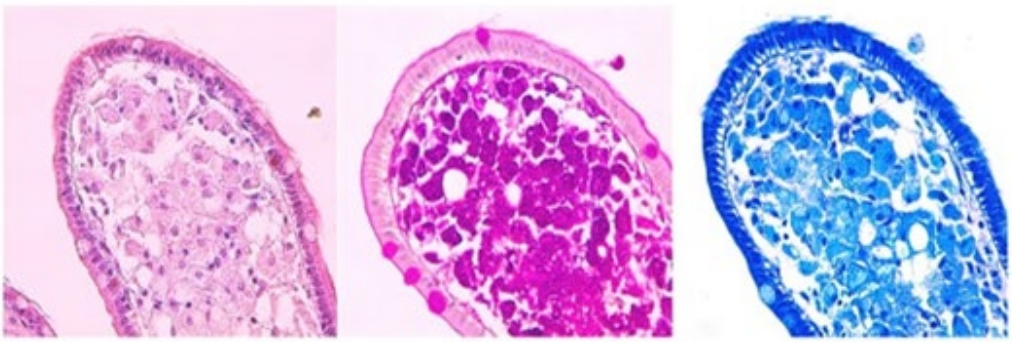

Figure 2: (He, pas y giemsa). Histochemistry techniques. Pas-positive macrophages with granular pattern and cytoplasmic location

4. Discussion and Conclusions

WD has a reputation as a great mimicker of many different illness. The first step in the evaluation of WD is to assess for alternative symptoms and more common diagnoses. Often WD is not suspected until such diagnoses have been ruled out [1]. Upper gastrointestinal endoscopy with biopsies of the small intestine has been the test of choice. It is generally the first test performed for the evaluation of WD in patients with gastrointestinal symptoms.

The diagnosis of WD can be made with the classic finding of PAS-positive macrophages from a small bowel biopsy. In the absence of this finding, the diagnosis can also be made when two different T. Whipplei tests (PCR testing, PAS staining or immunohistochemistry) from the same specimen or two tests from different specimens are positive. This disorder requires prolonged antibiotic therapy. Although the optimal regimen is uncertain, there has been proposed an initial phase of an intravenous antibiotic such us penicillin or ceftriaxona for two weeks, followed by maintenance therapy with oral trimethoprim-sulfamethoxazole given for 12 months [10]. The aim of this text is to encourage and take account that this disease should be considered in all patients with joint symptoms, chronic diarrhea and weight loss, particularly in males of European ancestry, and those with rheumatoid factor negative migratory poliarthritis that does not respond to immunosuppressive therapy [1].

Ethics Approval and Consent to Participate

This work has written and signed consent to publish the information from the patient.

Consent for Publication

This work has written institutional consent for publication.

Availability of Data and Materials

The datasets used and/or analyzed during the current are available from the corresponding author on reasonable request.

Competing Interests and Funding

None of the authors have financial or personal relationships with people or organizations that could inappropriately influence their work. There are no conflicts of interest to declare.

Author Contributions

All authors made substantial contributions to the information submitted and have read and approved the manuscript.

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