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DOI: 10.17235/reed.2022.9338/2022
Link: PubMed (Epub ahead of print)

Please cite this article as:
Valadas Ana, Fatela Narcisa, Sepúlveda Santos Tiago, Carvalho Maria Leonor. Whipple's disease – A typical endoscopic finding of a rare disease. Rev Esp Enferm Dig 2022. doi: 10.17235/reed.2022.9338/2022.

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IPD 9338

Whipple's disease – A typical endoscopic finding of a rare disease

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Received: 15/11/2022

Accepted: 16/11/2022

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Conflict of interest: the authors declare no conflict of interest.

Informed consent: patient consent for publication was obtained.

Data availability statement: all data generated or analyzed during this study are included in this article and the supplementary material files. Further enquiries can be directed to the corresponding author.

Author contributions: Ana de Matos Valadas: preparation of the manuscript. Narcisa Fatela: image preparation and intellectual contribution. Tiago Sepúlveda Santos and Maria Leonor de Carvalho: intellectual contribution and review of the manuscript.

CASE REPORT

A 49-year-old female presented with a five-month course of diarrhea, nocturn abdominal pain, asthenia and weight loss of 30 % of her body mass in the last three months. The patient also had a four-year medical history of bilateral mechanic gonalgia and arthralgias of the metacarpophalangeal and interphalangeal joints, despite treatment with prednisolone. On examination, the patient had hyperpigmentation of the face and thorax, low-grade fever

and a body mass index (BMI) of 15.8 kg/m^2. Diarrhea was documented with watery brownish stools seven times per day, despite loperamide, with no visible blood or mucous.

Since the upper gastrointestinal (GI) endoscopy and colonoscopy had no macroscopic abnormalities, the patient underwent a capsule endoscopy, which revealed a continuous mucosal lesion with lymphangiectasia, edema, villous atrophy and areas of denudation with hematinic punctate from the duodenum to the ileum (Fig. 1). A diagnosis of Whipple's disease was made with typical histology findings in duodenum material and a positive PCR for *Tropheryma whipplei*. Complete investigation revealed secondary cerebral demyelination and endocarditis of three valves, with negative blood cultures. Treatment with intravenous ceftriaxone for four weeks and oral cotrimoxazole for one year (2) resulted in complete resolution of the symptoms and endocarditis (Fig. 2).

DISCUSSION

This case shows the importance of recognizing the typical macroscopic feature of this rare condition, with a worldwide incidence of about 30 cases per year. Given the lack of literature on this disease, it is important to share the clinical findings to better diagnose this condition, considering that it can be fatal when not recognized and properly treated (3).

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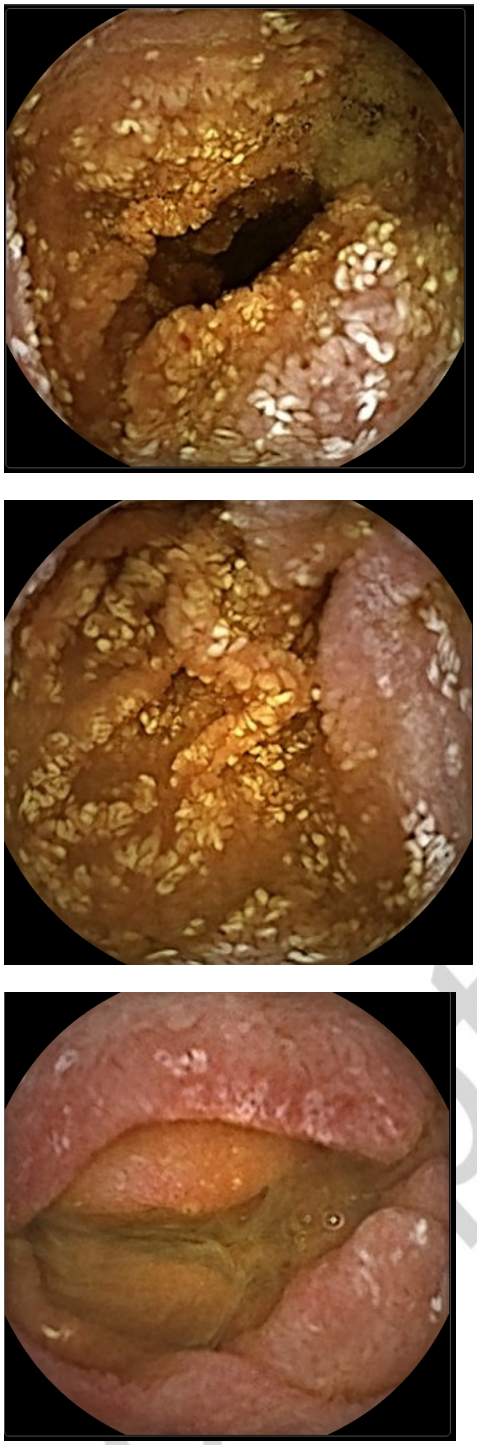

Fig. 1. Images from capsule endoscopy, suggestive of enteropathy and macroscopically characteristic of Whipple's disease. A. Lymphangiectasia in D2. B. Lymphangiectasia and areas of denudation with hematinic punctate in D3. C. Oedema of the ileum.

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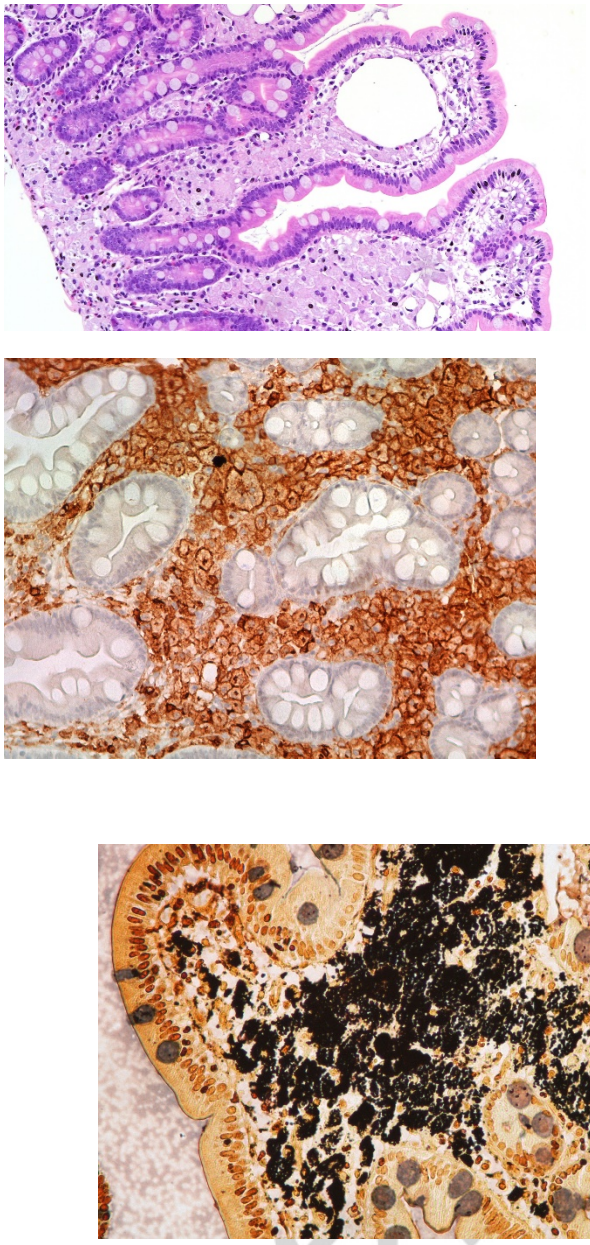

Fig. 2. Histological photos confirming the histopathological diagnosis of Whipple's disease. A. HE, 100x. The duodenal mucosa showed a diffuse infiltration of the lamina propria and submucosa by PAS+ foamy macrophages that blunted and distended the villi. Lymphangiectasia was also seen (arrow). B. CD68, 200x. The cells expanding the lamina propria showed a strong and diffuse membrane and cytoplasmic staining for CD68 immunohistochemistry marker, confirming their histiocytic nature. C. Warthin-Starry, 200x. The foamy macrophages showed intense staining, with a Warthin-Starry stain, of the intracellular material that gives their cytoplasm their characteristic granularity.