A less common cause of diarrhoea

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CASE REPORT

A 42-year-old man was admitted to the hospital because of watery diarrhoea and weight loss. He had been diagnosed with seronegative arthritis ten years earlier, for which he was taking methotrexate. At that time he had no other complaints, notably no diarrhoea. We cannot rule out the possibility that the arthritis was the primary presentation of the disease. He was not on NSAIDs. On physical examination no abnormalities were found. His laboratory studies showed an iron deficiency anaemia with normal white cell account and a C-reactive protein of 128 mg/l (normally <5 mg/l). As inflammatory bowel disease was suspected, colonoscopy was performed, revealing normal mucosa. Wireless capsule endoscopy (WCE) showed markedly abnormal mucosa, with a pseudopolypoid appearance and numerous small white spots (*figure 1*).

WHAT IS YOUR DIAGNOSIS?

See page 402 for the answer to this photo quiz.



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ANSWER TO PHOTO QUIZ (PAGE 401) A LESS COMMON CAUSE OF DIARRHOEA

DIAGNOSIS

The clinical presentation and images obtained with WCE support the diagnosis of Whipple's disease.¹ Histological findings from duodenal biopsies revealed periodic acid Schiff (PAS)-positive macrophages typical for this disease (*figure 2*). Bacterial polymerase chain reaction (PCR) was positive for *Tropheryma whipplei*. The patient was treated with a third-generation cephalosporin (ceftriaxone) intravenously for two weeks, and after discharge he continued treatment with trimethoprim-sulphamethoxazole for one year. Within two weeks his diarrhoea had disappeared. After two months, he had no more complaints of arthralgia and had gained weight. Whipple's disease is a rare, systemic disease caused by *Tropheryma whipplei*. Its clinical manifestation is diarrhoea, weight loss and fever. Extraintestinal disease often involves the brain, the heart





and the joints.² Patients without the classic symptoms of gastrointestinal disease may be misdiagnosed. Patients who are not treated or insufficiently treated can experience fatal outcome or irreversible neurological damage.

The diagnosis of Whipple's disease is made by the presence of PAS-positive macrophages in histological specimens from the small bowel. Organisms at different stages of degeneration are seen within phagosomes in macrophages which also contain abundant irregular membranous inclusions representing remnants of the bacterial capsule, which are the equivalent of the PAS-positive material seen at light microscopy. PCR assay for Tropheryma whipplei is positive, but can also be positive in asymptomatic subjects. There are several publications describing capsule endoscopy in Whipple's disease.3 Recommended therapy is a third-generation cephalosporin for 10 to 14 days followed by long-term treatment for one year with trimethoprimsulphamethoxazole.4 Follow-up of patients with Whipple's disease could be based on quantitative PCR. If patients have a good clinical response, they can simply be followed up with duodenal biopsies 6 months and 12 months after diagnosis. Antibiotic treatment can generally then be stopped if no PAS-positive material is identified.4

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