We report the case of a 43-year-old woman with a medical history of active smoking who presented with abdominal pain that had appeared 4 months ago. The physical examination revealed peri-umbilical pain associated with weight loss. Laboratory studies revealed elevated inflammatory markers (C-reactive protein: 54 mg/dL) and hypoalbuminemia (22 g/L). Abdominal computed tomography (CT) and magnetic resonance enterography (MRE) showed a thickening of the small bowel at the junction between the jejunum and ileum, at 30 cm, suggesting Crohn’s disease. Gastroscopy showed an appearance of duodenal villous atrophy, confirmed by Marsh stage 3c biopsy findings. Anti-transglutaminase antibodies were positive at 17 U/ml. 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET) detected a jejunoileal accumulation of 18F-FDG, with a standard uptake value (SUV) at of 8.1. Small-bowel capsule endoscopy confirmed the appearance of villous atrophy with the presence of sporadic ulceration (Fig. 1), then a completely ulcerated jejunoileal segment over several tens of centimeters, with necrotic pseudomembranous and hemorrhagic spots (Fig. 2, Video 1). Biopsies of this ulcerated area were performed with double-balloon enteroscopy (Fig. 3), histological examination revealing refractory type 2 celiac disease (RCD) characterized by an increased clonal population of abnormal intraepithelial lymphocytes (IEL). Abnormal IEL phenotype was described with disappearance of the classical markers such as CD4 or CD5 and the presence of NKp46 marker, but without any sign of high-grade lymphoma (Fig. 4). Systemic corticosteroid therapy was initiated but did not improve the patient’s symptoms. She developed an occlusive syndrome requiring surgical resection of the ulcerated jejunoileal segment (39 cm). The surgical follow-up was simple. The diagnosis of type 2 RCD without high-grade lymphoma was confirmed. Gluten-free diet and open-capule budesonide treatment were initiated with close surveillance of the patient due to the high risk of progression to T-cell lymphoma.

About 5% of patients with celiac disease do not respond to a gluten-free diet and are at risk of developing RCD. Type 2 RCD is characterized by infiltration of monoclonal intraepithelial T cells and is considered as a low-grade intraepithelial lymphoma with a high risk of progression to aggressive T-cell lymphoma [1]. In this situation, imaging (CT, MRE, PET) is inadequate because it lacks sensitivity and specificity. Exploration of lesions in the small bowel is complex and can lead to a prejudicial delayed diagnosis. Small-bowel capsule endoscopy and enteroscopy are relevant tests to explore suspected refractory celiac disease [2–3].
Competing interests
None

The authors
Jean-Philippe Le Mouel¹, Mathurin Fumery¹, Sami Hakim¹, Clara Yzet¹, Alexandra Dervaux², Xavier Dray³, Eric Nguyen-Khac¹
1 Department of Hepatogastroenterology, Amiens University Hospital, Amiens, France
2 Department of Pathology, Amiens University Hospital, Amiens, France
3 Endoscopy Unit, Department of Digestive Diseases, Sorbonne Université, APHP Hôpital Saint-Antoine, 75012 Paris, France

Corresponding author
Jean-Philippe Le Mouel, MD
Department of Hepatogastroenterology, Amiens University Hospital, Amiens, France
jeanphilippe.lemouel@wanadoo.fr

Video 1 Typical endoscopic appearance of ulcerative jejunitis secondary to refractory celiac disease type 2.

Fig. 3 Double-balloon enteroscopy: ulcerated jeunoileal segment.

Fig. 4 Increased clonal population of abnormal intraepithelial lymphocytes: CD3+/CD5−.
References


Bibliography

DOI https://doi.org/10.1055/a-1046-1593
Published online: 2019
Endoscopy
© Georg Thieme Verlag KG
Stuttgart · New York
ISSN 0013-726X

Endoscopy E-Videos is a free access online section, reporting on interesting cases and new techniques in gastroenterological endoscopy. All papers include a high quality video and all contributions are freely accessible online.
This section has its own submission website at https://mc.manuscriptcentral.com/e-videos