Langerhans cell histiocytosis: a rare case of large-bowel inflammatory lesions

Langerhans cell histiocytosis (LCH) is a rare disease characterized by clonal neoplastic proliferation of Langerhans cells, with typical expression of CD1a, S100 protein, and langerin [1, 2]. Gastrointestinal involvement is extremely rare and usually observed in systemic illness with poor prognosis and high morbidity [2–4].

Here, we report the case of a 48-year-old man affected by mucocutaneous Langerhans disease, presenting with severe weight loss, constipation, and nausea. He was previously diagnosed with LCH with pituitary localization in 2002 and underwent treatment with vinblastine, steroids, and mercaptopurine. An esophagogastroduodenoscopy was unremarkable, including duodenal and gastric biopsies. Ileocolonoscopy was performed using a high definition colonoscope with optical enhancement (OE mode) virtual chemoendoscopy (Optivista EPK-I7010 video processor; Pentax, Tokyo, Japan) [5]. During withdrawal, the OE mode 2 was activated to increase the detection of subtle mucosal changes, revealing multiple yellowish nodules of 5–10 mm in size, with central erosions or aphthous ulcers (▶ Video 1). OE mode 1 showed
both the pit and vascular patterns to be clearly characterized as nonadenomatous (Kudo II) and irregular or interrupted (Fig. 1, Video 1).

Histology on targeted biopsy samples demonstrated submucosal proliferation of histiocytes, with abundant eosinophilic cytoplasm, positive anti-CD1a, anti-CD68 R, and S100 protein immunohistochemistry, which are pathognomonic for colonic localization of LCH (Fig. 2).

This case is relevant for several reasons. First, we describe an unusual case of gastrointestinal LCH, which unconventionally manifested with symptoms that warranted endoscopic investigations. In addition, we provide for the first time detailed video and image documentation of intestinal LCH to further support clinicians and endoscopists during their everyday practice.

Endoscopy_UCTN_Code_CCL_1AD_2AC

Competing interests

None

The authors

Beatrice Marinoni1, Federica Cavalcoli1, Alessandro Del Gobbo2, Helmut Neumann3, Emanuela Passoni4, Maurizio Vecchi2, 5, Gian Eugenio Tontini1

1 Gastroenterology and Endoscopy Unit, Fondazione IRCCS Ca’ Granda Ospedale Maggiore Policlinico, Milan, Italy
2 Pathology Unit, Fondazione IRCCS Ca’ Granda Ospedale Maggiore Policlinico, Milan, Italy
3 First Medical Department, University Medical Center Mainz, Mainz, Germany
4 Dermatology Unit, Fondazione IRCCS Ca’ Granda Ospedale Maggiore Policlinico, Milan, Italy
5 Department of Pathophysiology and Transplantation, University of Milan, Milan, Italy

Corresponding author

Beatrice Marinoni, MD
Gastroenterology and Endoscopy Unit, Fondazione IRCCS Ca’ Granda Ospedale Maggiore Policlinico, Via della Commenda 12, 20122 Milan, Italy
Fax: +39-02-55033414
beatrice.marinoni@unimi.it

References