Antonello Trecca *Editor*

Atlas of Ileoscopy

A Collection of Clinical Cases

Springer

Atlas of Ileoscopy

Antonello Trecca
Editor

Atlas of Ileoscopy

A Collection of Clinical Cases

Springer

Editor
Antonello Trecca
Department of Operative Endoscopy
Usi Group
Rome, Italy

ISBN 978-88-470-5204-8 ISBN 978-88-470-5205-5 (eBook)

DOI 10.1007/978-88-470-5205-5

Springer Milan Heidelberg New York Dordrecht London

Library of Congress Control Number: 2012955511

9 8 7 6 5 4 3 2 1 2013 2014 2015 2016

Cover design:Ikona S.r.l., Milan, Italy
Typesetting: Graphostudio, Milan, Italy
Printing and binding: Grafiche Porpora, Segrate (Milan), Italy

Springer-Verlag Italia S.r.l. – Via Decembrio 28 – I-20137 Milan
Springer is a part of Springer Science+Business Media (www.springer.com)

Preface

Last year I was very proud to edit my first book dedicated to the diagnosis and treatment of ileal diseases, published under the simple title *Ileoscopy*.

The book's success derived from the participation of so many colleagues from several countries, who demonstrated their passion and competence in their contributions on this new topic of research in the field of modern gastroenterology and endoscopy.

Nonetheless, I never imagined that Springer would return with an even greater challenge: an atlas focused on this same topic, with brilliant images and concise but insightful discussions on the most important issues stemming from clinical practice!

But once again I was fortunate in being able to call upon my skilled colleagues, who, as before, did not disappoint in that they have been invaluable in sharing their knowledge. The result is a very interesting and user-friendly book that will not fail to attract the close attention of its readers, whether they are simply curious, wish to compare notes on a similar experience, consider a rare disease, view a particular image, update their skills, or, of course, share their passion for the world's most beautiful profession: medicine.

So please enjoy our book and let's hope that we meet again!

Rome, December 2012 Antonello Trecca

Contents

Surgery

Histology

Miscellaneous

Contributors

Giovanni Alfano Department of Surgery, Minimally Invasive and Robotic Surgery Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Luciana Ambrosiani Pathology Department, Valduce Hospital, Como, Italy

Andrea Anderloni Gastroenterlogy Unit, "Maggiore della Carità" Hospital, Novara, Italy

Vincenzo Arena Institute of Pathology Catholic University, Rome, Italy

Stefano Ascani Institute of Pathologic Anatomy, University of Perugia in Terni, St. Mary Hospital, Terni, Italy

Antonio Astolfi Department of Gastroenterology, Teramo, Italy

Alessandra Baccarin Gastroenterology Unit, Valduce Hospital, Como, Italy

Paolo Baldassari Department of Radiological Sciences, Oncology and Pathology, "Sapienza" University of Rome, Umberto I Hospital, Rome, Italy

Fabio Baldi Digestive Endoscopy Unit, General Surgery Department, Tarquinia General Hospital, Tarquinia (VT), Italy

Gabrio Bassotti Department of Clinical and Experimental Medicine, University of Perugia, Italy

Andrea Belli Abdominal Oncological Surgery, National Cancer Institute and G. Pascale Foundation, Naples, Italy

Alessandra Bizzotto Digestive Endoscopy Unit, Catholic University, Rome, Italy

Lorenzo Bonomo Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Vincenzo Bruzzese Gastroenterlogy Unit, Nuovo "Regina Margherita" Hospital, Rome, Italy

Maria Gabriella Brizi Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Emma Calabrese Department of Gastroenterology, Tor Vergata University of Rome, Rome, Italy

Tamara Caldaro Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Cono Casale Unit of Gastroenterology and Internal Medicine, Catholic University-Columbus, Rome, Italy

Giuseppe Cerno Department of Histopathology, Usi Group, Rome, Italy

Paola Cesaro Digestive Endoscopy Unit, Catholic University of Rome, Italy

Maria Ciolina Department of Radiological Sciences, Oncology and Pathology, ''Sapienza'' University of Rome, Umberto I Hospital, Rome, Italy

Sergio Coda Department of Endoscopy, Section of Gastroenterology and Hepatology, Imperial College London, London, UK

Guido Costamagna Digestive Endoscopy Unit, Catholic University, Rome, Italy

Antonio Crucitti General Surgery, Cristo Re Hospital, Rome, Italy

Giancarlo D'Ambrosio Department of Surgery, "Sapienza" University of Rome, Rome, Italy

Valentina D'Angelo Endoscopy Unit, National Cancer Institute and G. Pascale Foundation, Naples, Italy

Luigi Dall'Oglio Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Annibale D'Annibale Department of Surgery, Minimally Invasive and Robotic Surgery Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Paola De Angelis Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Mario de Bellis Endoscopy Unit, National Cancer Institute and G. Pascale Foundation, Naples, Italy

Roberto de Franchis University of Milan, Gastroenterology Unit, L. Sacco Hospital, Milan, Italy

Francesca De Laurentiis Department of Surgery, "Sapienza" University of Rome, Rome, Italy

Angelo De Padova Gastroenterology and Digestive Endoscopy Unit, "Morgagni-Pierantoni" Hospital, Forlì, Italy

Italo de Vitis Unit of Gastroenterology and Internal Medicine, Catholic University-Columbus, Rome

Anselmo Della Spoletina Department of Operative Endoscopy, Saint Mary Hospital, Terni, Italy

Davide Diacinti Department of Radiological Sciences, Oncology and Pathology, "Sapienza" University of Rome, Umberto I Hospital, Rome, Italy

Elena Di Girolamo Endoscopy Unit, National Cancer Institute and G. Pascale Foundation, Naples, Italy

Nadia Di Lorenzo Pathology Department, Valduce Hospital, Como, Italy

Giovanni Di Nardo Department of Pediatrics, Pediatric Gastroenterology and Liver Unit, "Sapienza" University of Rome, Rome, Italy

Maria C. Di Paolo Department of Surgery, Gastroenterology and Digestive Endoscopy Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Alessandra Farchione Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Giovanni Federici di Abriola Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Francesca Foschia Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Paola Francalanci Department of Pathology, Bambino Gesù Children's Hospital, Rome, Italy

Kuangi Fu Department of Gastroenterology, Juntendo University Nerima Hospital, Nerima City, Tokyo, Japan

Takahiro Fujii Takahiro Fujii Clinic, Digestive Endoscopy, Tokyo, Japan

Fabio Gaj Department of Surgery, "Sapienza" University of Rome, Rome, Italy

Antonio Gasbarrini Internal Medicine and Gastroenterology Unit, Catholic University, Rome, Italy

Viviana Gerardi Internal Medicine and Gastroenterology Unit, Catholic University, Rome, Italy

Ugo Grossi General Surgery, UCSC Roma, Rome, Italy

Cesare Hassan Gastroenterlogy Unit, Nuovo "Regina Margherita" Hospital, Rome, Italy

Federico Iacopini Gastroenterology Unit, S. Giuseppe Hospital, Albano L., Rome, Italy

Franco Iafrate Department of Radiological Sciences, Oncology and Pathology, ''Sapienza'' University of Rome, Umberto I Hospital, Rome, Italy

Marcella Iannitti Department of Radiological Sciences, Oncology and Pathology, ''Sapienza'' University of Rome, Umberto I Hospital, Rome, Italy

Alessandro Inserra General and Thoracic Surgery Unit, Bambino Gesù Children's Hospital, IRCCS, Rome, Italy

Prashant Kant Endoscopy Department, Leeds Teaching Hospitals NHS Trust, Leeds, West Yorkshire, UK

Ichiro Konuma Konuma Clinic, Nasushiobara City, Tochigi, Japan

Hironori Konuma Department of Gastroenterology, Juntendo University Nerima Hospital, Nerima City, Tokyo, Japan

Shin-ei Kudo Showa University Northern Yokohama Hospital, Digestive Disease Center, Yokohama, Kanagawa, Japan

Andrea Laghi Department of Radiological Sciences, Oncology and Pathology, "Sapienza" University of Rome, Umberto I Hospital, Rome, Italy

Luigi Larosa Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Roberto Lorenzetti Gastroenterlogy Unit, Nuovo "Regina Margherita" Hospital, Rome, Italy

Giorgio Lucandri Department of Surgery, Minimally Invasive and Robotic Surgery Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Makomo Makazu Endoscopy Division, National Cancer Center Hospital, Tokyo, Japan

Stefania Manenti Department of Pathology, Spedali Civili, Brescia, Italy

Raffaele Manta Digestive Endoscopy Unit, New Civil Hospital S. Agostino Estense of Modena, Modena, Italy

Gabriele Marinozzi Department of Operative Endoscopy, Saint Mary Hospital, Terni, Italy

Pietro Marone Endoscopy Unit, National Cancer Institute and G. Pascale Foundation, Naples, Italy

Rosa Marra Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Manuela Marzo Unit of Gastroenterology and Internal Medicine, Catholic University-Columbus, Rome

Takahisa Matsuda Endoscopy Division, National Cancer Center Hospital, Tokyo, Japan

Andrea Mazzari General Surgery, Cristo Re Hospital, Rome, Italy

Paolo Mazzocchi Department of Surgery, Minimally Invasive and Robotic Surgery Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Laura M. Minordi Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Takeshi Nakajima Endoscopy Division, National Cancer Center Hospital, Tokyo, Japan

Salvatore Oliva Department of Pediatrics, Pediatric Gastroenterology and Liver Unit, "Sapienza" University of Rome, Rome, Italy

Silvia Paggi Gastroenterology Unit, Valduce Hospital, Como, Italy

Amilcare Parisi Department of General and Digestive Surgery, Saint Mary Hospital, Terni, Italy

Vito Pende Department of Surgery, Minimally Invasive and Robotic Surgery Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Graziano Pernazza Department of Surgery, Minimally Invasive and Robotic Surgery Unit, San Giovanni-Addolorata Hospital, Rome, Italy

Lucio Petruzziello Digestive Endoscopy Unit, Catholic University of Rome, Italy

Alessandro Pichi Department of Radiological Sciences, Oncology and Pathology, ''Sapienza'' University of Rome, Umberto I Hospital, Rome, Italy

Francesca Rea Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Bjorn J. Rembacken Endoscopy Department, Leeds Teaching Hospitals NHS Trust, Leeds, West Yorkshire, UK

Enrico Ricci Gastroenterology and Digestive Endoscopy Unit, "Morgagni-Pierantoni" Hospital, Forlì, Italy

Maria Elena Riccioni Digestive Endoscopy Unit, Catholic University, Rome, Italy

Patrizia Rigato Pathology Unit, S. Giuseppe Hospital, Marino, Rome, Italy

Erminia Romeo Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Emanuele Rondonotti Gastroenterology Unit, Valduce Hospital, Como, Italy

Giovanni Battista Rossi Endoscopy Unit, National Cancer Institute and G. Pascale Foundation, Naples, Italy

Taiji Saga Department of Gastroenterology, Juntendo University Nerima Hospital, Nerima City, Tokyo, Japan

Yutaka Saito Endoscopy Division, National Cancer Center Hospital, Tokyo, Japan

Taku Sakamoto Endoscopy Division, National Cancer Center Hospital, Tokyo, Japan

Franco Scaldaferri Internal Medicine and Gastroenterology Unit, Catholic University, Rome, Italy

Nigel Scott Histopathology Department, Leeds Teaching Hospitals NHS Trust, Leeds, West Yorkshire, UK

Agostino Scozzarro Gastroenterology Unit, S. Giuseppe Hospital, Albano L., Rome, Italy

Takayoshi Shimizu Department of Gastroenterology, Juntendo University Nerima Hospital, Nerima City, Tokyo, Japan

Cristiano Spada Digestive Endoscopy Unit, Catholic University of Rome, Italy

Giancarlo Spinzi Gastroenterology Unit, Valduce Hospital, Como, Italy

Francesca Stella Gastroenterlogy Unit, Nuovo "Regina Margherita" Hospital, Rome, Italy

Gianfranco Tappero Department of Gastroenterlogy, Gradenigo Hospital, Turin, Italy

Pasquina M.C. Tomaiuolo General Surgery, Cristo Re Hospital, Rome, Italy

Filippo Torroni Digestive Endoscopy and Surgery Unit, Department of Surgery and Transplantation, IRCCS, Bambino Gesù Children's Hospital, Rome, Italy

Antonello Trecca Department of Operative Endoscopy, Usi Group Rome, Italy

Pasquale Trecca University Campus Bio Medico, Rome, Italy

Paolo Trentino Digestive Endoscopic Service, Department of General and Transplantation Surgery "Paride Stefanini", "Sapienza" University of Rome, Rome, Italy

Hiroya Ueyama Department of Gastroenterology, Juntendo University Nerima Hospital, Nerima City, Tokyo, Japan

Maxim Untesco Department of Pathology, Spedali Civili, Brescia, Italy

Riccardo Urgesi Department of Gastroenterology, Belcolle Hospital, Viterbo

Amorino Vecchioli Department of Imaging and Radiological Sciences, Catholic University, Rome, Italy

Vincenzo Villanacci Department of Pathology, Spedali Civili, Brescia, Italy

Marina Yarygina Department of Pathology, Spedali Civili, Brescia, Italy

Angelo Zullo Gastroenterlogy Unit, Nuovo "Regina Margherita" Hospital, Rome, Italy

The Asymptomatic Patient

Case 1
Ileoscopy in the Asymptomatic Patient

Antonello Trecca, Giuseppe Cerno, Pasquale Trecca,
Fabio Gaj and Gabriele Marinozzi

Background

In the field of modern gastroenterology, ileoscopy has opened up new frontiers in the diagnosis of intestinal diseases [1]. Exploration of the terminal ileum during total colonoscopy has gained much greater acceptance among endoscopists, based on its diagnostic accuracy in patients with intestinal diseases, including neoplasms [2]. Whether a routine terminal ileoscopy should be mandatory when performing a screening colonoscopy remains to be assessed. However, many authors seem to agree that, at the very least, an ileoscopy should always be attempted, because it allows an accurate diagnosis even in asymptomatic patients [3]. Here we present a clinical case in which ileoscopy enabled the definitive treatment of a rare tumor located in the terminal ileum.

Clinical Presentation

A 53-year-old woman underwent a screening colonoscopy as an outpatient procedure at our department. The terminal ileum was intubated while a total colonoscopy was performed. A sessile, (Paris classification 0-Is), submucosal lesion 5 mm in diameter was found (Fig. 1.1), located 5 cm from the Bauhin valve.

Virtual chromoendoscopy using the FICE (Fujinon Intelligent Chromoendoscopy) technique showed the round pitted pattern of the lesion and its regular vascular morphology (Fig. 1.2). Palpation with biopsy confirmed its elastic consistency and its mobility over the submucosal plane.

The lesion was completely excised with an endoscopic mucosal resection and the iatrogenic ulcer was closed with one resolution endoclip (Figs. 1.3, 1.4). The specimen was fixed and sent to the pathologist. Histology showed a well-differentiated neuroendocrine neoplasia, immunohistochemically positive for chromogranin A, with lateral and vertical free margins (Fig. 1.5), consistent with a diagnosis of grade 1 neuroendocrine tumor. The patient was discharged uneventfully.

After the initial diagnosis, a capsule endoscopy of the small bowel, a CT total body scan, and an octeotride scan were performed to stage the disease, all with negative findings. One year later, a second CT scan confirmed the absence of residual or recurrent disease. The patient is in good general conditions after 18 months of clinical follow-up.

A. Trecca (✉)
Department of Operative Endoscopy
Usi Group Rome, Italy
e-mail: atrecca@alice.it

A. Trecca (ed.), *Atlas of Ileoscopy*,
DOI: 10.1007/978-88-470-5205-5_1, © Springer-Verlag Italia 2013

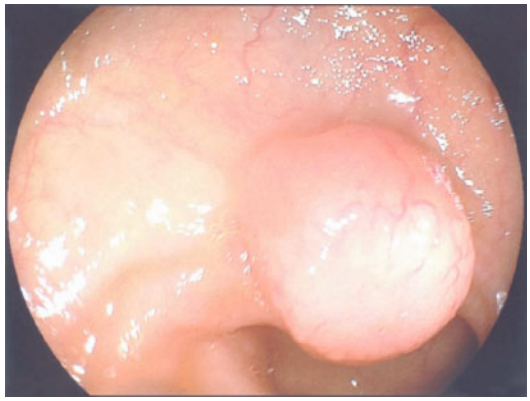

Fig. 1.1 Endoscopic view of a submucosal tumor of the terminal ileum

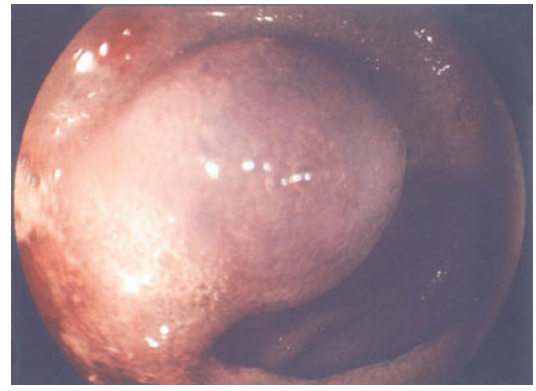

Fig. 1.2 Virtual chromoendoscpy (FICE) shows the normal vascular pattern of the tumor

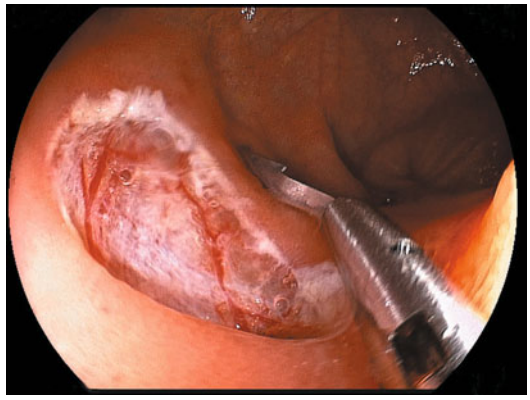

Fig. 1.3 Ulcer scare after endoscopic mucosal resection

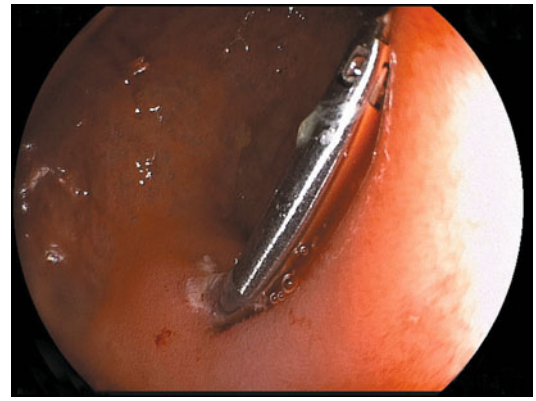

Fig. 1.4 Closure of the defect with a resolution clip

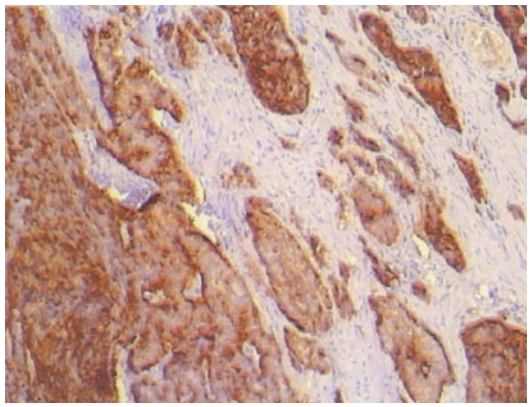

Fig. 1.5 Well differentiated carcinoid tumor, positive for chromogranin, ×200

Open Issues

Our experience demonstrates that a carcinoid of the terminal ileum can be detected even during a screening colonoscopy, underlining the importance of at least attempting an ileoscopy, even in asymptomatic patients.

Endoscopic treatment was carried out during the same session as the routine scheduled colonoscopy. The decision for immediate treatment was based on the small size of the lesion, such that a biopsy might have been insufficient for the definitive diagnosis, mainly due to the submucosal origin of the lesion

[4, 5]. Tissue sampling of the tumor can be performed with different techniques, such as the bite-on-bite (or stacked) biopsy, endoscopic-ultrasound-guided fine-needle aspiration (EUS-FNA) and EUS-guided tru-cut biopsy, but all of them have a low diagnostic accuracy. The elastic consistency of the lesion and its wide mobility over the submucosal plane convinced us to simultaneously treat the lesion, because only the resected specimen allows a definitive histological diagnosis.

In this patient, endoscopic treatment can be considered as definitive, confirmed at histological examination by the clear and free lateral and vertical margins and by the other imaging modalities, which excluded distant disease [6, 7]. Follow-up of the patient was negative and she was disease free at 18 months [8, 9].

References

1. Cherian S, Singh P (2004) Is routine ileoscopy useful?An observational study of procedure times, diagnostic yield and learning curve. Am J Gastroenterol 99:2324-2329
2. Trecca A (2011) Ileoscopy. Technique, diagnosis and clinical application. Springer, Milan, pp 13-14
3. Ansari A, Soon SY, Saunders BP et al (2003) A prospective study of the technical feasibility of ileoscopy at colonoscopy. Scand J Gastroenterol 38:1184-1186
4. Pape UF, Berndt U, Müller-Nordhorn J et al (2008) Prognostic factors of long-term outcome in gastroenteropancreatic neuroendocrine tumours. Endocr Relat Cancer 15:1083-1097
5. Van Tuyl SAC, van Noorden JT, Timmer J et al (2006) Detection of small-bowel neuroendocrine tumors by video-capsule endoscopy. Gastrointest Endosc 64:66-72
6. Johanssen S, Boivin M, Lochs H, et al (2006) The yield of wireless capsule endoscopy in the detection of neuroendocrine tumors in comparison with CT enteroclysis. Gastrointest Endosc 63:660-665
7. Ahlman H, Wangberg B, Jansson S et al (2000) Interventional treatment of gastrointestinal neuroendocrine tumours. Digestion 62(suppl 1):59-68
8. Goede AC, Winslet MC. (2003) Surgery for carcinoid tumours of the lower gastrointestinal tract. Colorectal Dis 5:123-128
9. Sutton R, Doran HE, Williams EMI et al (2003) Surgery for midgut carcinoid. Endocr Relat Cancer 10:469-481

Multiple Choice Questionnaire

1) Which is/are the absolute indication(s) to ileoscopy?

a. chronic diarrhea
b. right lower quadrant abdominal pain
c. abnormal imaging finding
d. all of the above

2) Which of the following serological tests helps to diagnose a carcinoid syndrome?

a. raised transaminases
b. leukocytosis
c. high levels of chromogranin A and 5-hydroxyindoleacetic acid
d. raised lipase and amylase
e. raised γ-glutamyl transpeptidase and alkaline phosphatase

3) What is the 5-year survival percentage for patients with all forms of gastrointestinal carcinoids?

a. 20%
b. 40%
c. 50%
d. 70%
e. 90%

1.d – 2.c – 3.d

The Symptomatic Patient

Case 2
Isolated Polypoid Primary Lymphangiectasia of the Terminal Ileum

Federico Iacopini, Patrizia Rigato, Emma Calabrese and Agostino Scozzarro

Background

Intestinal lymphangiectasia is a rare condition of impaired lymphatic flow. Several forms are recognized: primary or congenital malformation (Waldmann's disease) [1], or secondary to a related localized obstructing pathology [2, 3]. The elevated pressure of the lymphatic drainage in the intestinal wall results in the leakage of lymphatic fluid and the manifestation of an exudative enteropathy with hypoproteinemia, peripheral edema, low serum gammaglobulin and/or lymphocytopenia. However, asymptomatic cases also have been described [4].

Clinical Presentation

A 67-year-old man underwent colonoscopy due to a positive fecal occult blood test. He was asymptomatic, with arterial hypertension as his only comorbidity. The family history was unremarkable. The physical examination was negative and the laboratory examination showed a normal complete blood count and ferritin.

Colonoscopy findings were normal as well, but on ileoscopy a hard-elastic 5-mm pseudo-pedunculated polyp (Paris classification type 0-Isp) was seen located 3 cm above the ileo-cecal valve, with a small erosion (Fig. 2.1). Chromoendoscopy with indigo-carmine (0.4%) evidenced a roundish and small, tubular pit pattern (type IIIs by Kudo) (Fig. 2.2) and no adjacent subtle lesions (Fig. 2.3). No further lesions were found in the ileum explored for 30 cm proximally.

The ileal polyp was resected during the same procedure by endoscopic mucosal resection after it was elevated by the injection of normal saline into the submucosa (Fig. 2.4). Microscopic examination demonstrated dilated mucosal and submucosal lymphatic vessels with polyclonal normal plasma cells, shortened and widened villi, and marked edema of the lamina propria, compatible with intestinal lymphangiectasia (Fig. 2.5). No acute or chronic inflammation was observed in biopsy specimens from the colon.

Subsequent esophagogastroduodenoscopy (EGD), abdominal MRI, and small intestine contrast ultrasonography (SICUS) (Fig. 2.6) were negative. Total protein, albumin, gamma globulin, creatinin, electrolytes, and C-reactive protein were normal. A stool sample was negative for steatorrhea and α1-antitrypsin was normal.

F. Iacopini (✉)
Gastroenterology and Endoscopy Unit
S. Giuseppe Hospital
Albano L., Rome, Italy
e-mail. federico.iacopini@gmail.com

A. Trecca (ed.), *Atlas of Ileoscopy*,
DOI: 10.1007/978-88-470-5205-5_2, © Springer-Verlag Italia 2013

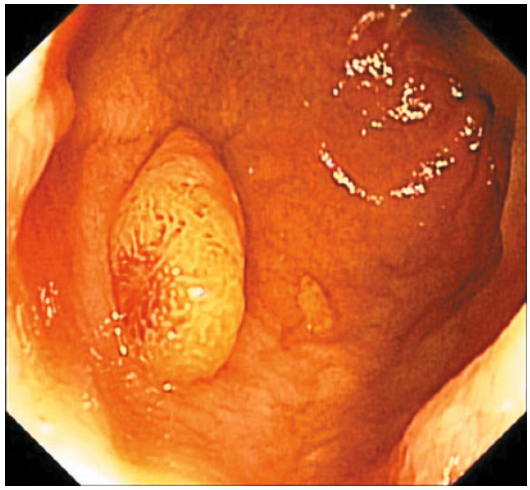

Fig. 2.1 Isolated polypoid primary lymphangiectasia of the terminal ileum

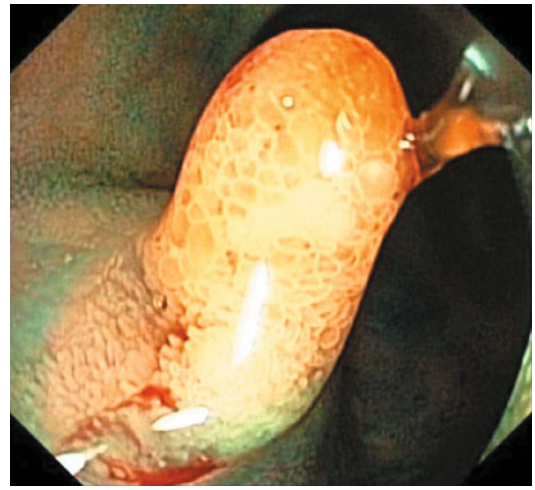

Fig. 2.2 Close-up view of the polypoid intestinal lymphangiectasia: roundish and small, tubular (type IIIs) pit pattern, and small mucosal erosion

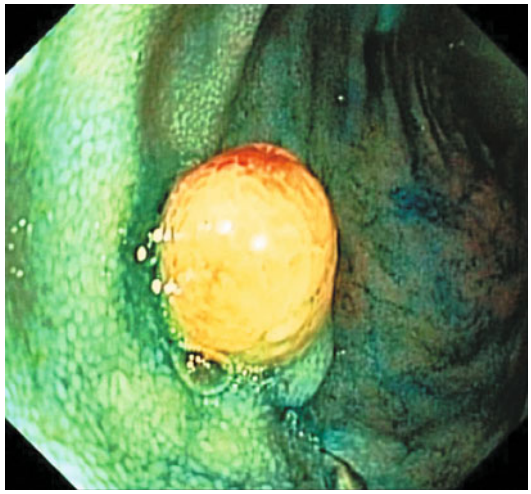

Fig. 2.3 Chromoendoscopy (indigo-carmine 0.4%) shows no subtle lesions adjacent to the polyp and normal villi

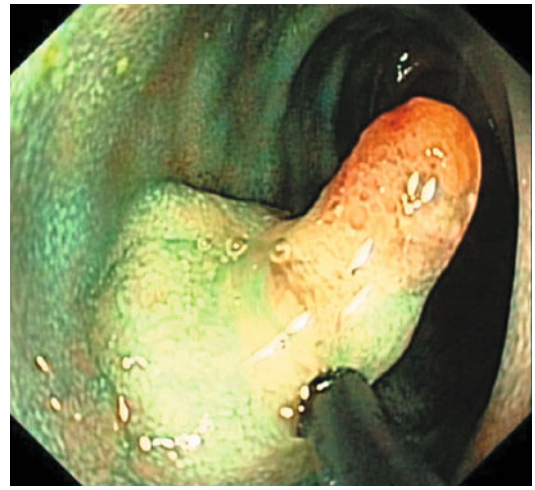

Fig. 2.4 Submucosal lifting of the polypoid intestinal lymphangiectasia before en bloc endoscopic mucosal resection

Isolated primary intestinal lymphangiectasia of the terminal ileum was diagnosed. During the following 36 months, the patient remained asymptomatic. The abdominal ultrasound was negative for lymphoadenopathy and/or masses.

Open Issues

The standard endoscopic features of intestinal lymphangiectasia are: (1) multiple scattered pinpoint white spots, (2) diffuse prominent villi plus whitish-discolored tips, and (3) focal small whitish macules or nodules [2, 5].

This is a rare case of an asymptomatic isolated polypoid primary intestinal lymphangiectasia of the terminal ileum. Thus far, it has been observed only in two previous cases, one primary [6] and the other secondary [7]. The pattern is considered as indicative of an advanced stage of the disorder, with an increased risk of lymphoma.

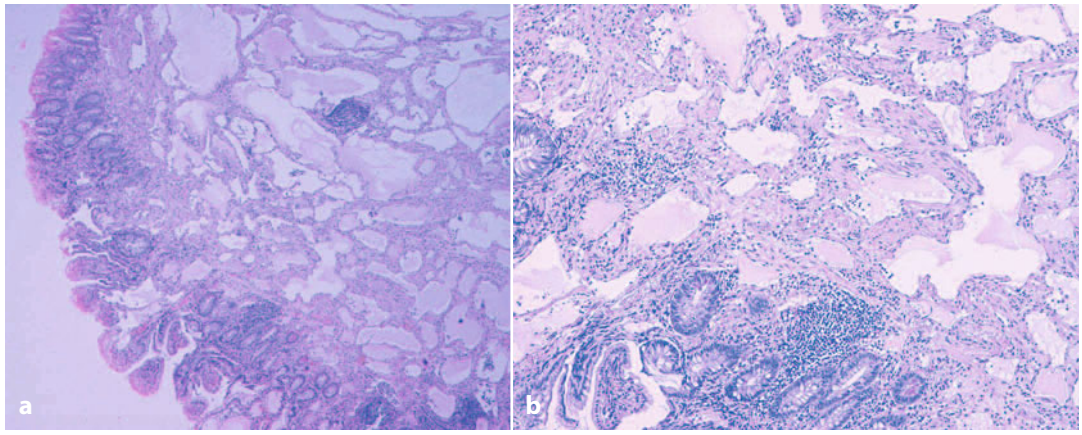

Fig. 2.5 Histological view of ileal lymphangiectasia: dilated mucosal and submucosal lymphatic vessels with cystic dilatations (lactocele); plump villi; surrounding lipid-rich macrophages (hematoxylin and eosin): **a** low-power view; **b** high-power view

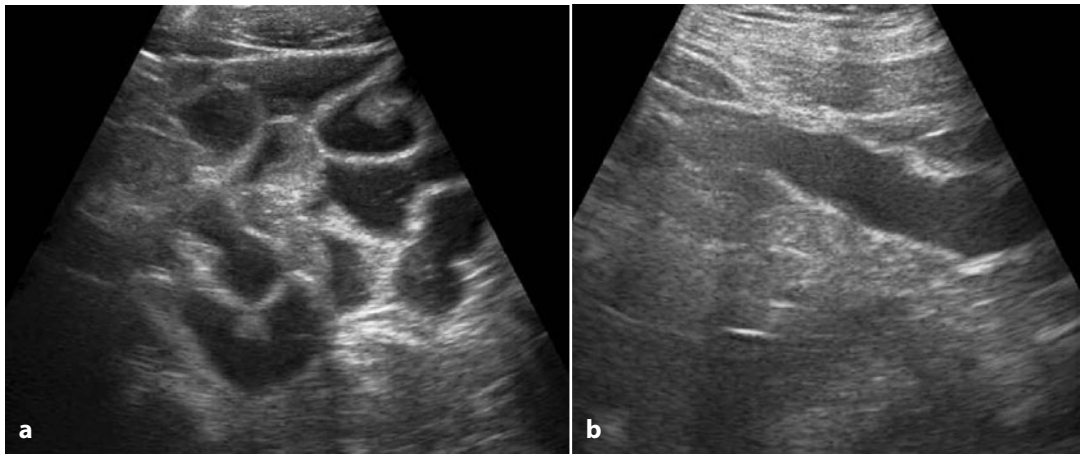

Fig. 2.6 Small intestine contrast ultrasonography (SICUS): **a** normal appearance of the ileal loops; **b** normal (2 mm) bowel wall at the terminal ileum

Features of intestinal lymphangiectasia are mainly visible in the duodenum. EGD, with the corresponding histology of intestinal biopsy specimens, is the main diagnostic approach. However, MRI, CT, or a dynamic examination by SICUS [8] and deep retrograde ileoscopy [9] are necessary to assess multifocality and to exclude secondary intestinal lymphangiectasia.

References

1. Waldmann TA, Steinfeld JL, Dutcher TF et al (1961) The role of the gastrointestinal system in "idiopathic hypoproteinemia". Gastroenterology 41:197-207
2. Kim JH, Bak YT, Kim JS et al (2009) Clinical significance of duodenal lymphangiectasia incidentally found during routine upper gastrointestinal endoscopy. Endoscopy 41:510-5
3. Biyikoglu I, Babali A, Cakal B et al (2009) Do scattered white spots in the duodenum mark a specific gastrointestinal pathology? J Dig Dis 10:300-4
4. Bellutti M, Monkemuller K, Fry LC et al (2007) Characterization of yellow plaques found in the small bowel during double-balloon enteroscopy. Endoscopy 39:1059-63
5. Salomons HA, Kramer P, Nikulasson S et al (1995) Endoscopic features of long-standing primary intestinal lymphangiectasia. Gastrointest Endosc 41:516-8

6. Hirano A, Matsumoto T, Esaki M et al (2010) Intestinal lymphangiectasia presenting with duodeno-jejunal polyposis: enteroscopic findings. Endoscopy 42 (Suppl) 2:E281-2
7. Safatle-Ribeiro AV, Iriya K, Couto DS et al (2008) Secondary lymphangiectasia of the small bowel: utility of double balloon enteroscopy for diagnosis and management. Dig Dis 26:383-6
8. Calabrese E, Zorzi F, Pallone F (2012) Ultrasound in Crohns Disease. Curr Drug Targets 13:1224-33
9. Iacopini G, Frontespezi S, Vitale MA et al (2006) Routine ileoscopy at colonoscopy: a prospective evaluation of learning curve and skill-keeping line. Gastrointest Endosc 63:250-6

Multiple Choice Questionnaire

1) Intestinal lymphangiectasia is
a. characterized by dilated lymphatic ducts in the bowel wall
b. associated with hypoproteinemia, edema, hypogammaglobulin, lymphocytopenia
c. primary or secondary
d. mainly diagnosed by endoscopy and histology
e. all of the above

2) Secondary forms of intestinal lymphangiectasia are
a. associated with systemic disorders
b. characterized by diffuse small bowel involvement
c. differentiated according to the endoscopic features
d. diagnosed by CT scan or MRI
e. related to advanced abdominal cancer

3) Endoscopic features of intestinal lymphangiectasia include
a. multiple scattered pinpoint white spots
b. diffuse prominent villi with whitish-discolored tips
c. focal small whitish macules or nodules
d. segmental or diffuse
e. all of the above

4) Intestinal lymphangiectasia is
a. characterized by exudative enteropathy but can be asymptomatic
b. treated with a low-fat diet with medium chain tryglicerides and vitamin supplementation
c. associated with a very low risk of lymphoma
d. all of the above
e. requires surgery for a definitive resolution

1.e – 2.d – 3.e – 4.d

Case 3
A Case of Unrecognized Complicated Celiac Disease

Riccardo Urgesi, Manuela Marzo, Cono Casale and Italo de Vitis

Background

Celiac disease (CD) is a genetic autoimmune disease in which affected individuals are unable to tolerate foods containing gluten, a specific protein of wheat, barley, and other cereals. The abnormal immune response in the intestine triggered by gluten generates a chronic inflammation and results in tissue damage to the small intestine, with the disappearance of the intestinal villi. If CD is not promptly diagnosed and treated, important and, in some cases, irreversible complications may occur [1]. In the most severe manifestations, CD can promote tumor development in the small bowel, such as intestinal lymphoma or adenocarcinoma. Especially in the initial phase of CD, the differential diagnosis will include lymphoma and other diseases [2]. Since the symptoms and endoscopic picture may be similar and non-specific, the diagnosis of CD is often challenging.

Clinical Presentation

In January 2007, a 53-year-old female without a previous medical history underwent a series of medical examinations to determine the cause of leg edema. The tests did not detect any evidence of cardiovascular disease but abdominal CT showed an "...important thickening of the third portion of the duodenum and the first jejunal loops with associated mesenteric lymphadenopathy (from 8 to 35 mm)...". She suffered weight loss (about 10 kg in 3 months), nausea, loss of appetite, and vomiting and was therefore admitted to the Department of Oncology. Blood tests including antibodies to CD and total immunoglobulins were normal. Ileocolonoscopy documented a diffusely edematous and fragile mucosa with large superficial ulcers covered with fibrin and involving only the terminal ileum. A diagnosis of Crohn's disease was then proposed. An entero-CT showed dilation of the proximal duodenum with obvious reduction of the caliber of the lumen at the level of the III duodenal segment. The involved duodenal walls were slightly thickened, irregular, and hyperdense until the duodenal-jejunal junction. The same pattern was evident at the first jejunal loops, where the wall thickness was about 7 mm, with mesenteric hyperdensity and multiple enlarged lymph nodes, (maximum diameter 3 cm) (Fig. 3.1). In January 2008, a push-and-pull enteroscopy confirmed these findings (Fig. 3.2). Histological examination was not definitive for Crohn's disease nor was it able to differentiate CD from autoimmune enteritis. Given the difficulty of

I. De Vitis (✉)
Unit of Gastroenterology & Internal Medicine
Catholic University-Columbus, Rome, Italy
E-mail: italodev@tin.it

A. Trecca (ed.), *Atlas of Ileoscopy*,
DOI: 10.1007/978-88-470-5205-5_3, © Springer-Verlag Italia 2013

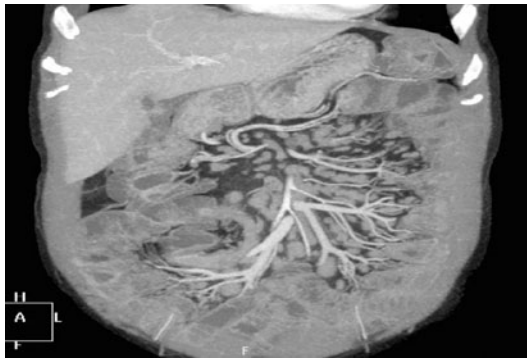

Fig. 3.1 Entero-CT showing dilation of the proximal duodenum with reduction of the caliber of the lumen at the level of the III duodenal segment

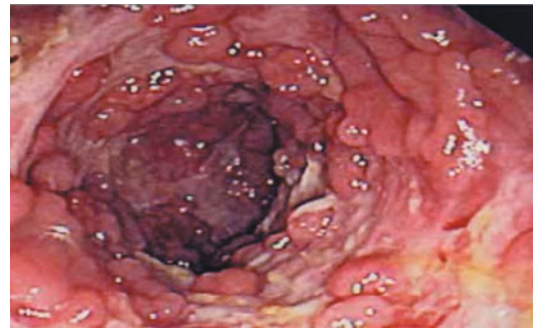

Fig. 3.2 Enteroscopic appearance of the first jejunal loop

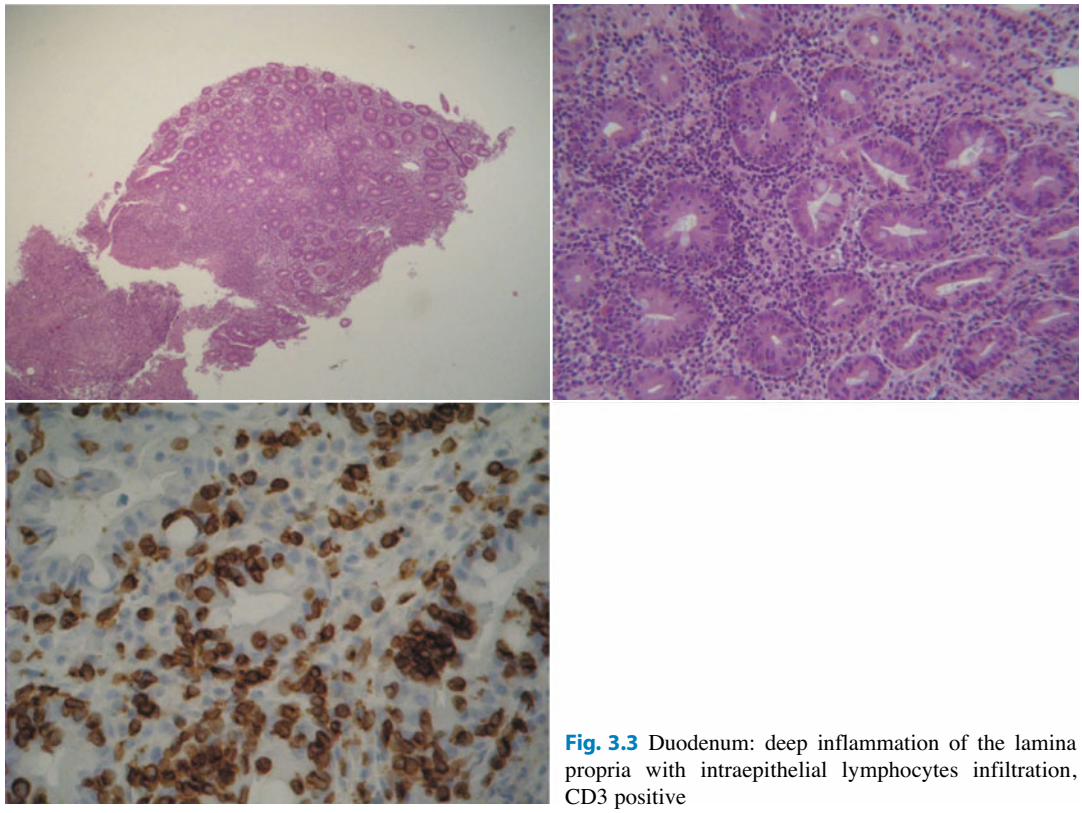

Fig. 3.3 Duodenum: deep inflammation of the lamina propria with intraepithelial lymphocytes infiltration, CD3 positive

this case the histological samples were sent to another reference center, which subsequently provided the following response: "Morphological aspect indicative of jejunal-ileitis with ulcerative negativity to CD8 and positivity for CD3 antigen, suggestive of a state of refractoriness strongly suspicious of a possible lymphomatous evolution" [3] (Figs. 3.3, 3.4). The patient also underwent DQ2/DQ8 haplotyping (DQ2 +), a bone marrow biopsy, lymphocyte subpopulation analysis, and an exploratory laparoscopy with lymph node biopsies, all not significant; and a CEUS (ultrasound with contrast medium) and diagnostic enteroclysis,

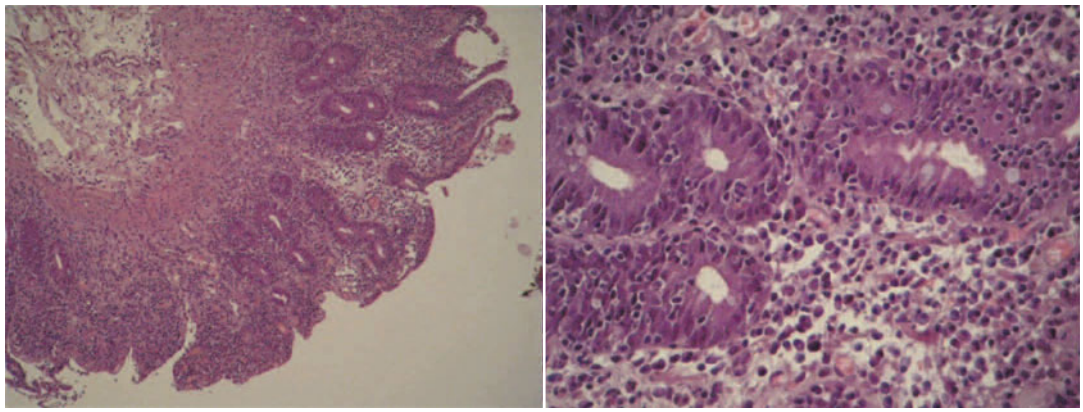

Fig. 3.4 Jejunum: histological view

both compatible with suspected ileal Crohn's disease with skip jejunal lesions; however "*…the appearance did not allow the exclusion of a possible jejunitis ulcerative.*" Based on the progressive deterioration of the woman's general condition and the inability to reach a final differential diagnosis between inflammatory bowel disease and suspected ulcerative jejunal ileitis in refractory celiac disease (or in lymphomatous transformation), the patient started steroid therapy and was placed on a gluten-free diet. After an initial clinical improvement followed by the discontinuation of steroid therapy while maintaining the strict gluten-free diet, there was a further significant worsening of symptoms. In June 2008, the patient was again hospitalized during which time her son was diagnosed with CD. She again underwent enteroscopy, with histology showing evidence of refractory CD with a lymphomatous evolution. Specific chemotherapy was then started, but her general condition worsened. The patient died due to gastrointestinal bleeding, impossible to treat.

Open Issues

The notable features of this case are that the clinical, radiological, and endoscopic features at the onset were all typical for Crohn's disease, particularly aphthoid ulcer, cobblestone pattern, and the deep mucosal lesions observed during push enteroscopy. Only the histological pattern was dubious for refractory CD. But the patient's non-responsiveness to steroid therapy and to a gluten-free diet should alert the physician to the likely evolution of the disease to intestinal lymphoma. This case shows that different diseases may have a common endoscopic picture and that T cell lymphoma can be synchronous with the onset of CD [4].

Sometimes the obvious diagnosis is not the correct one!

References

1. Holmes GK, Prior P et al (1989) Malignancy in coeliac disease: effect of a gluten free diet. Gut 30:333-338
2. Corrao G, Corazza GR, Bagnardi V et al (2001) Club del Tenue study Group. Mortality in patients with coeliac disease and their relatives:a cohort study. Lancet 358:356-61
3. De Mascarel A, Belleanne'e G, Stanislas S et al (2008) Mucosal intraepithelial T- lymphocytes in refractory celiac disease: a neoplastic population with a variable CD8 phenotype. Am J Surg Pathol 32:744-51
4. Cellier C, Delabesse E, Helmer C and French Coeliac Disease Study Group (2000) Refractory spurie, coeliac disease and enteropathy-associated T-cell lymphoma. Lancet 356:203-208

Multiple Choice Questionnaire

1) Celiac disease is

a. an allergy
b. a temporary condition typical of childhood
c. a syndrome characterized by damage to the small intestinal mucosa caused by gliadin
d. an infectious disease
e. a malabsorption condition related to the pancreas

2) The most frequent complication of celiac disease is

a. enteropathy-associated T-cell lymphoma
b. refractory sprue
c. ulcerative jejunitis
d. all of the above
e. none of the above

3) The only current treatment for celiac disease is

a. immunosuppressant drugs
b. a vaccine
c. a lifelong, strict, gluten-free diet
d. a zonulin inhibitor
e. antibodies against IL-15

4) The enteropathy-associated T-cell lymphoma is

a. MALT lymphoma
b. burkitt-like lymphoma
c. T cell lymphoma
d. immunoproliferative small intestinal disease (IPSID)
e. gastrointestinal stromal tumor

1.c – 2.d – 3.c – 4.c