

We are IntechOpen, the world's leading publisher of Open Access books Built by scientists, for scientists

6,900

Open access books available

186,000

International authors and editors

200M

Downloads

Our authors are among the

154

Countries delivered to

TOP 1%

most cited scientists

12.2%

Contributors from top 500 universities



WEB OF SCIENCE™

Selection of our books indexed in the Book Citation Index
in Web of Science™ Core Collection (BKCI)

Interested in publishing with us?
Contact book.department@intechopen.com

Numbers displayed above are based on latest data collected.
For more information visit www.intechopen.com



Appendiceal MALT Lymphoma in Childhood – Presentation and Evolution

Antonio Marte^{1,*}, Gianpaolo Marte², Lucia Pintozzi¹ and Pio Parmeggiani¹

¹*Pediatric Surgery, 2nd University of Naples, Naples*

²*General Surgery, 2nd University of Naples, Naples
Italy*

1. Introduction

Lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma) was first described by Isaacson et al. in 1983 (Isaacson & Wright, 1984). According to the WHO lymphoma classification, the indolent B cell lymphoma of MALT type is classified as a marginal zone lymphoma, thus called because it originates from the B lymphocytes normally present in a distinct anatomical location (marginal zone) of the secondary lymphoid follicles (Harris et al., 2001). MALT lymphomas comprise up to 40% of adult non-Hodgkin lymphomas (NHL); the median age at occurrence is 60 years, with a female predominance (Anonymous, 1997). In paediatric age MALT lymphomas are very rare. We report on a case of MALT lymphoma involving the appendix in a 6-year-old immunocompetent girl and its evolution toward an inflammatory bowel disease (IBD) at a middle-term follow-up.

2. Case report

P.A., a 6-year-old girl, was referred to our institution in May 2005 with a diagnosis of appendicitis. The girl had been complaining of right lower abdominal pain for 6 months. More recently, the pain was exacerbated by walking and coughing. Abdominal ultrasound showed a slight effusion of the pelvic fossa. Her postnatal history showed some period of constipation spaced by regular daily evacuations. Blood examinations showed neutrophil leucocytosis. The patient underwent laparoscopic appendectomy using the three-trocar technique, three endo-loops and the Liga-Sure for the hemostasis. During the laparoscopic exploration no hyperplastic mesenteric lymphnode was found. The appendix appeared moderately hyperemic with a slight enlargement of two-thirds of its distal portion (Fig. 1).

The postoperative course was uneventful and the girl was discharged on day 1, without any complications. The appendix underwent a routine histological examination. The morphological appearance showed thickened lamina propria and submucosa, which were occupied by pseudonodules of immunocompetent cells (Fig. 2), characterized by lymphocytes with small nuclei with a narrow cytoplasmic rim and plasma cell (Fig. 3). Immunohistochemical studies revealed positivity for CD20 (CD20, pan B cell), and

* Corresponding Author

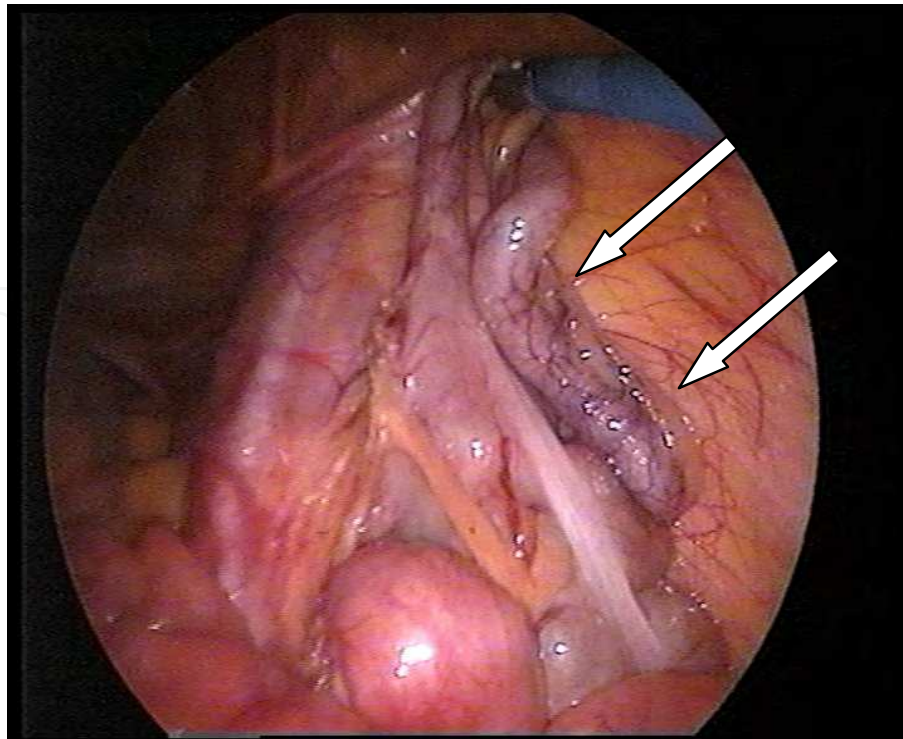


Fig. 1.

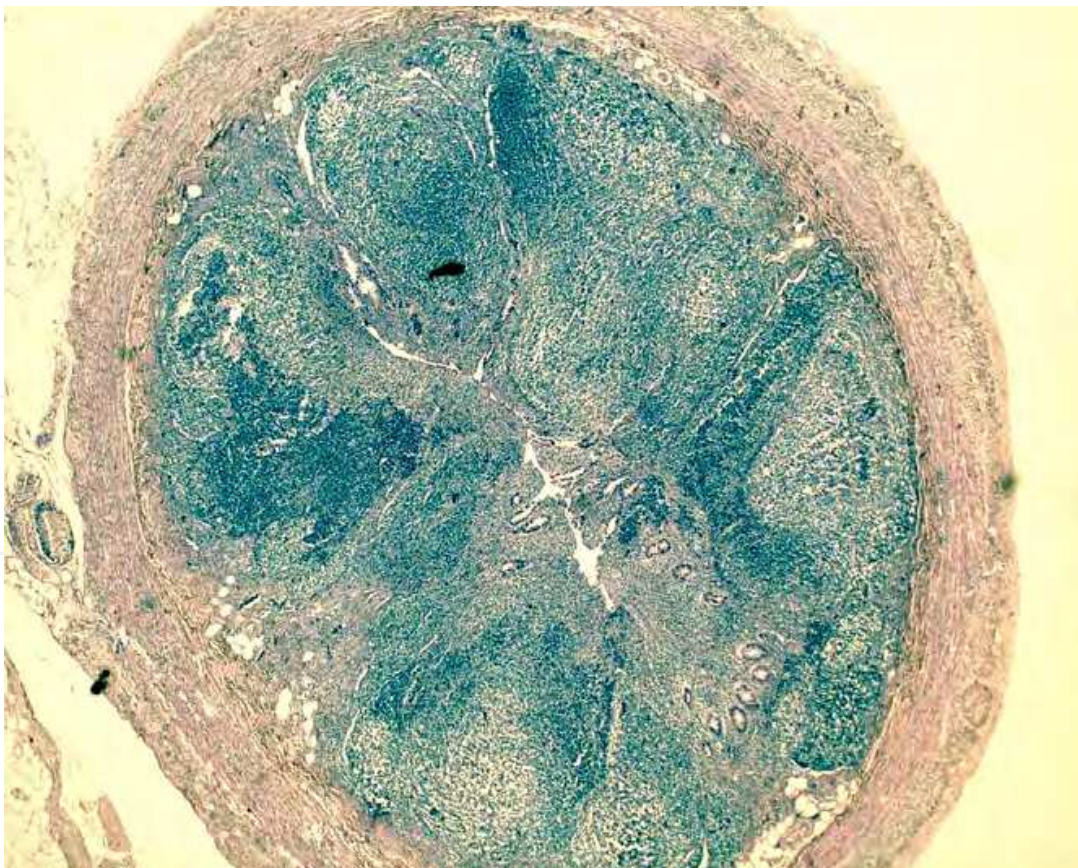


Fig. 2. Hematoxylin-eosin staining. The wall of appendix is thickened, and occupied by a compact nodular formation. The serous and muscular tunic appear thin.

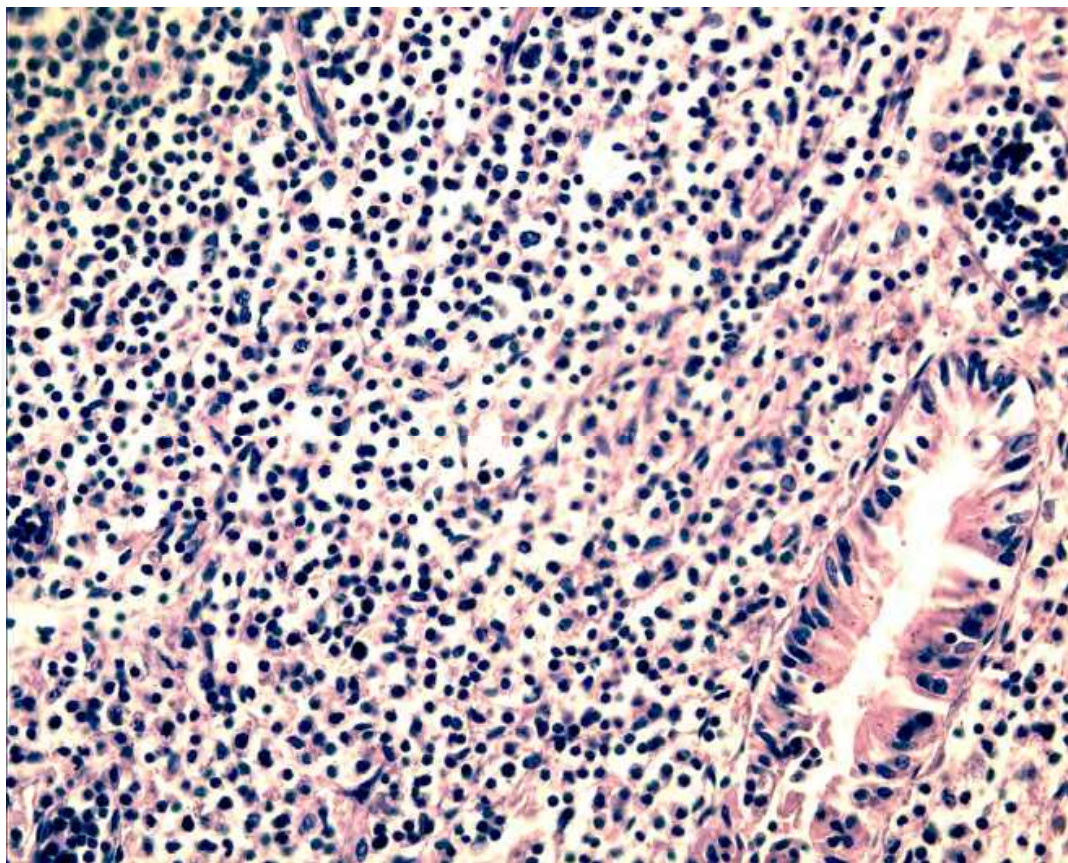


Fig. 3. Hematoxylin–eosin staining. Hyperdense lymphocytic population with small nuclei and narrow cytoplasmic rim and some plasmacytoid and monocytoid elements with great and hyperchromatic nuclei.

negativity for CD5 (CD5, Pan T cell, and B cell subsets) and CD10 using monoclonal antibodies, and positivity for anti-k (immunoglobulin light chain) using polyclonal antibodies, in addition to a low positivity to Ki-67 (proliferation-associated marker). Extensive further examination revealed that the lymphoma was restricted to the distal portion of the appendix (stage IA) and was not associated with any specific infection.

Abdominal MRI, OGDS, and capsule endoscopy of the ileum were all negative; the search for *H. pylori* was also negative. No chemotherapy was performed. After a 15-months follow-up, the patient was doing well (Marte et al., 2008). Calprotectin and clinical evaluation were repeated yearly showing no problem and the patient was asymptomatic. 3 year after, the yearly follow-up showed a slight increase of fecal calprotectin values (40µg/g) with recurrent abdominal pain and occasional episodes of diarrhea. The girl underwent a new clinical evaluation, small bowel radiological contrast study, videocapsule, OGDS, and colonoscopic examination. No fever or weight loss was present; erythrocyte sedimentation rate, C-reactive protein level were slightly higher. Perinuclear antineutrophil cytoplasmic antibodies (pANCA) and Anti-Saccharomyces cerevisiae (ASCA) antibodies are not increased too.

The small bowel x-ray contrast, OGDS and videocapsule study demonstrated no abnormalities. Colon biopsies revealed a mild nonspecific IBD extending till 90 cm from the anal verge. (Fig.4,5,6,7)

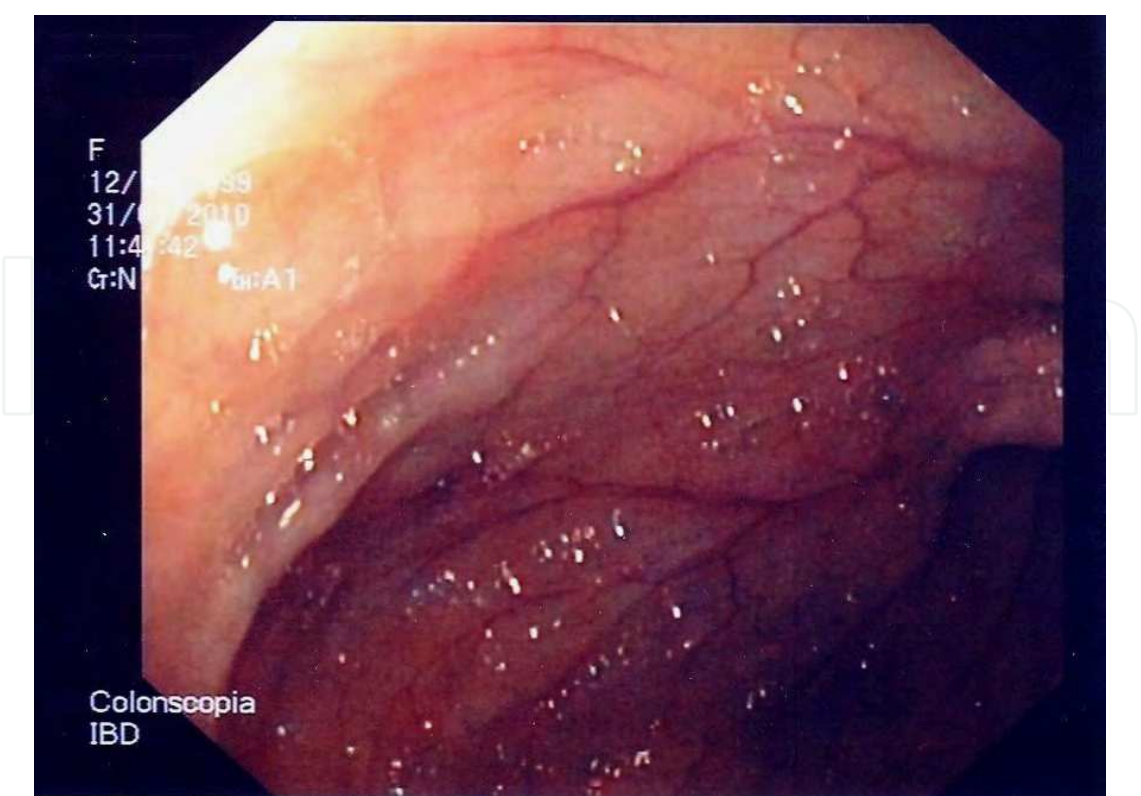


Fig. 4. Colonoscopy. Rectum and sigmoid colon: mucosal redness, nonspecific inflammatory pattern.

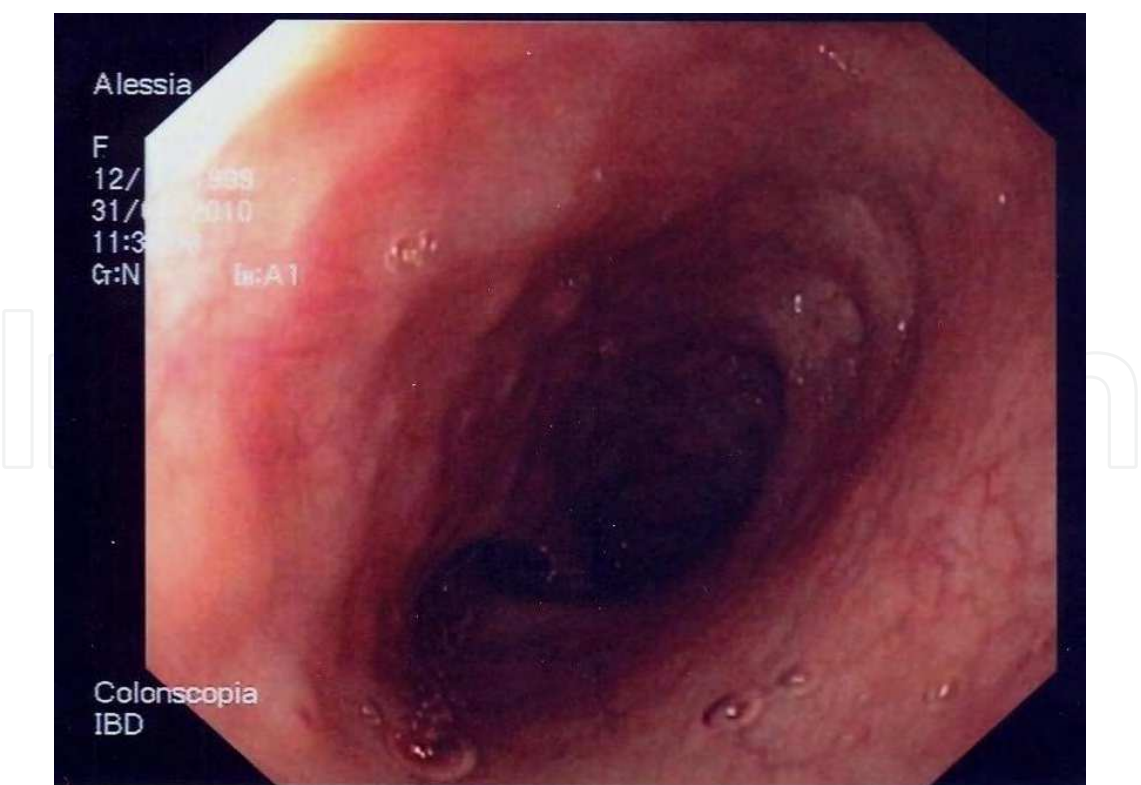


Fig. 5. Colonoscopy. Rectum and sigmoid colon: mucosal redness, nonspecific inflammatory pattern.

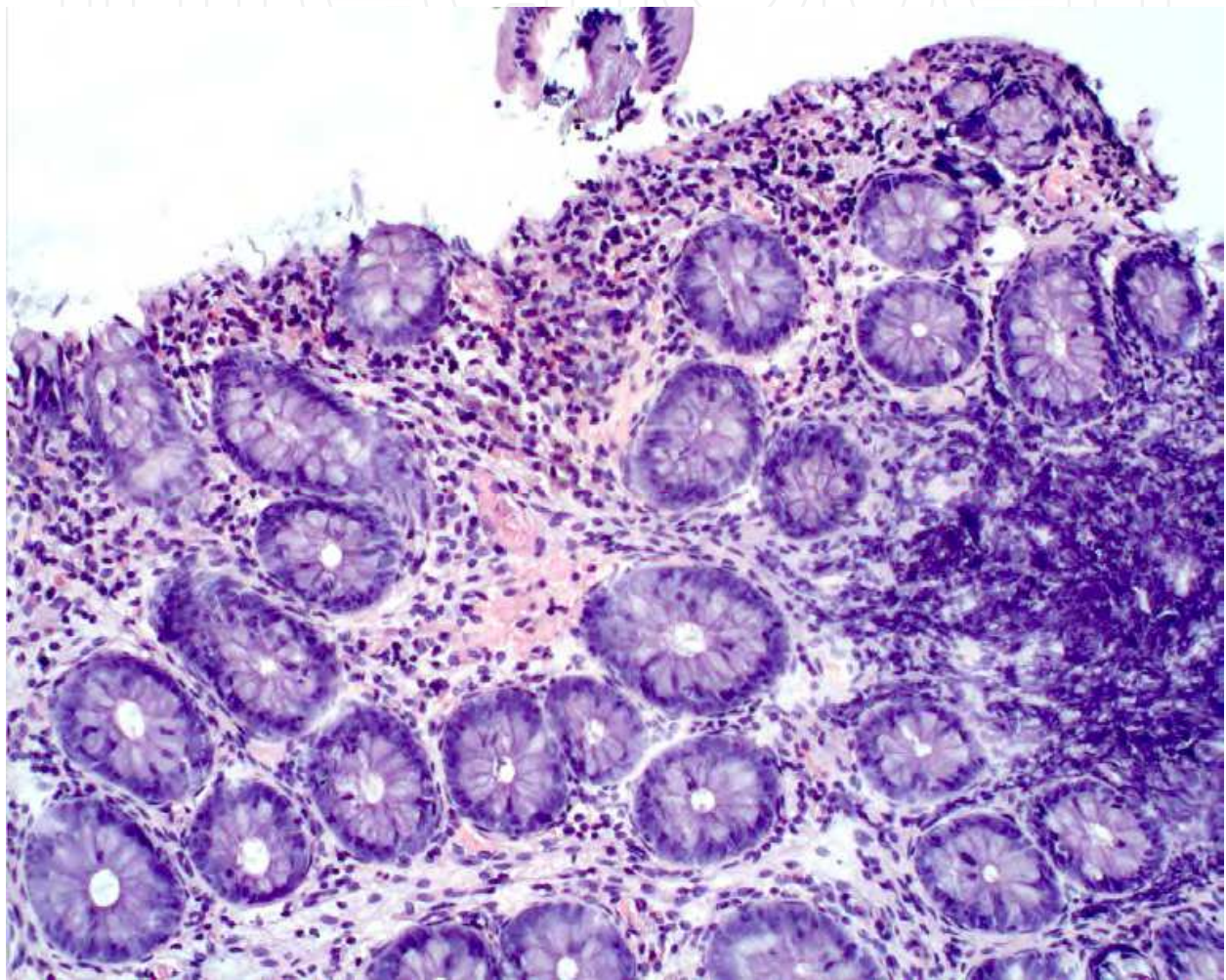


Fig. 6. Absent mucosal surface epithelium with focal reduction of the glands. The lamina propria is edematous, site of microbleeds and is infiltrated by elements of immunocompetent and heaps of eosinophils. (20 X). 90 cm from the anal verge.

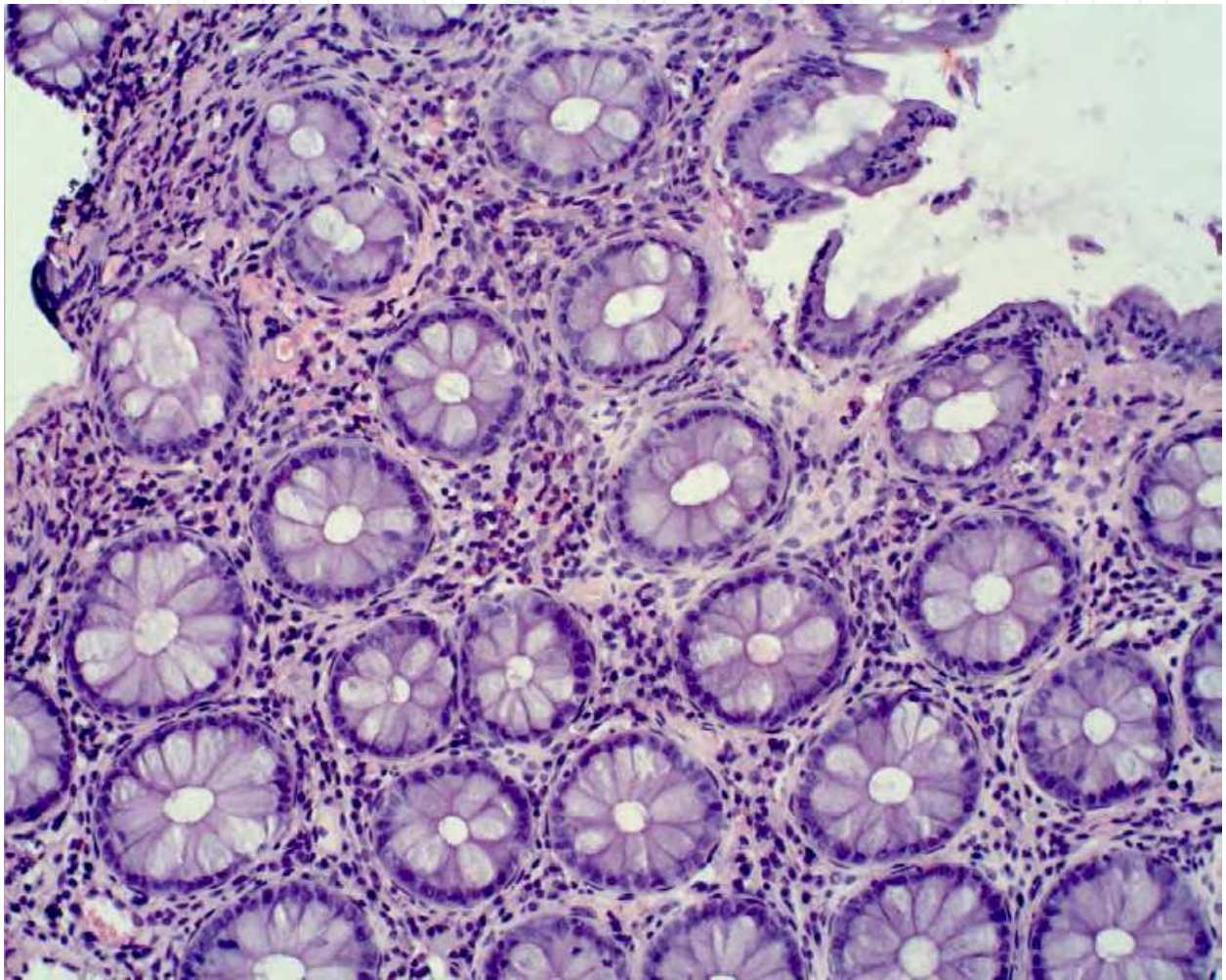


Fig. 7. Rectum (20X): 2 small erosions of the mucosa, glandular patrimony preserved, but low in mucus cells. The lamina propria is diffusely infiltrated by immunocompetent cells (lymphocytes and plasma cells).

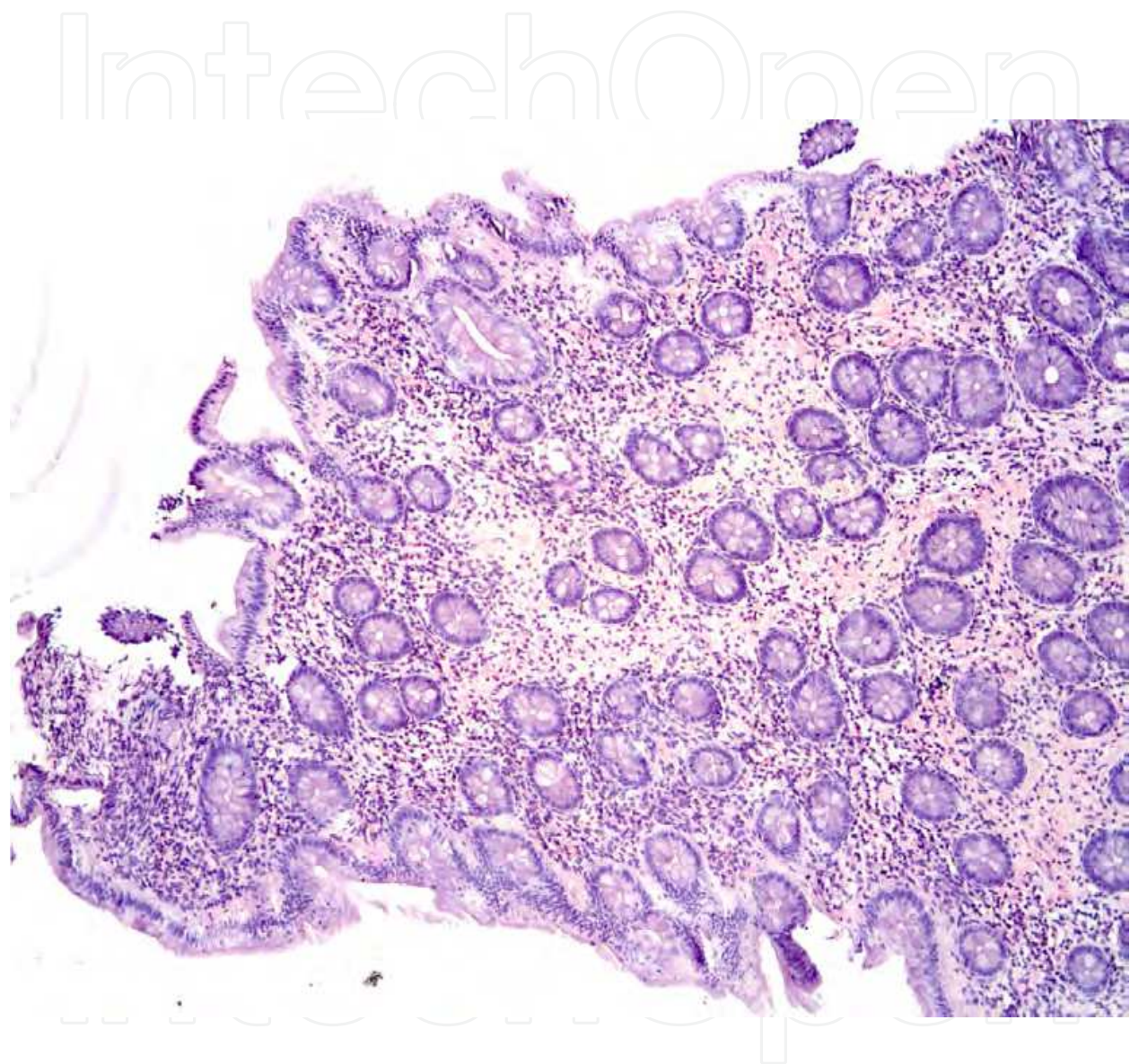


Fig. 8. Recto-sigmoid junction (10X). Mucosal home-based micro-inflammatory polyps. The glandular portion is moderately reduced. The lamina propria is edematous and infiltrated by immunocompetent elements.

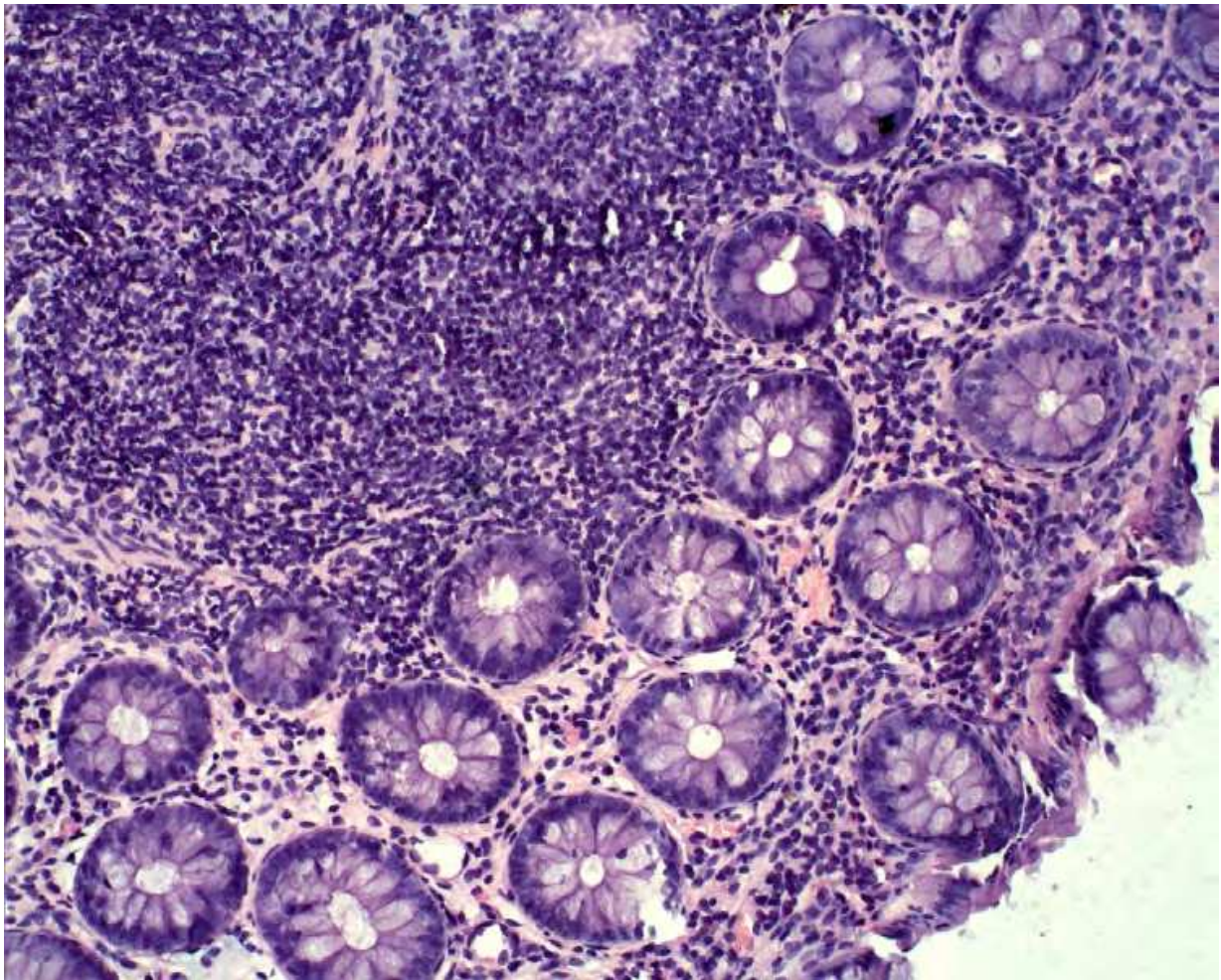


Fig. 9. Mucosal lymphocytes nodule. Glands, well structured, present a reduction of mucus cells. The lamina propria is infiltrated by immunocompetent elements. (20x). 15 cm from the anal verge.

A 6 weeks cycle of Mesalazine (5ASA), 2gr/day was administered to the patient obtaining the induction of remission and then repeated every 2 months for the prevention of recurrences. At present the patient is doing well and a strict clinical serologic follow-up with calprotectin, P-Anca and ASCA is scheduled every 6 months and, yearly, colonoscopy.

3. Discussion

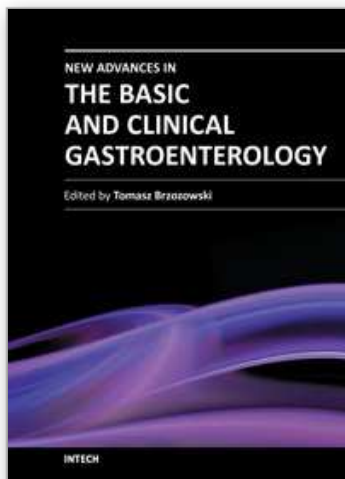
Current knowledge of MALT lymphoma is largely based upon studies in adults. MALT lymphoma is rare in children; the available evidence consists mostly of isolated case reports, except for one series of ten cases (Corr et al., 1997), and another including a total of 48 cases (children and young adults) (Taddesse-Heath et al., 1997) and a pediatric NHL trial recruiting children and adolescents from Germany, Austria and Switzerland (Kaatsch et al., 2004). MALT often develops within the context of a pre-existing inflammatory response due to infection or to autoimmune disorder. Many studies show the relationship between *H. pylori* infection and gastric MALT lymphoma (Isaacson & Whright, 1984; Kurugoglu et al., 2002); some authors have reported a regression of MALT lymphoma in parotid gland (Alkan et al., 1996), lip gland (Berrebi et al., 1998), small intestine (Fischbach et al., 1997) and

rectum (Matsudo et al., 1997) following *H. pylori* eradication. Other risk factors for MALT lymphoma include autoimmune diseases like Hashimoto thyroiditis or Sjogren syndrome, and *Borrelia burgdorferi* for skin lymphoma. A further prerequisite for the development of MALT lymphoma in children may be the presence of HIV infection (Teruya-Feldestein et al., 1995; Mo et al., 2004). In some patients no risk factors can be identified. The most common sites are the stomach and salivary glands. Others sites are: ocular adnexa, the lungs, thyroid and the skin (Zucca et al., 2000). Some retrospective analyses of histopathological results of appendectomy specimens performed for acute appendicitis in a large sample of patients, including children, report a prevalence of appendiceal malignant tumors ranging from 0.4 (Tchana et al., 2006) to 1.5% (Ravi et al., 2006). Among the malignant tumors, carcinoids have the highest incidence (Tchana et al., 2006; Ravi et al., 2006) and 70–90% of these tumors are discovered incidentally because they are usually restricted to the distal appendix (Akerstrom, 1989; Aranha & Greenle, 1980). From a review of the literature we found only one case of appendix lymphoma in paediatric age presenting with intussusception symptoms (Karabulut et al., 2005). Our report probably represents the first case of MALT lymphoma of the appendix found accidentally in a child during an appendectomy. MALT lymphomas manifest with aspecific symptoms. In our case, the clinical presentation was characterized by recurrent abdominal pain, and the only element of suspicion was the enlargement of the distal portion of the appendix. The subsequent evolution to a mild form of IBD could be considered as an evolution of the appendiceal malt-lymphoma for which the phenomenon should be considered a prodromal presentation of a more extensive bowel disease which require a close follow-up and specific therapy (Aomatsu et al., 2011). Otherwise we can't exclude that the subsequent IBD could be an autonomous, subsequent disease considered that, also in this case, there are no data in the Literature. Furthermore, given the previous appendiceal malt-lymphoma, the efficacy of mesalazine alone, without the use of immunosuppressive drugs, can be considered a very favorable factor in our case. In conclusion, even if the occurrence of malignant appendiceal pathology in children is rare (Setty & Termuhlen, 2010), the probability that it is asymptomatic is very high. According to our experience, our case suggests that histological examination should always be performed following appendectomy in children and that if a MALT lymphoma were discovered, a close follow-up is strongly recommended, not only for the MALT lymphoma recurrence but also for its possible evolution towards an inflammatory bowel disease.

4. References

- Akerstrom G (1989). Surgical treatment of patients with the carcinoid syndrome, *Acta Oncol* 28(3):409–414.
- Alkan S, Karcher DS, Newman MA, Cohen P (1996). Regression of salivary gland MALT lymphoma after treatment for *H. pylori*, *Lancet* 348:268–269.
- Anonymous (1997). A clinical evaluation of the international lymphoma study group classification of non Hodgkin's lymphoma. The non Hodgkin's lymphoma classification project, *Blood* 89:3909–3918.
- Aomatsu T, Yoden A, Matsumoto K, Kimura E, Inoue K, Andoh A, Tamai H (2011). Fecal calprotectin is a useful marker for disease activity in pediatric patients with inflammatory bowel disease, *Dig Dis Sci.* Aug;56(8):2372-7.
- Aranha GV, Greenle HB (1980). Surgical management of carcinoid tumors of the gastrointestinal tract, *Am Surg* 46(8):429–435.

- Berrebi D, Lescoeur B, Faye A et al (1998). MALT lymphoma of labial minor salivary gland in an immunocompetent child with a gastric *Helicobacter pylori* infection, *J Pediatr* 133:290–292.
- Corr P, Vaithilingum M, Thejpal R, Jeena P (1997). Paroid MALT lymphoma in HIV infected children, *J Ultrasound Med* 16:615–617.
- Fischbach W, Tacke W, Greiner A et al (1997). Regression of immunoproliferative small intestinal disease after eradication of *H. pylori*, *Lancet* 349:31–32.
- Harris NL, Jaffe ES, Stein H, Vardiman JW (2001). Pathology and genetics of tumors of the haemopoietic and lymphoid tissues, WHO classification of tumors, *International Agency For Research On Cancer Press*, Lyon, pp 157–160.
- Isaacson P, Wright DH (1984). Extranodal malignant lymphoma arising from mucosa associated lymphoid tissue, *Cancer* 53:2515–2524.
- Kaatsch P, Spix C (2004). *Annual report 2004 (1980–2003)*. German childhood Cancer Registry.
- Karabulut R, Sonmez K, Turkyilmaz Z, Yilmaz Y, Akyurek N, Basaklar AC, Kale N (2005). Mucosa associated lymphoma tissue lymphoma in the appendix, a lead point for intussusceptions, *J Pediatr Surg* 40(5):872–874.
- Kurugoglu S, Mihmanli I, Celkan T, AkiH, Aksoy H, Korman U (2002). Radiological features in paediatric primary gastric MALT lymphoma and association with *H pylori*, *Pediatr Radiol* 32:82–87.
- Marte A, Sabatino MD, Cautiero P, Accardo M, Romano M, Parmeggiani P (2008). Unexpected finding of laparoscopic appendectomy: appendix MALT lymphoma in children, *Pediatr Surg Int*. Apr;24(4):471–3.
- Matsumoto T, Lida M, Shimizu M (1997). Regression of mucosa associated lymphoid tissue lymphoma of rectum after eradication of *H. pylori*, *Lancet* 350:115–116.
- Mo JQ, Dimashkieh H, Mallery SR et al (2004) MALT lymphoma in children: case report and review of the literature, *Pediatr Dev Pathol* 7:407–413.
- Ravi Marudanayagam, Geraint T Williams, Brian I Rees (2006). Review of the pathological results of 2660 appendicectomy specimens, *J Gastroenterol* 41:745–749.
- Setty BA, Termuhlen AM (2010). Rare pediatric non-Hodgkin lymphoma, *Curr Hematol Malig Rep*. Jul;5(3):163–8.
- Taddesse-Heath L, Pittalunga S, Sorbara L et al (2003). Marginal zone B-cell lymphoma in children and young adults, *Am J Surg Pathol* 27:522–531.
- Tchana SV, Detry O, Polus M, Thiry A, Detroz B, Maweja S, Hamoir E, Defechereux T, Cimbra C, De Roover A, Meurisse M, Honore P (2006). Carcinoid tumor of the appendix: a consecutive serie from 1237 appendectomies, *World J Gastroenterol* 7;12(41):6699–701.
- Teruya-Feldstein J, Temeck BK, Sloas MM et al (1995). Pulmonary malignant lymphoma of mucosa associated lymphoid tissue (MALT) arising in a pediatric HIV-positive patient, *Am J Surg Pathol* 19:357–363.
- Zucca E, Conconi A, Roggero E et al (2000). Non gastric MALT lymphomas: a survey of 369 european patients. The international extranodal lymphoma study group, *Ann Oncol* 11:99.



New Advances in the Basic and Clinical Gastroenterology

Edited by Prof. Tomasz Brzozowski

ISBN 978-953-51-0521-3

Hard cover, 546 pages

Publisher InTech

Published online 18, April, 2012

Published in print edition April, 2012

The purpose of this book was to present the integrative, basic and clinical approaches based on recent developments in the field of gastroenterology. The most important advances in the pathophysiology and treatment of gastrointestinal disorders are discussed including; gastroesophageal reflux disease (GERD), peptic ulcer disease, irritable bowel disease (IBD), NSAIDs-induced gastroenteropathy and pancreatitis. Special focus was addressed to microbial aspects in the gut including recent achievements in the understanding of function of probiotic bacteria, their interaction with gastrointestinal epithelium and usefulness in the treatment of human disorders. We hope that this book will provide relevant new information useful to clinicians and basic scientists as well as to medical students, all looking for new advancements in the field of gastroenterology.

How to reference

In order to correctly reference this scholarly work, feel free to copy and paste the following:

Antonio Marte, Gianpaolo Marte, Lucia Pintozzi and Pio Parmeggiani (2012). Appendiceal MALT Lymphoma in Childhood - Presentation and Evolution, New Advances in the Basic and Clinical Gastroenterology, Prof. Tomasz Brzozowski (Ed.), ISBN: 978-953-51-0521-3, InTech, Available from:
<http://www.intechopen.com/books/new-advances-in-the-basic-and-clinical-gastroenterology/appendiceal-malt-lymphoma-in-childhood-presentation-and-evolution>

INTECH
open science | open minds

InTech Europe

University Campus STeP Ri
Slavka Krautzeka 83/A
51000 Rijeka, Croatia
Phone: +385 (51) 770 447
Fax: +385 (51) 686 166
www.intechopen.com

InTech China

Unit 405, Office Block, Hotel Equatorial Shanghai
No.65, Yan An Road (West), Shanghai, 200040, China
中国上海市延安西路65号上海国际贵都大饭店办公楼405单元
Phone: +86-21-62489820
Fax: +86-21-62489821

© 2012 The Author(s). Licensee IntechOpen. This is an open access article distributed under the terms of the [Creative Commons Attribution 3.0 License](#), which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

IntechOpen

IntechOpen