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Case Report

Ileal mucosa-associated lymphoid tissue lymphoma diagnosed after emergency surgery: A case report and literature review

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ABSTRACT

Introduction and importance: Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue, also called MALT lymphoma, is one of the entities of marginal zone lymphomas. These lymphomas are originated from indolent B-cell lymphomas and involve many organs such as the gastrointestinal tract, salivary gland, skin, lung, thyroid or breast. Ileal MALT lymphoma is relatively rare and clinical symptoms are usually atypical.

Case presentation: We report a case of a 99-year-old man who admitted to the emergency department with increasing and colicky periumbilical pain, vomiting and constipation. Non-contrast-enhanced computed tomography suggested small bowel obstruction due to phytobezoar. Intraoperatively, surgeon discovered the tumor at the site of phytobezoar. Histologically, there was a diffuse infiltration comprised of small to medium sized lymphocytes with monocytoid features. Immunohistochemical result confirmed CD20 positive B-lymphocytes and the Ki-67 proliferation index was 10%. Ileal mucosa-associated lymphoid tissue lymphoma was diagnosed based on histological findings and immunohistochemistry.

Discussion: MALToma of the gastrointestinal tract is related to chronic antigenic, inflammatory bowel disease and malabsorption syndromes. However, the etiology of ileal MALToma is unclear. Moreover, symptom of ileal MALToma is really not typical and overleaped in the context of small intestinal obstruction. It should be differentiated small intestinal MALToma from immunoproliferative small intestinal disease and an alpha heavy chain disease.

Conclusion: Ileal MALT lymphoma remains little known in many previous studies. It is really difficult to preoperatively diagnose. The combination of clinical presentation, postoperative histology and immunohistochemistry contribute to diagnosis and carry out appropriate management.

1. Introduction

Marginal zone lymphomas (MZLs) which are derived from indolent B-cell lymphomas include three different MZL entities: splenic MZL (SMZL), nodal MZL (NMZL) and extranodal marginal zone lymphoma (EMZL) of mucosa-associated lymphoid tissue (MALT) also known as MALT lymphoma according to the latest update of the World Health Organization (WHO) classification [1,2]. Among them, EMZL is the most common entity, comprising approximately 70% of all MZLs [3].

MALT lymphoma, also called MALToma, involves many organs such as gastrointestinal (GI) tract, salivary gland, skin, lung, thyroid or breast [4]. The most common site of GI is the stomach, accounting for 60%–75%, followed by the small intestine, cecum, colon and rectum [5]. Thus, MALT lymphoma of the small bowel is relatively rare compared to MALT lymphoma of the stomach. The clinical presentation of MALToma of GI is poorly specific and vague. The symptoms vary from weight loss, dyspepsia, nausea, vomiting to gastrointestinal bleeding or even spontaneous perforation of gastrointestinal. The optimal diagnosis of MALT lymphoma is based on a combination of the clinical exam, histopathological findings, immunohistochemical markers and even molecular diagnostics [6]. In this article, we report a case of a 99-year-old man who was hospitalized with the symptom of bowel obstruction. Ileal

Abbreviations: MZLs, Marginal zone lymphomas; SMZL, Splenic marginal zone lymphomas; NMZL, Nodal marginal zone lymphomas; EMZL, Extranodal marginal zone lymphoma; MALT, Mucosa-associated lymphoid tissue; GI, Gastrointestinal; CT, Computed tomography.

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MALToma was diagnosed by emergency surgery, histopathology and immunohistochemistry. This case report has been reported in line with the SCARE Criteria [7].

2. Case presentation

A 99-year-old man was admitted to the emergency department with increasing and colicky periumbilical pain, vomiting and constipation for three days. The past medical history of patient included hypertension and atrial fibrillation. The patient had no history of prior abdominal surgery or family history of colonic malignancies. Physical examination showed abdominal distension, severe tenderness of the entire abdomen without guarding or rigidity. Laboratory tests revealed a serum creatinine of 124 μmol/L and a serum urea of 12.7mmol/L. Blood tests, liver function tests and pancreatic enzymes were normal. The patient had elevated C-reactive protein level of 133.48 mg/L.

A plain abdominal X-ray showed several air fluid levels. A non-contrast-enhanced CT was shown dilated loops of jejunum and proximal ileum with a width of 3.4cm and air fluid level in the bowel dilatation. CT images also revealed an intraluminal mass and abnormal wall thickening (approximately 1.5cm) at the site of obstruction. The intraluminal mass was a round, well-defined, heterogeneous with a mottled air. The terminal ileum and large bowel were collapsed (Fig. 1). These CT images suggested small bowel obstruction due to phytobezoar. The doctor explained to the patient in detail and the patient consented to perform the surgery. Afterwards the decision to exploratory laparotomy was performed by experienced gastrointestinal surgeon. The surgery took one hour and our patient did not receive blood transfusions. During the surgery, a tumor at proximal ileum was detected. Surgeon removed phytobezoar and revealed the tumor. The ileum in accompany with the tumor was removed and surgeon performed end-to-end ileal anastomosis. There was no complication during the surgery. The patient recovered well postoperatively and discharged on the 6th postoperative day. There was no recurrence of symptoms at one month postoperative follow-up.

On gross examination, a 25cm part of the proximal ileum contained a 5cm length tumor in the central portion. The ileal mucosa was described as pink, smooth. Tumor lesion was pale yellow tissue extending into the mesentery and hemorrhagic mucosa (Fig. 2). On microscopic examination, the lesion showed infiltration of small atypical cells, which were similar to centrocyte–like lymphocytes. There was a diffuse infiltrate comprised of small to medium sized lymphocytes with monocytoid features and the destruction of gland structures (Fig. 3). Immunohistochemistry showed positivity for CD20 and negative for CD3, CD5, CD10, CD23, CD79a, Bcl-6 and Cyclin D1. The percentage of Ki-67 positive was 10%. With these histological findings and immunohistochemistry, the patient was diagnosed with ileal MALT lymphoma. Consequently, MALToma of the ileum led to stagnation of food and obstruction of the bowel in this patient. The patient was transferred to the oncology department for further treatment.

3. Discussion

The age – standardized incidence rate of small bowel MALT lymphoma remains poorly known in many previous studies as well as in the literature. It was reported that the highest incidence rate of MALT lymphoma was between the age of 50 and 60 years [8]. According to the study of Won JH, the median age of colorectal MALT lymphoma was 62 (range 26–87) in literature reviews published in English between 1993 and 2017 [5]. Our patient was 99 years old and admitted to emergency department due to the symptoms of bowel obstruction. Small bowel obstruction is one of the most common emergency diagnosis in elderly patients, which require acute medical care and emergency surgery. The differential diagnoses for such small intestinal obstruction include adhesions, tumors, herniations or even postoperative adhesions. However, malignant small bowel obstruction, gall stone ileus and hernia incarceration are more frequent etiologies in the elderly [9].

Computed tomography is considered as a step forward in detecting causes of bowel obstruction. In our patient, the estimated glomerular filtration rate was 20.3 ml/min/1.73m^2 which was at high risk for contrast-induced nephropathy [10]. In addition, the age of 99 is considered as increasing the risk of an acute adverse reaction to contrast [11]. Therefore, our patient was indicated to perform non-contrast CT. Millet et al. showed unenhanced computed tomography is a safe and utility method for diagnosis in elderly patients with acute abdominal pain [12].

Endoscopy is considered as a valid tool which has an impact on the diagnosis of MALT lymphoma of the GI. Upper gastrointestinal endoscopy can detect the gastric MALT lymphoma. However, the diagnosis of small bowel MALToma required advanced endoscopy such as video capsule or double balloon enteroscopy**.** According to few cases reported in the literature, endoscopic feature of ileal MALToma is a solitary mass resembling a carcinoma and more common than multiple masses. Although these techniques have the possibility of visualization the small intestine, they have the limitation on obtaining biopsy specimens. Capsule endoscopy, therefore, is useful for determining the stage and for the follow-up [13,14]**.**

Many studies indicate that MALToma of the GI relates to chronic antigenic, inflammatory bowel disease and malabsorption syndromes such as *H. pylori* gastritis or *Campylobacter jejuni* in the proximal small bowel [1,15]. *H. pylori* is known as the etiology of gastric MALT lymphoma. In contrast, other case reports of ileal MALToma indicated no

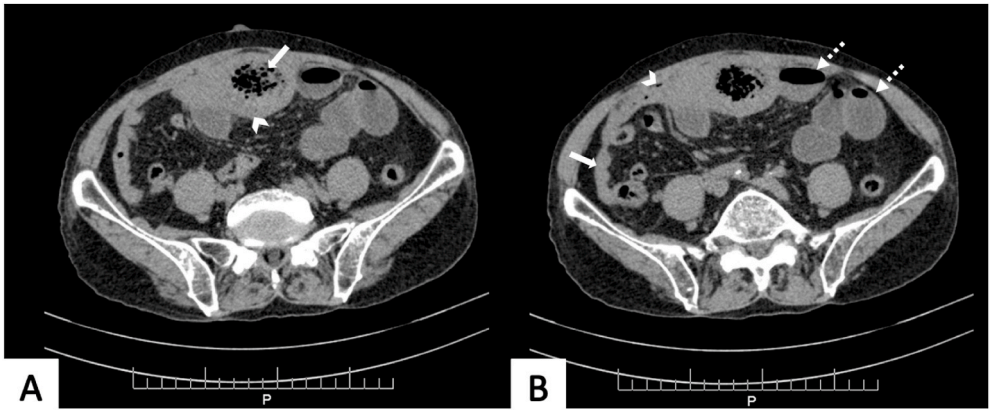

Fig. 1. Non-contrast-enhanced axial computed tomography (CT). A. CT showed an intraluminal round phytobezoar with a mottled gas pattern (arrow) at the site of obstruction, below the umbilical region and abnormal wall thickening (arrow head); B. CT illustrated an abrupt change in caliber at the transition point (arrow head) between distended loops with air – fluid levels (intermittent arrows) and the collapsed large bowel (arrow).

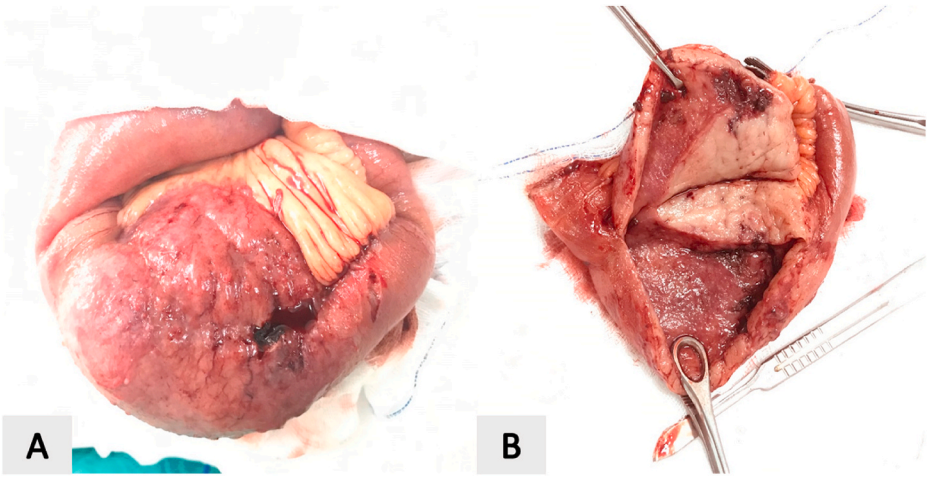

Fig. 2. A. The ileal tumor extending into the mesentery was detected during operation; B. Gross specimen: the ileal mucosa was pink, smooth with a focal area of hemorrhage, and the tumor lesion was pale yellow tissue. (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

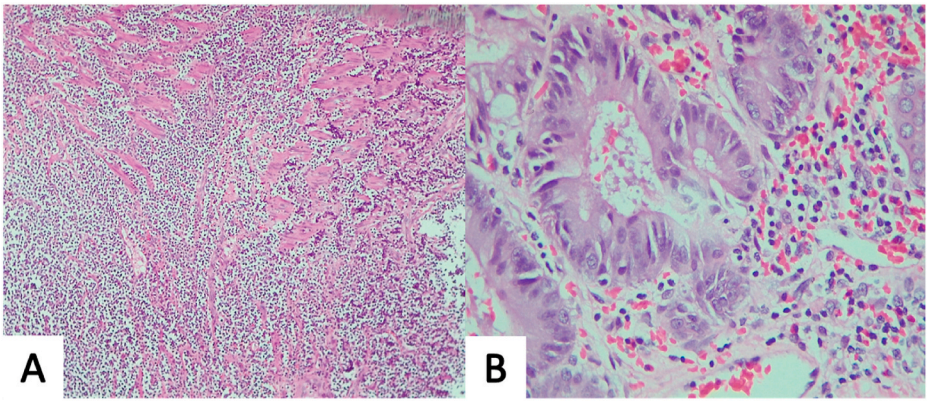

Fig. 3. Histological findings. A. Infiltration of small atypical cells, which were similar to centrocyte – like lymphocytes, is seen; B. There were a diffuse infiltration comprised of small to medium sized lymphocytes with monocytoid features, lymphoplasmacytoid cells and the destruction of gland structures.

presence of *H. pylori*. Thus, the etiology of ileal MALToma remains unknown and hypothesis is related to genetic alterations [16]. The differential diagnosis of MALT lymphoma in small bowel includes immunoproliferative small intestinal disease (associated with Campylobacter jejuni infection) and an alpha heavy chain disease which usually occurs in the proximal intestine [17]. This case report is an unusual case in which MALT lymphoma is found during emergency operation in elderly patient with bowel obstruction, no known history of a chronic inflammatory or autoimmune disease.

Although MALTomas can arise from different anatomical sites, these tumors have the same histopathology characteristics. These are the presence of lymphoepithelial lesions (defined by the infiltration of neoplastic lymphocytes and the destruction of epithelial structures) and sheets of neoplastic small lymphocytes which infiltrate around reactive secondary lymphoid follicles in a marginal zone distribution [18]. In our patient, histopathology result confirmed both the proliferation of small atypical lymphocytes and the infiltration of glands by monocytoid cells.

Immunohistochemical result indicated that the tumor was composed a large of B-cells based on marker CD20 positive. In our case, Cyclin D1 was negative. This helped to exclude mantle cell lymphoma. Similarly, the tumor was different from follicular lymphoma because of the negative Bcl-6 [16]. Ki-67 is defined as a protein expressed in late G1, S, G2 and M-phase and also called as cell proliferation marker. This index was proportional to tumor progression and malignancy. Many studies have indicated that Ki-67 in less than 5% of tumoral cells or the lack of Ki-67 expression had a better prognosis and a longer survival [19,20]. As similar to Ki-67, mitotic index (MI) is considered to reflect the proliferative activity of tumors and used as a prognostic factor. However, mitotic index is not a good prognostic factor in indolent lymphoma [20].

4. Conclusion

Ileal MALT lymphoma is relatively rare compared to others GI MALT lymphomas such as gastric MALToma. The differentiate between ileal MALToma from others tumors of this site, which base on clinical examination and radiology modalities, is really not easy. Histopathological diagnosis and immunohistochemistry are accurate diagnostic tools. This helps to early intervene and plan an optimal treatment after removal tumor such as targeted agents and chemotherapy.

Declaration of competing interest

The authors have no conflicts of interest to declare.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.amsu.2021.102973.

Human and animal rights

Institutions of the authors do not require Ethics committee approval for a case report or case series containing information of fewer than three patients.

Ethical approval

Institutions of authors do not require ethical approval for a case report.

Informed consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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M. D.P. conception and design of the manuscript, data analysis and writing the paper.

M. T.N. data collection and data analysis.

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Guarantor

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