Case Report

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Uncommon presentation of gastric mucosa-associated lymphoid tissue lymphoma in a 13-year-old girl: acute vomiting of blood as the initial symptom

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Introduction and importance: Extranodal marginal zone lymphoma (EMZL lymphoma), also known as mucosa-associated lymphoid tissue (MALT) lymphoma, is a rare B-cell lymphoma that rarely affects children. The involvement of infectious agents, especially *H. pylori*, has been observed in the formation and progression of MALT lymphoma in the stomach. Hematemesis as the primary clinical manifestation is uncommon, highlighting the need for case studies with this presentation. This article uses SCARE2023 criteria as a framework to sort out a case report in order.

Case presentation: A 13-year-old female patient was admitted in August 2022 with an episode of hematemesis. She had a prior diagnosis of anaemia and was found positive for *H. pylori*. Despite treatment, she developed symptoms of chronic non-atrophic gastritis and had recurring episodes of hematemesis. Physical and diagnostic examinations revealed B-cell lymphoma localized in the gastric antrum. The primary diagnosis was extranodal MALT lymphoma with unique plasma cell differentiation.

Clinical discussion: The presentation of gastric MALT lymphoma can be variable, with definitive diagnosis often achieved via endoscopic biopsy. *H. pylori* plays a significant role in the onset and progression of this lymphoma, emphasizing the importance of its eradication for treatment. Effective outcomes can be achieved through anti-*H. pylori* treatment, although it is essential for clinicians to ensure its complete eradication post-treatment.

Conclusion: Paediatric presentation of gastric MALT lymphoma, especially with hematemesis as the primary symptom, is rare and can be easily misdiagnosed. Compared to adults, children generally exhibit a better prognosis with effective H. pylori treatment. It is vital for medical professionals to recognize the differences in presentation between children and adults to ensure accurate diagnosis and treatment.

Keywords: children, infection, Helicobacter pylori, mucosa-associated lymphoid tissue lymphoma

Introduction and importance

Extranodal marginal zone lymphoma (EMZL lymphoma), also known as mucosa-associated lymphoid tissue (MALT) lymphoma, is an indolent B-cell lymphoma typically diagnosed around the median age of 60 years[1]. The occurrence of MALT lymphoma in children is extremely rare. Over the past four decades, both nationally and internationally, only 13 cases of gastric MALT lymphoma in children have been reported. These cases have mainly been observed in school-age and adolescent children[2]. This relatively uncommon low-grade non-Hodgkin lymphoma comprises about 7–8% of all B-cell lymphomas and can manifest in various extranodal sites including the stomach, lung, ocular adnexa, and skin[3].

Certain infectious agents, such as *H. pylori*, play a central role in the development of MALT lymphoma in the stomach[3–5].

HIGHLIGHTS

- Rare paediatric presentation of mucosa-associated lymphoid tissue lymphoma with hematemesis as the primary symptom.
- Association of *H. pylori* in the development and progression of mucosa-associated lymphoid tissue lymphoma emphasized.
- Multi-point biopsy and endoscopic ultrasonography crucial for accurate diagnosis.
- Effective outcomes achieved in children through comprehensive anti-*H. pylori* treatment.
- Importance of post-treatment surveillance to prevent lymphoma relapse or progression.

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Prolonged exposure to *H. pylori* induces the gradual formation of mucosa-associated lymphoid tissue, leading to the activation and rapid proliferation of B cells. Consequently, pathological B lymphocytes evolve into gastric mucosa-associated lymphoid tissue lymphoma. Clinically, MALT lymphoma presents with symptoms such as gastroesophageal reflux disease, upper abdominal pain, discomfort, anorexia, weight loss, or occult gastrointestinal bleeding, which occurs in about 19% of cases. However, this particular case presents a relatively uncommon scenario where overt blood loss, such as hematemesis, serves as the initial symptom. Such a presentation is infrequent in clinical practice, and its non-specific nature often leads to misdiagnosis.

A thorough review of recent literature on MALT lymphoma revealed no cases where hematemesis was the primary clinical manifestation[6]. This report aims to fill that knowledge gap.

Case presentation

A 13-year-old female patient was admitted to our facility in August 2022, presenting with an episode of hematemesis. A review of her medical history indicated that she had been diagnosed with anaemia in November 2021 at another hospital, where her haemoglobin levels were recorded at a low of 51 g/l. Concurrently, She had a positive carbon 13 breath test and a bone biopsy suggested hyperplastic anaemia. In response, she underwent a blood transfusion and was treated with a triple anti-*H. pylori* regimen, consisting of amoxicillin, omeprazole, and clarithromycin. This intervention led to the alleviation of her symptoms. Post-discharge, she was instructed to regularly monitor her haemoglobin levels, which subsequently fluctuated between 60 and 90 g/l. By January 2022, she was diagnosed at an external facility with chronic non-atrophic gastritis accompanied by bile reflux, based on gastroscopic findings. However, her condition deteriorated, leading to her readmission due to the recurrence of hematemesis.

On physical examination at our facility, the patient appeared pale, had mild abdominal tenderness, and experienced postprandial vomiting, with evidence of blood in her gastric contents. There were no other obvious physical abnormalities. 13 urea breath test (13C-UBT) result of 9.1, which is positive and has *Helicobacter pylori* infection. A comprehensive gastroscopy (Fig. 1B) displayed a congested and oedematous mucosa within the gastric antrum, characterized by alternating red and white zones—predominantly red. No signs of active bleeding were noted. The mucosa was coated with white moss-like patches, giving it a rough and uneven appearance. Furthermore, there were multiple irregular ulcers, each measuring ~0.5 × 0.6 cm. A subsequent rapid urease test was positive. Histopathological examination of biopsy samples confirmed B-cell lymphoma, localized to the anterior wall, posterior wall, and the corner of the gastric antrum.

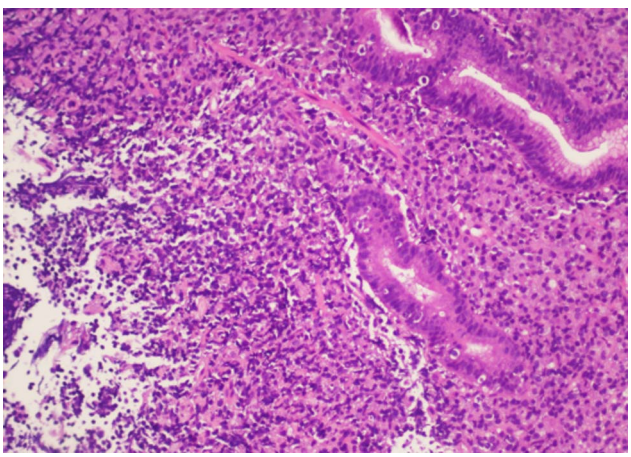

Figure 1. Pathological tissue showing small round mature lymphocyte infiltration.

Subsequent to the initial findings, positron emission tomography/computed tomography (PET/CT) scans were performed. These scans depicted impaired filling of the gastric cavity, mild thickening of the gastric wall, and a patchy increase in FDG metabolism, which was particularly prominent in the gastric antrum. Additionally, the scans showed heightened FDG metabolism in a localized thickened region of the nasopharynx. Bilateral slightly enlarged lymph nodes in the neck with modestly elevated FDG metabolism were also noted. A soft tissue density shadow was evident in the right anterior mediastinum, exhibiting a mild increase in patchy FDG metabolism. Moreover, there was a diffuse elevation in FDG metabolism throughout the skeletal system. Taking all these findings into account and using the Lugano staging system as reference, the patient was categorized under St. Jude stage I[7]. Considering the entirety of her medical history, the primary diagnosis was extranodal MALT lymphoma with a unique plasma cell differentiation.

In terms of treatment, the patient was prescribed an anti-*H. pylori* regimen, which included rabeprazole, colloidal bismuth pectin, amoxicillin, and clarithromycin. Concurrently, erythropoietin and iron supplementation were initiated to address her anaemia. Following this regimen, the patient was further treated with amoxicillin, furazolidone, and lansoprazole. By January and July 2023, 13C-UBT was conducted which yielded normal results, specifically 1.9 and 2.1, respectively. Additionally, a gastroscope examination in August 2023 confirmed the absence of any abnormalities within the digestive tract.

Clinical discussion

Currently, there is no standardized endoscopic or macroscopic classification system for primary gastric lymphoma[5]. Gastric MALT lymphoma often presents variably and nonspecifically under endoscopic examination. However, definitive diagnosis can be achieved through endoscopic biopsy and subsequent pathological examination[6]. Gastric MALT upper gastrointestinal endoscopic manifestations are diverse, encompassing mucosal redness, lump or polypoid lesion with or without ulcer, benign gastric ulcer, tubercle and thickened gyrate gastric folds[8,9]. To improve the accuracy and sensitivity of endoscopic biopsy[[10]], the M-system from the magnifying endoscope is used[11]. Therefore, multi-point biopsy in the lesions with abnormal manifestations is performed to help improve the diagnosis. In this case, the child's gastroscopy showed mainly non-specific ulcers, and a multi-point biopsy helped obtain a definitive diagnosis. In addition, endoscopic ultrasonography and *H. pylori* examination also have certain auxiliary diagnostic value for evaluating the depth of invasion and prognosis[12].

A Japanese study established a positive correlation between gastric lymphoma and *H. pylori* infection[13]. *H. pylori*-induced gastritis can recruit CD4 + lymphocytes and mature B cells to the lamina propria of the gastric wall. *H. pylori* antigens drive T cell activation, B-cell

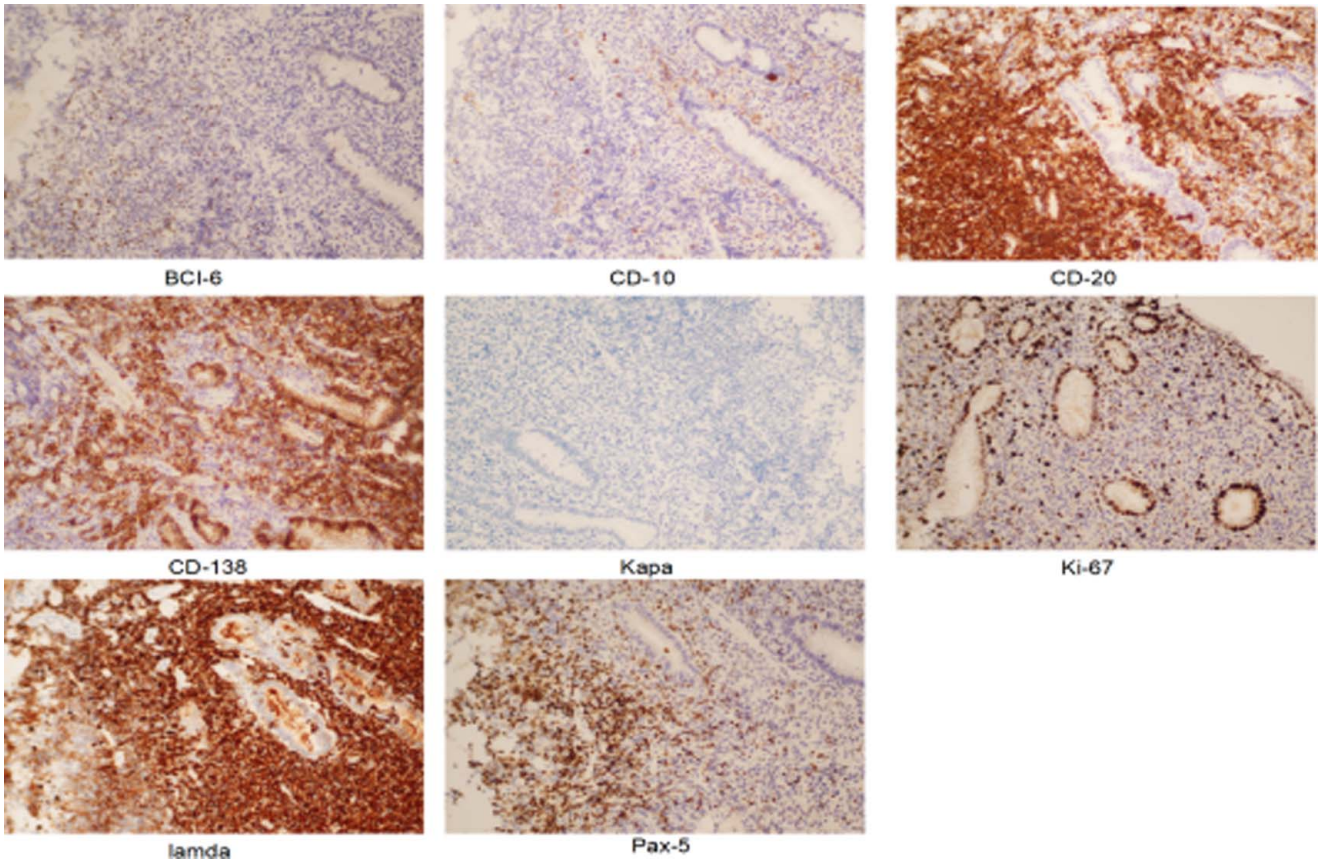

Figure 2. Immunohistochemistry: Bcl-6 (focus +), CD10 (−), CD20 (partial +), CD138 (+), Kappa (−), Ki-67 (about 30% +), lambda (+), Pax-5 (partial +).

proliferation, and lymphoid follicle formation. Prolonged stimulation can lead to monoclone lymphomas. In 80% of cases, eradication of H. pylori early in the disease results in complete resolution of lymphoma[14]. Therefore, triple or quadruple therapy is the recommended first-line therapy for eradication of *H. pylori*. A long-term multicenter retrospective cohort study showed that radical radiotherapy was effective and less toxic in patients with gastric MALT lymphoma[15]. Consequently, radiotherapy and chemotherapy are viable options for patients showing no remission or experiencing worsening symptoms. Surgical resection, due to its inability to eradicate lymphoma, is generally reserved for cases with severe complications like perforation and obstruction[16]. Immunotherapy may be more advantageous for refractory and recurrent cases[17]. Monoclonal antibodies combined with CD20 have shown promising outcomes when used alongside other treatment modalities. Literature also reports the use of adjuvant chemotherapy following anti-*H. pylori* treatment as a common clinical approach. Fortunately, in this case, a single anti-*H. pylori* treatment yielded a favourable response, consistent with existing literature[18]. Notably, this patient initially received anti-*H. pylori* treatment but later developed gastric MALT lymphoma. This might be attributed to resistance to the initial *H. pylori* treatment or incorrect medication adherence. Because it is different from the Western meal system, the Chinese eating habit is a shared meal system, which makes Helicobacter pylori very easy to recur and spread. At the same time, the children and their families have incomplete understanding of the disease, which may lead to not taking drugs on time and in accordance with the dosage, and under the influence of various factors, although anti-*Helicobacter pylori* treatment has been treated, the effect may not be satisfactory and not completely eradicated, resulting in the recurrence or progression of MALT. Therefore, clinicians must be vigilant in post-treatment surveillance for *H. pylori* eradication.

Conclusion

This case report of gastric MALT lymphoma in a paediatric patient with hematemesis as a primary symptom emphasizes the rarity and diagnostic challenges of this condition in children. It highlights the critical need for paediatricians and oncologists to consider atypical presentations in differential diagnoses, particularly when common gastrointestinal disorders are suspected. The successful management through gastroscopic pathological examination and *H. pylori* treatment underscores the potential for favourable outcomes. This report contributes to the limited literature on paediatric gastric MALT lymphoma, offering insights for improved diagnosis and treatment. It calls for further research to understand the disease's pathophysiology in children and to develop age-specific management protocols.

Ethical approval

No ethical review is required.

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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Author contribution

X.J. is responsible for conceptualization, formal analysis, and writing—original draft. L.L. is responsible for conceptualization and data curation. X.J. is responsible for conceptualization, data curation and writing—review and editing. X.J. is responsible for conceptualization, data curation and writing—review and editing. P.G. is responsible for conceptualization and data curation. W.S. is responsible for conceptualization and writing—review and editing. D.Z. is responsible for conceptualization, data curation, writing—original draft and writing -review and editing.

Conflicts of interest disclosure

The authors declare that they have no conflicts of interest. The work has been reported in line with the SCARE criteria.

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Guarantor

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Data availability statement

The data in this article are publicly available.

Provenance and peer review

The article was not invited.

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