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CASE REPORT

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Acute pancreatitis revealing duodenal papilla MALT lymphoma: a case report and review of the literature

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ABSTRACT

Mucosa-associated lymphoid tissue (MALT) lymphoma is a rare subtype of non-Hodgkin lymphoma, predominantly found in the gastrointestinal tract, particularly the stomach. However, its localization at the ampulla of Vater is exceptionally rare and presents considerable diagnostic challenges. This atypical presentation can easily mimic more common conditions, such as ampullary tumors or pancreaticobiliary disorders, making accurate diagnosis and management particularly complex. We present the case of a 70-year-old woman who initially presented with obstructive jaundice and acute pancreatitis, ultimately revealing a diagnosis of MALT lymphoma at the duodenal papilla. She was managed with combination of Helicobacter pylori eradication therapy followed by chemotherapy. This case emphasizes the importance of considering lymphoproliferative disorders in the differential diagnosis of biliary obstruction and pancreatitis, highlighting the need for heightened awareness among clinicians. Through this unique case, we aim to contribute to the limited literature on MALT lymphoma at the ampulla of Vater, illustrating both the complexities of diagnosis and the potential for successful management strategies in rare gastrointestinal lymphomas.

PLAIN LANGUAGE SUMMARY

This case report is about a rare cancer called MALT lymphoma, which was found in a part of the small intestine where the bile duct connects. A 70-year-old woman had symptoms like yellowing of the skin and eyes, stomach pain, and weight loss, which are also seen in pancreatic cancer. Doctors thought it might be pancreatic cancer after seeing signs of a mass in the pancreas on tests. But further tests showed an unusual growth in the small intestine. A biopsy confirmed it was MALT lymphoma, a type of cancer that affects the immune system. MALT lymphoma usually happens in the stomach but is very rare in the small intestine. The patient was treated with antibiotics for a bacteria called *Helicobacter pylori*, followed by chemotherapy. She felt much better after treatment and has been symptom-free for two years.

ARTICLE HIGHLIGHTS

- MALT lymphoma is an exceptionally rare form of non-Hodgkin lymphoma, with even rarer occurrences found in the duodenal papilla, leading to significant diagnostic challenges.
- This case presents a 70-year-old woman whose initial presentation included cholestatic jaundice, epigastric pain, weight loss, with lipase levels, raising concerns for a pancreatic tumor.
- Imaging studies, including CT and MRI, revealed a pancreatic mass, biliary dilation, and lymphadenopathy, all suggestive of malignancy, though these findings did not provide a definitive diagnosis.
- Endoscopic retrograde cholangiopancreatography and subsequent duodenoscopy revealed a protruding lesion at the duodenal papilla, suggestive of an ampullary tumor.
- Histopathological examination of biopsy specimens identified lymphoepithelial lesions and small lymphoid cells, supporting the diagnosis of MALT lymphoma.
- Immunohistochemical staining was positive for CD20 and Bcl-2, with CD10 and CD5 negative, which further supported the diagnosis of MALT lymphoma.

ARTICLE HISTORY

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MALT; lymphoma; pancreatitis; chemotherapy; jaundice

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- The management approach involved a two-step strategy, beginning with H. pylori eradication therapy followed by R-CHOP chemotherapy,
- The patient had an excellent response to treatment, with resolution of symptoms, normalization of laboratory markers, and no recurrence of symptoms over a two-year follow-up period.
- The rarity of MALT lymphoma at the ampulla of Vater emphasizes the need for high clinical suspicion and thorough diagnostic evaluation to ensure an accurate diagnosis and appropriate treatment.

1. Introduction

Mucosa-associated lymphoid tissue (MALT) lymphoma is a rare subtype of non-Hodgkin lymphoma characterized by the abnormal proliferation of B cells in the marginal zone of lymphoid tissues. It is often characterized by a relatively indolent growth pattern, although more aggressive forms can occur. The lymphoma most commonly affects the stomach, where a well-established association with Helicobacter pylori infection exists [1]. MALT lymphoma typically occurs in older adults, particularly between the ages of 50 and 60, with a slight female predominance [2,3].

While gastric MALT lymphoma is the most prevalent form, non-gastric MALT lymphomas are significantly less common and can develop in various anatomical sites, including the salivary glands, thyroid, lungs, and other regions of the gastrointestinal tract [4]. One notably rare manifestation is MALT lymphoma at the ampulla of Vater. This unusual location can lead to diagnostic challenges, as it may mimic more prevalent conditions like ampullary tumors or pancreaticobiliary disorders [5].

In this report, we present a compelling case of a 70-year-old woman whose presentation of acute pancreatitis ultimately revealed a diagnosis of MALT lymphoma at the duodenal papilla. This case emphasizes the importance of considering lymphoproliferative disorders in the differential diagnosis of biliary obstruction and pancreatitis. Additionally, we will explore the potential mechanisms by which MALT lymphoma can lead to acute pancreatitis and the treatment strategies that may be employed. Through this case, we aim to raise awareness among clinicians regarding this rare but significant entity.

2. Case presentation

A 70-year-old woman with no significant medical history presented to the emergency department with jaundice and signs of cholestasis, including pruritus and clay-colored stools, persisting for three months. She reported a progressive onset of epigastric pain that had intensified over the past two days, described as a dull ache radiating to the back. Additionally, she experienced significant weight loss of approximately 5 kg during this period.

Physical examination revealed marked scleral icterus and excoriations on the skin from scratching, along with tenderness in the epigastric region. There were no signs of hepatosplenomegaly or ascites, and the patient was afebrile with stable vital signs.

Initial laboratory tests showed significant abnormalities, including elevated lipase at 455 U/L (normal range: 0–160 U/L), total bilirubin at 377 µmol/L (normal range: 5–17µmol/L), and direct bilirubin at 250 µmol/L (normal range: 0–6 µmol/L). Additionally, CA 19-9 was notably elevated at 611 U/mL (normal range: <37 U/mL).

Imaging studies, including abdominal CT scan, demonstrated stage C pancreatitis according to the Balthazar classification, alongside biliary dilation and retroperitoneal and mesenteric lymphadenopathy (Figure 1). MRI showed a spiculated appearance in the head of the pancreas and biliary dilatation (Figure 2).

A preliminary diagnosis of pancreatic head cancer was considered due to the patient's clinical presentation and imaging findings. An endoscopic retrograde cholangiopancreatography (ERCP) was performed; however, cannulation of the main bile duct was unsuccessful. Subsequent duodenoscopy revealed a protruding, non-ulcerative lesion at the duodenal papilla, suggestive of an ampullary tumor (Figure 3).

Biopsies examination revealed small lymphoid cells with occasional lymphoepithelial lesions (LEL) (Figure 4). Immunohistochemical staining showed intense expression of CD20 and Bcl-2, while CD3, CD5, and CD10 were negative (Figure 5). Although IgH-BCL2 FISH testing, which could have helped exclude the presence of BCL2 translocations characteristic of follicular lymphoma (FL), was not carried out due to technical limitations, the

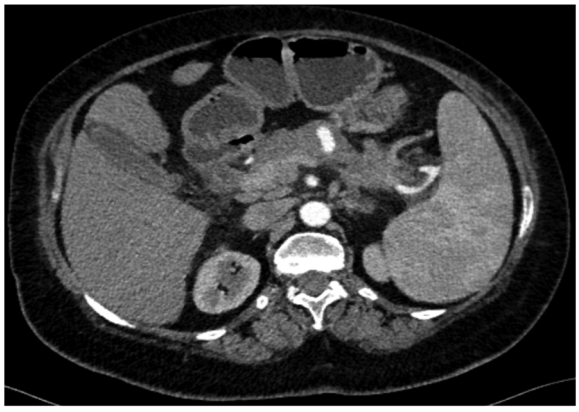

Figure 1. The CT scan reveals a swollen pancreas with loss of physiological lobulations and densification of peripancreatic fat, consistent with stage C pancreatitis.

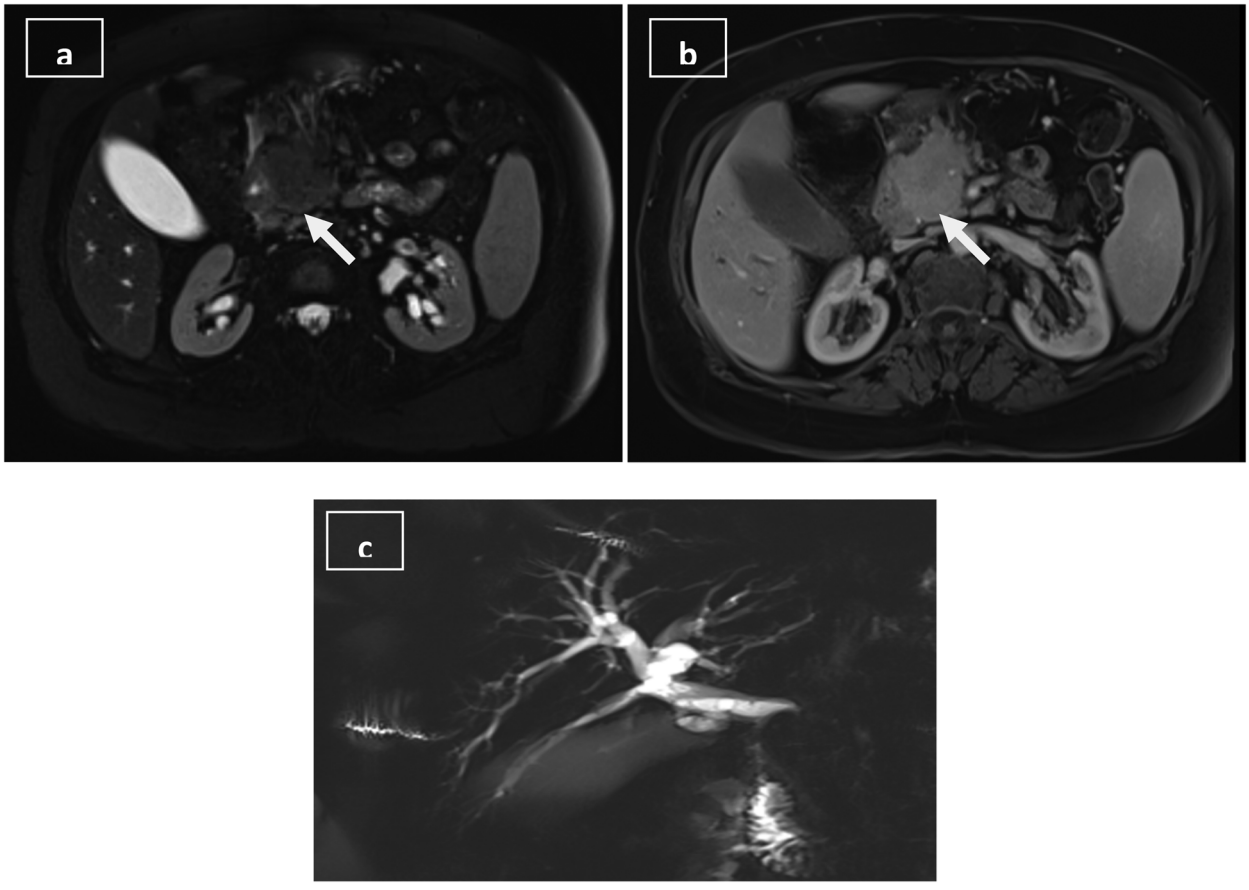

Figure 2. MRI with T2-weighted (a) and T1-weighted with gadolinium contrast (b) axial sections demonstrating pancreatic enlargement and biliary tract dilation. Axial MRI images using T2-weighted (a) and T1-weighted with gadolinium contrast (b) sequences show a swollen, spiculated appearance of the pancreatic head (arrow). Additionally, MRI imaging of the biliary tract demonstrates notable intra- and extra-hepatic bile duct dilation (c), raising concerns of obstructive pathology, which prompted further diagnostic evaluation.

overall histological and immunohistochemical findings, combined with the clinical presentation, strongly supported a diagnosis of MALT lymphoma.

Staging with a thoracoabdominal CT scan showed no distant localization but did reveal local lymphadenopathy. The patient was classified as II1E according to the Ann Arbor classification revised by Musshof.

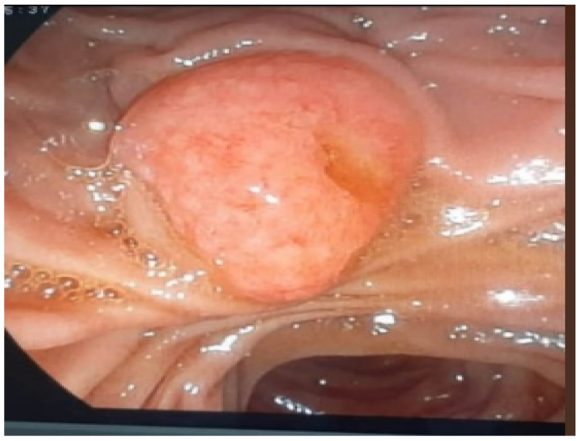

Figure 3. Duodenoscopy revealing the presence of a protruding lesion at the duodenal papilla. Duodenoscopy revealed a protruding, non-ulcerative lesion at the duodenal papilla, raising suspicion for an ampullary tumor. This finding prompted a biopsy, which ultimately confirmed the diagnosis of MALT lymphoma.

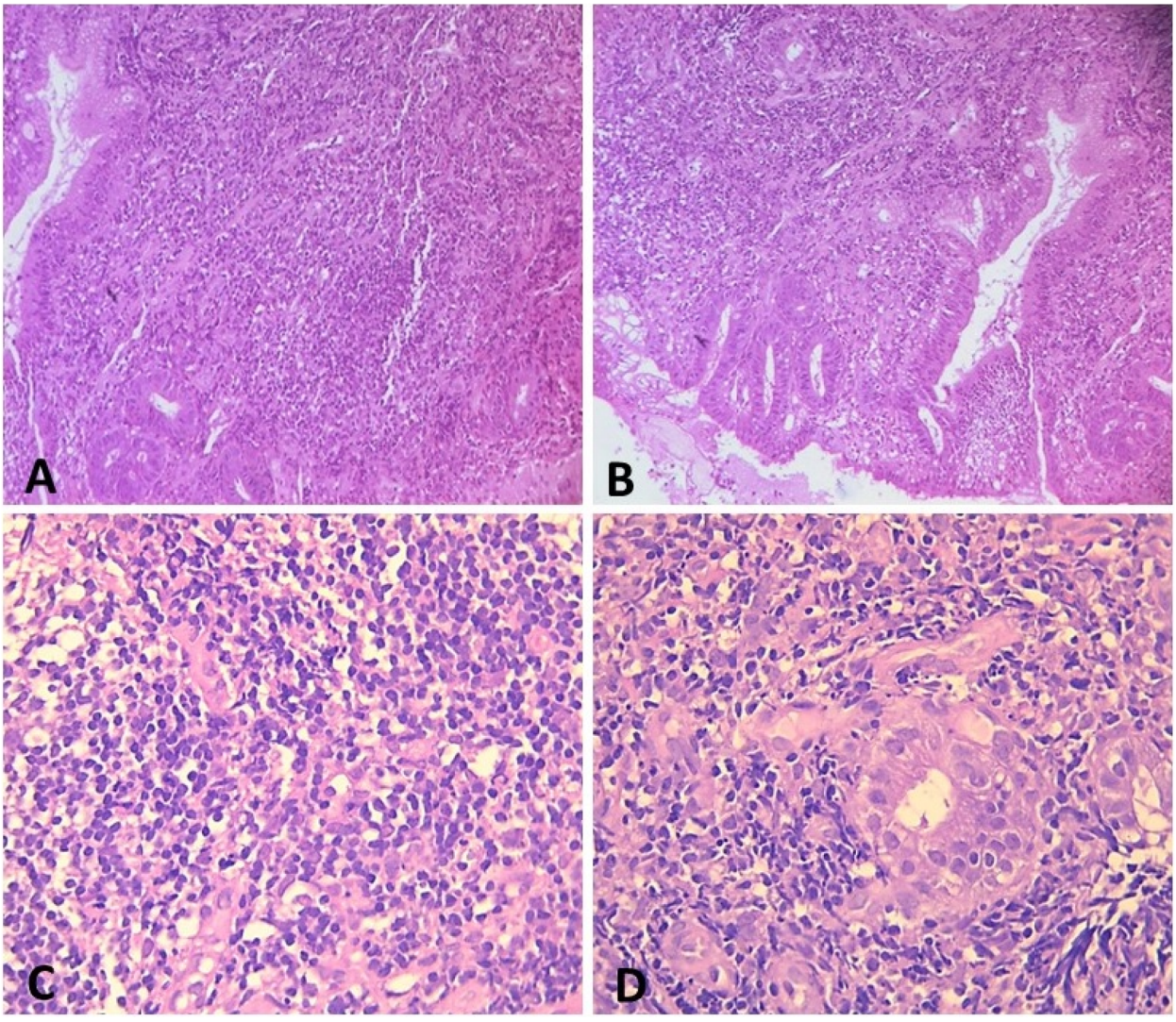

Figure 4. Histology: Hematoxylin and eosin stain(HE) sections of the duodenal biopsy showing patchy involvement by a dense and atypical lymphoplasmacytic infiltrate with lymphoepithelial lesions. A-B (HE x 10), C-D (HE x 40). These features are consistent with MALT lymphoma, especially when observed in the mucosa and submucosa of the duodenal papilla.

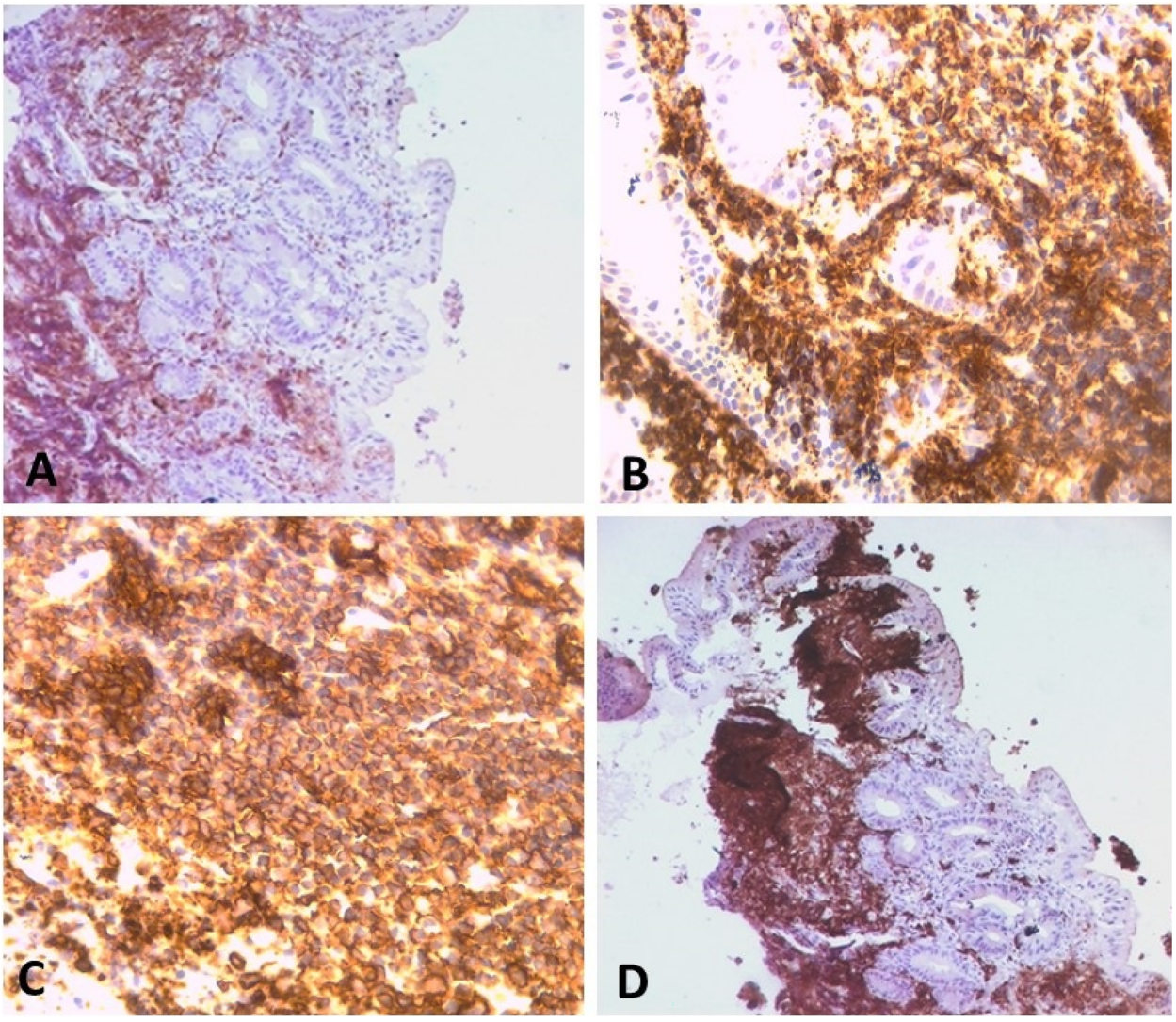

Figure 5. Immunohistochemical staining showed an intense expression of CD20: A (x10), B-C (x40) and Bcl-2: D (x10). These findings confirmed the diagnosis of MALT lymphoma in the duodenal papilla. These findings support the diagnosis of MALT lymphoma and help differentiate it from other types of lymphoid malignancies.

The patient underwent standard therapy for acute pancreatitis. Follow-up laboratory tests showed a marked decrease in bilirubin levels (78 µmol/L) and CA 19-9 (26 U/mL) probably due to papilla manipulation during ERCP.

The patient was first treated with *Helicobacter pylori* (H.pylori) bismuth therapy followed up by chemotherapy, using R-CHOP combination immunochemotherapy (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisone). She underwent six cycles of R-CHOP. The patient has now been free of symptoms for 2 years.

3. Discussion

Duodenal lymphoma is exceptionally uncommon, comprising less than 2% of all gastrointestinal lymphomas. The most frequently affected regions include the bulb of the duodenum, the second portion, and occasionally multiple sites. Within primary duodenal lymphomas, MALT lymphoma represents approximately 13.8% [6]. Even more rare is MALT lymphoma localized specifically at the ampulla of Vater; our review of the English literature identified only six documented cases involving this site (Table 1). These cases present with varied symptoms, including obstructive jaundice, abdominal pain, weight loss, and incidental findings during investigations for other complaints. Notably, only one case presented with acute pancreatitis and obstructive jaundice, which parallels our own case [2].

Table 1. Summary of previous cases of MALT lymphoma at the ampulla of Vater.

Author	Year	Patient age	Gender	Symptoms	Endoscopic aspect	Treatment modality	Outcome
J Pawade et al. [7]	1994	57	Female	Obstructive jaundice	Protuberant tumor at the ampulla of Vater with multiple mucosal polyps in the duodenum	Surgery (Whipple's procedure)	Complete remission post-surgery
L Zhu et al. [8]	1995	53	Female	Heartburn, abdominal distension, nausea	Irregular ampullary mucosa, nodularity	None (observation)	No evidence of tumor progression
Ventrucci et al. [9]	1998	65	Female	Recurrent postprandial abdominal pain	Polypoid mass on the ampulla of Vater	Chemotherapy	No evidence of lymphoma post-treatment
Toyoda et al. [10]	2001	62	Male	Asymptomatic; incidental finding	Multiple granules around major papilla	H. pylori eradication	Regression after H. pylori eradication
Isomoto et al. [11]	2003	46	Male	Asymptomatic; incidental finding	Whitish granules around ampulla	Triple therapy for H. pylori, radiotherapy	Complete remission after radiotherapy
Simkova et al. [2]	2015	72	Female	Obstructive jaundice and acute pancreatitis	Exophytic ulcerating infiltration in the region of the papilla of Vater	Chemotherapy	Complete remission

H.pylori: Helicobacter pylori.
This table presents a review of six documented cases of MALT lymphoma localized at the ampulla of Vater. It includes patient demographics, clinical presentation, diagnostic findings, and treatment outcomes. The ages of patients ranged from 46 to 72 years, and the most common presentations included obstructive jaundice and abdominal pain.

The diagnostic dilemma in cases of duodenal papillary involvement is considerable. In fact, cholestatic jaundice and acute pancreatitis are frequent clinical findings in many pancreato-biliary disorders, including malignant neoplasms such as pancreatic cancer and ampullary tumors. As a result, distinguishing MALT lymphoma from more common conditions requires a high level of clinical suspicion and thorough diagnostic evaluation. Early imaging studies in our patient suggested a pancreatic mass, with biliary dilation and retroperitoneal lymphadenopathy, which raised concern for malignancy. However, the definitive diagnosis of MALT lymphoma was achieved only after the subsequent finding of a protruding lesion at the duodenal papilla and the histological examination of biopsies obtained via duodenoscopy, which revealed small lymphoid cells with LEL. Immunohistochemical analysis further confirmed this diagnosis, demonstrating intense expression of CD20 and Bcl-2 while being negative for CD3, CD5, and CD10. While FL could be considered due to the strong Bcl-2 expression, several histological and immunohistochemical findings, in conjunction with the clinical presentation, led us to exclude FL in favor of MALT lymphoma. Although IgH-BCL2 FISH testing, which would confirm BCL2 translocations characteristic of FL, was not possible due to technical limitations, the overall findings strongly support the diagnosis of MALT lymphoma. The biopsy revealed LELs, a feature commonly associated with MALT lymphoma, but not typically seen in FL, where follicular architecture is more characteristic. Immunohistochemical staining showed strong Bcl-2 expression, which can be present in both MALT lymphoma and FL, but it was negative for CD10, a marker typically expressed in FL. Additionally, CD5 was absent, further differentiating the lymphoma from FL. These findings, along with the presence of lymphoepithelial lesions and the lack of CD10 and CD5, made FL an unlikely diagnosis. Thus, despite the inability to perform IgH-BCL2 FISH testing, the histological and immunohistochemical features strongly pointed to MALT lymphoma, ruling out FL.

The management of duodenal MALT lymphoma, especially when localized to the ampulla of Vater, differs from more common gastrointestinal lymphomas, such as gastric MALT lymphoma, where eradication of H. pylori has been a cornerstone of treatment. In gastric MALT lymphoma, the eradication of H. pylori can induce regression of the lymphoma in a significant proportion of cases [12]. However, the role of H. pylori eradication in duodenal MALT lymphoma remains a subject of debate. Some studies suggest that similar to gastric cases, duodenal MALT lymphoma may regress following H. pylori eradication, while others have found no correlation, indicating that the pathogenetic mechanisms in duodenal MALT lymphoma may differ from those seen in gastric cases [10,11,13].

Local treatments, including surgical resection and radiotherapy, are also options for managing duodenal MALT lymphoma [11,14,15]. The risks of radiotherapy (perforation, bleeding, and renal toxicity) are minimal,

with incidences comparable to or lower than those of surgery [11]. Some authors suggest that systemic chemotherapy should have a central role in treatment [16].

In our case, a two-step approach was employed, beginning with H. pylori eradication therapy, which is typically recommended in suspected MALT lymphoma cases with H. pylori association. However, given the patient's acute clinical presentation with obstructive jaundice and pancreatitis, it was deemed urgent to initiate chemotherapy without waiting for the response to H. pylori therapy. We chose to proceed with the R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone), which is the standard treatment for non-Hodgkin lymphoma, including MALT lymphoma. The decision to rapidly initiate chemotherapy was also supported by the limited evidence for the efficacy of H. pylori eradication alone in duodenal MALT lymphoma and the poor prognosis associated with delays in therapy for advanced-stage lymphomas.

The patient's response to the combination of H. pylori therapy and subsequent chemotherapy was favorable, with resolution of symptoms and normalization of laboratory markers during follow-up. The patient has remained symptom-free for two years, suggesting a good long-term prognosis following appropriate treatment. This case reinforces the need for individualized management strategies, especially for rare and complex lymphoma presentations like MALT lymphoma of the ampulla of Vater.

4. Conclusion

In conclusion, duodenal MALT lymphoma is an exceptionally rare form of lymphoma, and even rarer still is its localization to the ampulla of Vater. The diagnosis of this condition requires a high index of suspicion and a comprehensive diagnostic work-up, including imaging, endoscopy, and histopathological examination. Management is complex and should be individualized, particularly in light of the limited evidence for the role of H. pylori eradication in duodenal lymphoma. This case contributes to the growing but still limited body of literature on MALT lymphoma in uncommon gastrointestinal locations, highlighting the importance of early recognition, accurate diagnosis, and appropriate treatment for improving patient outcomes. It also underscores the necessity for clinicians to consider lymphoproliferative disorders in the differential diagnosis of biliary obstruction and pancreatic inflammation.

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Authors' contributions

Wafa Dahmani, Houssem Hassan: conceptualization and writing; Aya Hammami: formal analysis. Aida Ben Slama, Ahlem Brahem: supervision. Hanene Jaziri, Nour Elleuch and Mehdi Ksiaa: validation.

Disclosure statement

No relevant affiliations or financial involvement with any organization or entity with a financial interest in or financial conflict with the subject matter or materials discussed in the manuscript have been used. This includes employment, consultancies, honoraria, stock ownership or options, expert testimony, grants or patents received or pending, or royalties.

References

Papers of special note have been highlighted as either of interest (•) or of considerable interest (••) to readers.

[1] Wotherspoon AC, Diss TC, Pan L, et al. Regression of primary low-grade B-cell gastric lymphoma of mucosa-associated lymphoid tissue type after eradication of *Helicobacter pylori*. Lancet. 1993;342(8871):575–577. doi: 10.1016/0140-6736(93)91409-f

[2] Simkova I, Urbanek K, Prochazka V, et al. Acute pancreatitis as the first manifestation of duodenal MALT lymphoma. Biomed Pap Med Fac Univ Palacky Olomouc Czech Repub. 2015;159(4):688–694. doi: 10.5507/bp.2015.047

[3] Ndzengue A, Khurana R, Mora M, et al. Gastric marginal zone B cell lymphoma of the duodenum. Case Rep Gastroenterol. 2011;5(3):578–582. doi: 10.1159/000331137

* *Marked as "of interest" because this study contributes valuable insights into the rare occurrence of MALT lymphoma in the duodenum, helping to contextualize the presentation of this lymphoma in other gastrointestinal sites.*

[4] Zucca E, Bertoni F. The spectrum of MALT lymphoma at different sites: biological and therapeutic relevance. Blood. 2016;127(17):2082–2092. doi: 10.1182/blood-2015-12-624304

** *Marked as "of considerable interest" as it provides an overview of the biological and therapeutic implications of MALT lymphoma at various sites, which is critical for understanding the treatment of this lymphoma in the duodenal papilla.*

[5] Joo YE, Park CH, Lee WS, et al. Primary non-Hodgkin's lymphoma of the common bile duct presenting as obstructive jaundice. J Gastroenterol. 2004;39(7):692–696. doi: 10.1007/s00535-004-1367-0

* *Marked as "of interest" due to its examination of non-Hodgkin's lymphoma in the biliary tract, providing context for the presentation of lymphoma in a nearby anatomical region and aiding in differential diagnosis.*

[6] Iwamuro M, Tanaka T, Okada H. Review of lymphoma in the duodenum: An update of diagnosis and management. World J Gastroenterol. 2023;29(12):1852–1862. doi: 10.3748/wjg.v29.i12.1852

** *Marked as "of considerable interest" for its comprehensive update on the diagnosis and management of duodenal lymphoma, which is highly relevant to the presentation of MALT lymphoma in this case.*

[7] Pawade J, Lee SC, Ellis DW, et al. Primary lymphoma of the ampulla of Vater. Cancer. 1994;73(8):2083–2086. doi: 10.1002/1097-0142(19940415)73:8<2083::aid-cncr2820730812>3.0.co;2-b

** *Marked as "of considerable interest" because it establishes the link between Helicobacter pylori infection and the development of gastric lymphoma, an important factor in understanding potential pathogenetic mechanisms in MALT lymphoma.*

[8] Zhu L, Slee GR, Domenico DR, et al. B-cell lymphoma of ampulla of Vater: observation for six years. Pancreas. 1995;10(2):208–210. doi: 10.1097/00006676-199503000-00018

** *Marked as "of considerable interest" due to its findings that Helicobacter pylori eradication may lead to the regression of primary lymphoma of the ampulla of Vater, providing a potential treatment strategy for MALT lymphoma in this location.*

[9] Ventrucci M, Gherlinzoni F, Sabattini E, et al. Case report: primary MALT-lymphoma of the papilla of Vater. Dig Dis Sci. 1998;43(1):214–216. doi: 10.1023/a:1018865213404

[10] Toyoda H, Yamaguchi M, Nakamura S, et al. Regression of primary lymphoma of the ampulla of Vater after eradication of Helicobacter pylori. Gastrointest Endosc. 2001;54(1):92–96. doi: 10.1067/mge.2001.114963

[11] Isomoto H, Kamihira S, Matsuo E, et al. A case of mucosa-associated lymphoid tissue lymphoma of the ampulla of Vater: successful treatment with radiation therapy. Eur J Gastroenterol Hepatol. 2003;15(9):1037–1041. doi: 10.1097/00042737-200309000-00016

[12] Parsonnet J, Hansen S, Rodriguez L, et al. Helicobacter pylori infection and gastric lymphoma. N Engl J Med. 1994;330(18):1267–1271. doi: 10.1056/NEJM199405053301803

[13] Nagashima R, Takeda H, Maeda K, et al. Regression of duodenal mucosa-associated lymphoid tissue lymphoma after eradication of *Helicobacter pylori*. Gastroenterol. 1996;111(6):1674–1678. doi: 10.1016/s0016-5085(96)70032-x

[14] Tsang RW, Gospodarowicz MK, Pintilie M, et al. Stage I and II malt lymphoma: results of treatment with radiotherapy. Int J Radiat Oncol Biol Phys. 2001;50(5):1258–1264. doi: 10.1016/s0360-3016(01)01549-8

[15] Montalbán C, Castrillo JM, Abraira V, et al. Gastric B-cell mucosa-associated lymphoid tissue (MALT) lymphoma. Clinicopathological study and evaluation of the prognostic factors in 143 patients. Ann Oncol. 1995;6(4):355–362. doi: 10.1093/oxfordjournals.annonc.a059184

[16] Hammel P, Haioun C, Chaumette MT, et al. Efficacy of single-agent chemotherapy in low-grade B-cell mucosa-associated lymphoid tissue lymphoma with prominent gastric expression. J Clin Oncol. 1995;13(10):2524–2529. doi: 10.1200/JCO.1995.13.10.2524