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A RARE TUMOUR OF THE AMPULLA OF VATER

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ABSTRACT

Primary lymphoma of the ampulla of Vater is an exceptionally rare entity that may clinically mimic more common periampullary malignancies. Duodenal involvement is uncommon among gastrointestinal lymphomas, and mucosa-associated lymphoid tissue (MALT) lymphoma at this site is particularly rare. Diagnosis is often challenging due to the submucosal location of the lesion and the limited yield of conventional endoscopic biopsies.

A 59-year-old woman presented with painless obstructive jaundice, weight loss and cholestatic liver enzyme abnormalities. Abdominal computed tomography (CT) scan revealed a periampullary mass with regional lymphadenopathy, biliary dilatation and a solitary lesion in the left hepatic lobe. Endoscopic retrograde cholangiopancreatography demonstrated a deformed ampulla with ulcerated periampullary mucosa. However, repeated endoscopic biopsies were inconclusive. Definite diagnosis was established only after endoscopic ultrasound-guided fine needle biopsy (EUS-FNB) of both the primary duodenal lesion and the hepatic lesion, which demonstrated B-cell non-Hodgkin lymphoma with immunohistochemical features consistent with MALT lymphoma. Staging investigations, including positron emission tomography -CT and bone marrow biopsy, showed stage IV disease.

Ampullary MALT lymphoma is an extremely rare cause of obstructive jaundice and may be indistinguishable from other ampullary or pancreatic neoplasms. This case highlights the diagnostic limitations of conventional endoscopic biopsies and the pivotal role of EUS-FNB in obtaining adequate tissue samples from deeper layers. Early and accurate diagnosis is essential, as appropriate treatment can result in excellent clinical outcomes. EUS-FNB should be considered when initial biopsies are nondiagnostic in suspected periampullary malignancies, as it may facilitate the diagnosis of rare entities such as MALT lymphoma.

KEYWORDS

Vater papilla, primary MALT lymphoma

LEARNING POINTS

- MALT lymphoma is a rare subtype of non-Hodgkin lymphoma, and involvement of the ampulla of Vater is exceptionally uncommon.
- Obstructive jaundice is the most common clinical presentation of ampullary lymphoma.
- Endoscopic ultrasound-guided tissue acquisition is particularly valuable when conventional endoscopic biopsies are nondiagnostic.

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INTRODUCTION

Primary lymphoma of the ampulla of Vater is a rare entity, with only a limited number of cases reported in the literature. The gastrointestinal (GI) tract is the most common site of primary extranodal lymphoma, accounting approximately 30-40% of cases[1]. The stomach is the most frequently involved organ of GI tract, whereas the duodenum is among the least commonly affected sites[1,2]. The most common histological subtypes include mucosa-associated lymphoid tissue (MALT) lymphoma and high-grade B-cell lymphomas[1,3].

MALT lymphoma is a common subtype of non-Hodgkin's lymphoma; however, duodenal MALT lymphoma, particularly involving the ampulla of Vater, is exceptionally rare[4]. The most common clinical presentation is obstructive jaundice, and the condition should be differentiated from pancreatic adenocarcinoma, cholangiocarcinoma and ampullary carcinoma. Diagnosis is challenging because conventional endoscopic biopsies may be nondiagnostic owing to the predominantly submucosal location of the tumour[5].

Endoscopic ultrasound (EUS) is particularly useful because it enables tissue acquisition from deeper layers and from secondary lesions when present[6]. We report a diagnostically challenging case of MALT lymphoma of the ampulla of Vater in which repeated endoscopic biopsies were inconclusive.

CASE DESCRIPTION

A 59-year-old Greek woman with no significant medical history was referred to our hospital with jaundice, dark urine, pale stools, and an unintentional 5-kg weight loss over a period of 2 months. She denied abdominal pain or other associated symptoms. Laboratory investigations on admission, demonstrated cholestatic liver dysfunction with aspartate aminotransferase 145 IU/l (normal 8-40 IU/l), alanine aminotransferase 229 (normal 5-42 IU/l), alkaline phosphate (ALP) 780 (normal 35-104 IU/L), γ-glutamyl transpeptidase 964 (normal 5-36 IU/l), total bilirubin 13.6 mg/dl (normal 0.3-1.3 mg/dl) and direct bilirubin 10.5 mg/dl (normal <0.5 mg/dl). CA 19-9 was mildly elevated at 95 U/ml (normal <37 U/ml), whereas carcinoembryonic antigen was within normal limits.

Abdominal computed tomography (CT) scan revealed a hypodense intra-abdominal solid lesion of a maximum diameter of 4.5 cm *(Fig. 1)* which represented a mass of lymph nodes and other smaller lymph nodes around this main lesion, and a solitary hypodense lesion at the left hepatic lobe. It also revealed a hypodense lesion in the second part of duodenum. The common bile duct was dilated, whereas the pancreatic duct was normal.

Endoscopic retrograde cholangiopancreatography was performed, and a plastic biliary stent was placed to relieve the obstruction. The ampulla of Vater appeared deformed and tumorous, while the adjacent periampullary mucosa was oedematous and ulcerated *(Fig. 2)*. Multiple biopsy samples were obtained from both the ampulla and the surrounding duodenal mucosa. Gastric biopsies were also taken to assess for *Helicobacter pylori* infection, which was negative. Histopathological examination was inconclusive, revealing only lymphocytic infiltration and necrotic tissue. Two additional endoscopic biopsy procedures were subsequently performed, but the results remained nondiagnostic.

Given the persistent diagnostic uncertainty, EUS-guided fine needle biopsy (EUS-FNB) was performed to obtain tissue samples from deeper layers of the primary lesion and from potential metastatic sites. Although the regional lymph nodes were inaccessible, biopsy samples were



Figure 1. Lymph node mass close to D2-3 portion of duodenum.

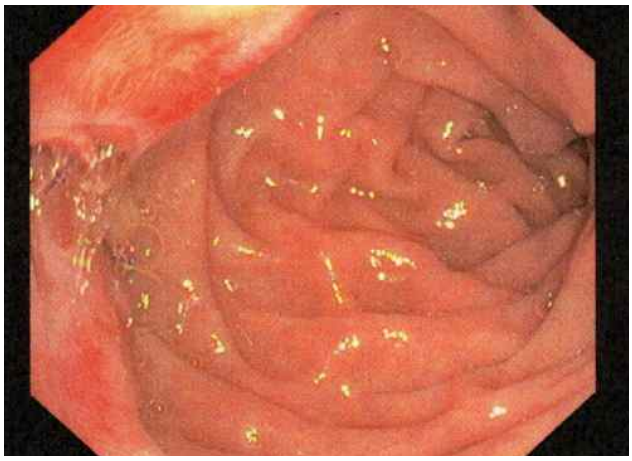


Figure 2. Tumorous ampulla of Vater.

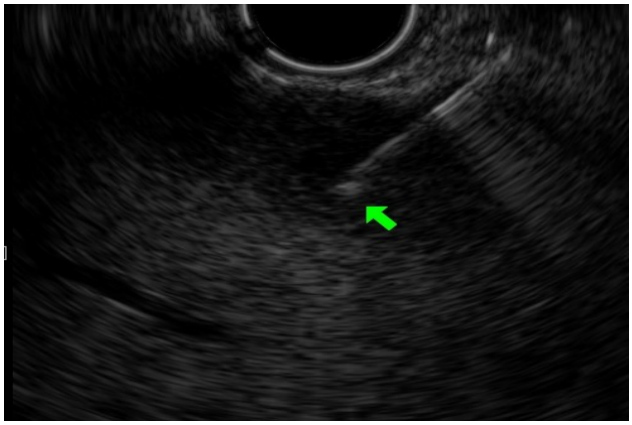


Figure 3. Left liver lobe lesion (arrow) seen on endoscopic ultrasound-guided fine needle biopsy.

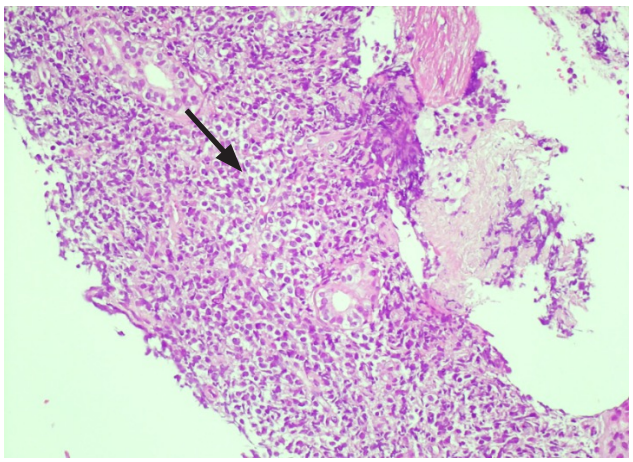


Figure 4. Increased number of lymphocytes with monocytoid differentiation (arrow) (20×).

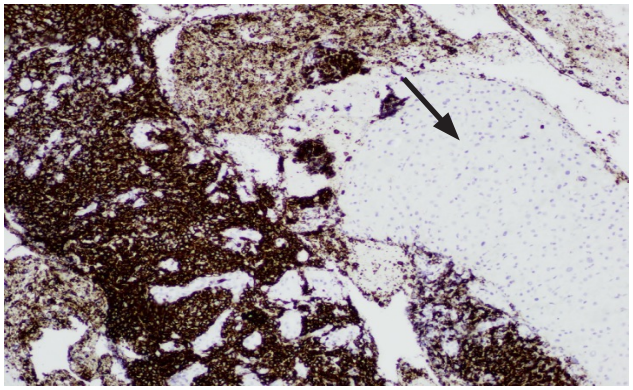


Figure 5. Higher magnification (10×). CD20 positive cells, adjacent to liver tissue (arrow).

successfully obtained from the deeper layers of the periampullary lesion and the left hepatic lobe lesion *(Fig. 3)*. Histopathological examination demonstrated extensive disruption of the normal hepatic architecture by infiltration of small lymphoid cells exhibiting monocytoid and plasmacytoid differentiation *(Fig. 4)*. Scattered larger atypical lymphoid cells were also observed. Immunohistochemical analysis showed positivity for CD20, CD79a, bcl2, MUM1, SIg/CIg(κ), while CD23, CD10, CD30, bcl6 and CD3 were negative *(Fig. 5)*. These findings were consistent with hepatic infiltration by extranodal marginal zone lymphoma, as classified by the World Health Organization in 2022. Duodenal biopsy specimens obtained through EUS were diagnostic of a small lymphocytic lymphoma; however, due to limited tissue quantity, further immunophenotypic characterization and definitive lymphoma subtyping could not be performed.

Staging computed tomography of the chest and pelvis showed no evidence of additional disease involvement. Positron emission tomography-CT did not reveal extra-abdominal disease, and bone marrow biopsy was negative for infiltration. The patient was therefore classified as having Lugano stage IV disease.

The patient was treated with systemic immunochemotherapy using the R-CHOP regimen (rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone) combined with radiotherapy. Doxorubicin was safely administrated after normalization of liver function tests following biliary decompression. After four cycles of R-CHOP, follow-up computed tomography demonstrated marked reduction of the ampullary mass, regional lymphadenopathy, and hepatic lesion, indicating a significant therapeutic response. The patient ultimately achieved complete radiological remission and remains under regular haematological follow-up.

DISCUSSION

Primary lymphoma of the ampulla of Vater is an exceptionally rare disease. Duodenal lymphoma accounts for less than 2% of all GI lymphomas and most commonly involves the duodenal bulb or the second portion of the duodenum. Among the primary duodenal lymphomas, approximately 14% of cases are MALT lymphoma. Involvement of the ampulla of Vater is extremely uncommon, with only few cases reported in the literature. In previously reported cases, conventional endoscopic biopsies were sufficient to establish the diagnosis. In contrast, our case required EUS-guided tissue acquisition after three consecutive endoscopic biopsies failed to provide a definitive diagnosis.

Painless obstructive jaundice, which was the presenting symptom in our patient, is the most common manifestation of ampullary and periampullary tumours and is more frequently associated with adenocarcinoma. Consequently, the initial diagnostic suspicion is often pancreatic or biliary malignancy[5]. Less common clinical manifestations include GI bleeding and diarrhoea[3,5].

The diagnosis of ampullary lymphoma can be particularly challenging because lymphomatous infiltration is frequently submucosal, rendering superficial endoscopic biopsies nondiagnostic. In addition, extensive necrosis may further compromise histopathological interpretation. EUS is especially valuable in such cases because it allows tissue acquisition from deeper layers, regional lymph nodes and the left hepatic lobe. In our patient, EUS-FNB enabled successful sampling of both the ampullary lesion and the hepatic lesion, ultimately leading to the diagnosis[7,8]. Furthermore, EUS provides accurate locoregional staging and facilitates targeted biopsy of accessible lymph nodes and metastatic lesions[7].

Endoscopic findings in GI MALT lymphoma are variable and may include mucosal nodularity, ulceration or polypoid lesions. Multiple synchronous lesions within the GI tract may also occur[8]. In the present case, the combination of a tumorous ampulla of Vater with ulcerated periampullary mucosal involvement was an exceptionally rare endoscopic presentation.

Treatment options for MALT lymphoma include radiotherapy, immunotherapy, chemotherapy or combined-modality treatment. The prognosis of primary ampullary MALT lymphoma varies depending on disease extent and lymph node involvement[1,9]. The R-CHOP regimen remains

a widely used therapeutic approach in advanced-stage disease[9].

In conclusion, primary duodenal MALT lymphoma is a rare entity, and involvement of the ampulla of Vater is exceptionally uncommon[2]. Imaging modalities and advanced endoscopic techniques, particularly EUS, play a central role in both diagnosis and staging[2,5]. This case highlights the importance of considering lymphoma in the differential diagnosis of periampullary lesions and demonstrates the crucial role of EUS-FNB when conventional biopsies are inconclusive.

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