CASE REPORT

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An Aggressive Form of MALT Lymphoma of the Stomach with Pancreas Infiltration

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ABSTRACT

Introduction: MALT lymphoma accounts for 7-8% of all B-cell lymphomas and at least 50% of primary gastric lymphoma, with the highest incidence at between 50 and 60 years of age. Aggressive forms are rare, as are indications for multi-visceral resection. Case study: A patient, 33 years old, was admitted to the tertiary hospital due to a biopsy at a small community hospital confirming adenocarcinoma of the stomach. She was Helicobacter pylori positive. CT showed thickening of the fundus and corpus wall, up to 2.7. cm., with numerous lymph nodes, along the small curvature and in the peripancreatic region, up to 1.5 cm in size. There was close contact between the changed and tumorous posterior wall of the stomach and the anterior surface of the pancreas. Neoplasm of the stomach was found that had infiltrated the body and tail of the pancreas and spleen hilum. Infiltration of the left crura of the diaphragm was also found, ex tempore biopsy showed inflammatory infiltration without elements of neoplasm. Total gastrectomy with omentectomy, and subtotal pancreatectomy and splenectomy were performed. Definitive patho-histological diagnosis confirmed MALT lymphoma of the stomach with pancreas infiltration, but no tumor cells were found on the spleen. Additional staining and immunohistological examination of the specimen from the community hospital showed that this was a misdiagnosis of carcinoma, and the specimen also contained MALT lymphoma. Discussion: MALT lymphoma frequently occurs in the stomach. For patients with MALT, systematic staging is indicated. If MALT is considered in the differential diagnosis, multiple random systematic biopsies within the stomach wall are needed to optimize diagnostic accuracy. Samples should be subject to immune phenotype analysis6. The main tumor cells of MALT are: CD 20+, CD 5-, CD 10-, CD 23-, CD 43+-. It is obvious that this kind of analysis cannot be accomplished in a small community hospital in a poor country such as Bosnia and Herzegovina, and suspicion of MALT indicates referral to a tertiary center. Although the long term risk of transformation of MALT lymphoma into the aggressive form is low9, this case of the aggressive form of MALT indicates the importance of systematic staging.

Key words: Mucosa-associated lymphoid tissue lymphoma, Helicobacter pylori, adenocarcinoma.

1. INTRODUCTION

MALT lymphoma accounts for 7-8% of all B-cell lymphomas and at least 50% of primary gastric lymphomas, with the highest incidence between 50 and 60 years of age (1).

Simultaneous occurrence of gastric carcinoma with omental MALT lymphoma is very rare (2). Both primary gastric carcinoma and MALT lymphoma are thought to be associated with H. pylori infection (3). Helicobacter pylorus is an important causative mechanism for the development of gastric MALT syndrome, with a positive rate of 90% in the early phase of gastric MALT syndrome (4).

Diagnosis should be confirmed by biopsy. Conventional pinch biopsies may miss the diagnosis, since gastric MALT can infiltrate the submucosa without affecting the mucosa (5). We report a patient with diagnosed gastric carcinoma, but the definitive diagnosis was MALT lymphoma with infiltration of the pancreas. Multivisceral resection of the stomach, pancreas and spleen was performed.

2. CASE REPORT

The patient, 33 years old, was admitted to the tertiary hospital as biopsy had confirmed adenocarcinoma of the stomach. Gastroscopy and biopsy were performed at a small community hospital (Figure 1). She had weight loss and epigastric pain. She was Helicobacter pylori positive. CT showed thickening of the fundus and corpus wall, up to 2.7. cm, with numerous lymph nodes along the small curvature and in the peripancreatic region, up to 1.5 cm. There was close contact between the changed tumorous posterior wall of the stomach and the anterior surface of the pancreas (Figure 2).

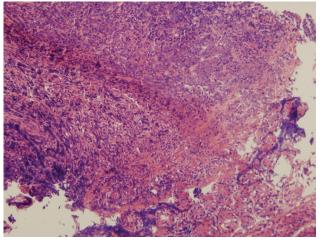


Figure 1. The biopsy specimen diagnosed as the adenocarcinoma. The lymphoma cells are centrocyte-like, with clear cytoplasm leading to a mocytoid appearance (HE 20x)



Figure 2. CT thickening of the fundus and corpus wall, up to 2.7 cm.

The patient was transferred to the University hospital. Neoplasm of the stomach was found that infiltrated the body and tail of the pancreas and spleen hilum (Figure 3). Infiltration of the left crura of the diaphragm was also found, and ex tempore biopsy showed inflammatory infiltration, without the elements of neoplasm.

Total gastrectomy with omeentectomy, subtotal pancreatectomy and splenectomy were performed. The postoperative period was normal and the patient was discharged on the tenth day. Definitive patho-histological diagnosis confirmed MALT lymphoma of the stomach with pancreas infiltration, but no tumor cells were found on the spleen. Additional staining and immuno-

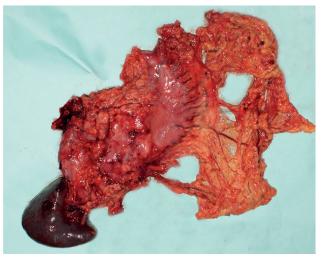


Figure 3. Specimen from the stomach with the body and tail of pancreas and spleen.

histological examination of the specimen from the community hospital showed that it was a case of the misdiagnosis of carcinoma, and the specimen also contained MALT lymphoma (Figure 4 and 5). Chemotherapy was prescribed for the patient.

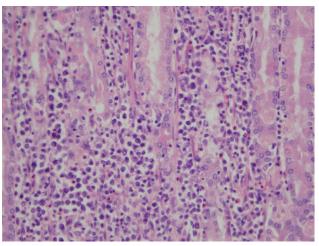


Figure 4. Lymphoepithelial lesions. The cells of this MALT lymphoma have clear cytoplasm leading to a monocytoid appearance (HE 40x)

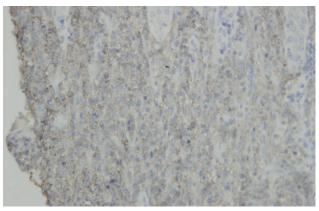


Figure 5. Immunophenotyping shows tumor cell positivity for CD20 B-cell marker (CD20 40x)

3. DISCUSSION

MALT lymphoma frequently occurs in the stomach. For patients with MALT, systematical staging is indicat-

ed. If MALT is considered in the differential diagnosis, multiple random systematic biopsies within the stomach wall are needed to optimize diagnostic accuracy. Samples should be subject to immune phenotype analysis6. The main tumor cells of MALT are: CD 20+, CD 5-, CD 10-, CD 23-, CD 43+-.

It is obvious that this kind of analysis cannot be accomplished in a small community hospital in a poor country such as Bosnia and Herzegovina, and suspicion of MALT indicates referral to a tertiary center.

MALT lymphoma is thought to be associated with H. pylori infection (3). Helicobacter pylori infection leads to the formation of lymphoid tissue within the stomach, and lymphomagenesis (6). T cell dependence explains the tendency of gastric MALToma to remain localized in the stomach (7).

However, in this case pancreas infiltration was found. Although the long term risk for transformation of MALT lymphoma into the aggressive form is low (8), this case of the aggressive form of MALT indicates the importance of systematic staging.

The US National Comprehensive Cancer Network recommends eradication of H. Pylori as the first line of treatment for H. pylori positive gastric MALT lymphoma, and radiotherapy as the second line of treatment for H. Pylori negative or refractory limphoma (9). Surgical resection of the superficial lesion is also an option, with low morbidity, and should be followed with low doses of chemotherapy. However in this case, multivisceral resection was performed without morbidity and mortality, and should be an option in the aggressive form of MALToma.

 Author's contribution: Edin Mulalic: substantial contribution to conception and design, substantial contribution to acquisition of data, substantial contribution to analysis and interpretation of data, drafting the article. Samir Delibegovic: critically revising the article for important intellectual content.

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