Cancer Association of South Africa (CANSA)

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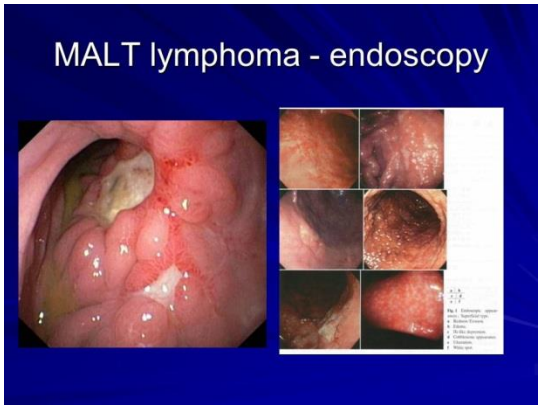
Fact Sheet on Mucosa Associated Lymphoid Tissue (MALT) Lymphoma

Introduction

Like all lymphomas, Mucosa Associated Lymphoid Tissue (MALT) Lymphoma, is a cancer of the lymphatic system which is part of the body's immune system. It develops when white blood cells, called B-lymphocytes, become abnormal and begin to grow in an uncontrolled manner.



[Picture Credit: MALT Lymphoma]

MALT Lymphoma

MALT lymphoma or Marginal Zone Lymphomas are B-cell lymphomas. They are not very common and account for a small percentage of non-Hodgkin's lymphomas (NHLs). There are 3 main types of marginal zone lymphomas:

- extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma). MALT lymphomas may also be called maltomas.
- Nodal marginal zone lymphoma
- Splenic marginal zone lymphoma

Extranodal marginal zone lymphomas start in tissues or organs outside of the lymph nodes (extranodal). MALT lymphoma develops in mucosa-associated lymphoid tissue, in the mucosa or tissue that lines body organs or body cavities including:

- gastrointestinal (GI) tract
  - The stomach is the most common location for MALT lymphoma, but it can also occur in the small bowel and colon
- lungs
- eyes, including the orbit (bony cavity that the eyeball sits in)
- skin
- salivary glands

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- thyroid gland
- breasts

Mullangi, S. & Lekkala, M.R. 2021.
"Mucosa-associated lymphoid tissue (MALT) lymphoma is a subtype of the marginal zone lymphoma (MZL), which is non-Hodgkin lymphoma. It is also called extranodal marginal zone lymphoma (EMZL). It is frequently noted in the stomach but virtually can involve any mucosal site like the salivary gland, ocular adnexa, lung, and elsewhere. EMZL is clinically distinct from the other types of MZL, the splenic marginal zone lymphoma (SMZL), and the nodal marginal zone lymphoma (NMZL) with specific diagnostic criteria, clinical behavior, different genetic features, and therapeutic implications. Other commonly used terminologies for MALT lymphoma are "MALToma," "MALT-type lymphoma," and "pseudo-lymphoma." In this review, we interchangeably use MALT lymphoma with EMZL."

Omote, R., Gion, Y., Omote, S., Tari, A., Tanaka, T., Nishikori, A., Yoshino, T. & Sato, Y. 2020.
"Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT) lymphoma of the stomach is mainly associated with Helicobacter pylori infection, and H. pylori eradication therapy is often effective. However, 20-30% of the cases of MALT lymphoma are resistant to the eradication therapy, and translocation of the API2-MALT1 gene is often found in these cases. Most cases without translocation of API2-MALT1 are localized to the stomach, whereas some cases with this translocation are a more advanced stage of MALT lymphoma that spreads to other organs. The c-Met receptor is a prognostic factor involved in infiltration and metastasis in many malignant tumors, including gastric, pancreatic, lung, and kidney cancer. In the present study, the expression of c-Met in 43 cases of gastric MALT lymphomas was immunohistochemically examined and compared with clinicopathological factors. To elucidate the significance of c-Met in MALT lymphoma, the expression intensity of c-Met in 22 API2-MALT1 translocation-positive and 21 API2-MALT1 translocation-negative cases was scored, compared, and examined. The immunohistochemistry analysis revealed strong staining for c-Met in 21 API2-MALT1 translocation-positive cases and in 1 translocation-negative case (P = 0.00). This result indicates the relationship between strong expression of c-Met and the progression of MALT lymphoma with API2-MALT1 gene translocation."

Incidence of MALT Lymphoma in South Africa
The National Cancer Registry (2017) does not provide any information regarding the incidence of MALT Lymphoma.

Signs and Symptoms of MALT Lymphoma
Symptoms depend on where the MALT lymphoma starts. The most common symptoms of MALT lymphoma that starts in the stomach is indigestion or heartburn. Some people also have weight loss, feeling or being sick and stomach (abdominal) pain.

In most people gastric MALT lymphoma is found during tests for persistent indigestion – although only a very small percentage of people with indigestion or heartburn will have lymphoma.

A few people with gastric MALT lymphoma go to their doctor with other symptoms, such as abdominal pain, nausea (feeling sick), vomiting (possibly with specks of blood in the vomit) and weight loss. Some people will have symptoms of anaemia, such as tiredness and shortness of breath,

because the stomach lining has been bleeding, but this is quite rare. Severe abdominal pain or the finding of a lump or mass in the abdomen would also be unusual.

Diagnosis of MALT Lymphoma
For a gastric MALT lymphoma to be diagnosed, the stomach lining has to be examined and biopsied. Tests for *H. pylori* infection are also needed to confirm the diagnosis.

Endoscopy - MALT lymphoma is usually discovered unexpectedly during an endoscopy examination of the stomach. This is a test in which a flexible tube with a light and a tiny camera in its tip is passed down through the mouth into the stomach.

Biopsy - It is very difficult, and often impossible, to tell the difference between lymphoma and the much more common kind of stomach cancer just by looking at these ulcers or nodules during the endoscopy examination, so small samples of the stomach lining – biopsies – will be taken during the endoscopy.

Cytogenetics tests. These specialised tests help the doctors to predict how well the lymphoma is likely to respond to therapy.

Testing for *H. pylori* organisms - It is very important that the presence of the *H. pylori* organism is confirmed so that a firm diagnosis can be made.

Salar, A. 2019.
"Marginal zone lymphomas of the MALT type are a type of B-cell neoplasms that involve extranodal tissues and have an indolent clinical behaviour. The stomach is the most common site and most patients are infected by Helicobacter pylori. An increase in the resistance of this bacterium to several antibiotics has been observed in the last years and this fact has determined the review of treatment guidelines. In areas with resistance to clarithromycin greater than 15%, classical triple therapy should be abandoned and quadruple regimens with or without bismuth are currently recommended. Thus, these new guidelines for eradication treatment should be applied to patients with gastric MALT lymphoma associated with H. pylori infection."

Staging of MALT Lymphoma
The aim of staging is to assist the treating physician in deciding on the treatment regimen.

Treatment of MALT Lymphoma
The usual initial treatment for gastric MALT lymphoma is a course of treatment to eradicate the *H. pylori* infection. This will successfully treat the lymphoma in most people who showed evidence of *H. pylori* infection in their tests.

Treatment is usually most successful when the tumour has not extended very far through the stomach wall and has not spread to the lymph nodes.

Yan, W., Wu, V., Liao, A-J., Yang, W. & Wang, H-H. 2021.
"Primary pulmonary mucosa-associated lymphoid tissue (MALT) lymphoma is rare, and the optimal frontline treatment has not taken shape so far. It is still debatable whether the watch-and-wait (W&W) policy is beneficial to patients, especially in the early stage. This study was to compare the efficacy of W&W with rituximab single agent or combined chemotherapy (R/R-Chemo) on primary pulmonary MALT patients with localized disease. Clinical characters and effect on 28 patients with primary pulmonary MALT (IE phase) were analyzed. Among the 28 patients, 14 were grouped into W&W cohort, and 14 were immediately treated with R/R-Chemo. The median follow-up duration was 62 months. The estimated median time to treatment failure (TTF) in the W&W cohort and immediate R/R-Chemo cohort was 29 months and 59 months, which were not significantly different (P = 0.667). The estimated median time of overall survival (OS) in the W&W cohort and immediate R/R-Chemo cohort was 78 months and 76 months, which were also not statistically significant (P = 0.696). Concerning prognosis, there is no difference between patients with primary pulmonary MALT (IE phase) treated with W&W and with timely R/R-Chemo."

Follow-up Upon Completion of Treatment
After the course of eradication therapy the patient should have a test (usually a breath test) to check for *H. pylori* about 4–6 weeks after the treatment has finished. About 3–6 months after the treatment has finished the patient will usually have another endoscopy.

About Clinical Trials
Clinical trials are research studies that involve people. They are conducted under controlled conditions. Only about 10% of all drugs started in human clinical trials become an approved drug.

Clinical trials include:

- Trials to test effectiveness of new treatments
- Trials to test new ways of using current treatments
- Tests new interventions that may lower the risk of developing certain types of cancers
- Tests to find new ways of screening for cancer

The South African National Clinical Trials Register provides the public with updated information on clinical trials on human participants being conducted in South Africa. The Register provides information on the purpose of the clinical trial; who can participate, where the trial is located, and contact details.

For additional information, please visit: www.sanctr.gov.za/

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http://www.cancerresearchuk.org/about-cancer/type/non-hodgkins-lymphoma/about/types/mucosaassociated-lymphoid-tissue-lymphoma

Chronic Illness Support
http://patient.info/doctor/non-hodgkins-lymphoma-pro

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