



# A Rare Case of Mucosa Associated Lymphoid Tissue (MALT) Lymphoma in the Esophagus

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## Abstract

We report a very rare case of primary Mucosa Associated Lymphoid Tissue (MALT) lymphoma of the esophagus. A middle-aged man was referred to our hospital for further examination and treatment of oesophageal tumor. Although a physical examination and laboratory data showed no significant abnormalities, endoscopic observation revealed slightly elevated submucosal tumour-like lesions in the esophagus, and CT show esophageal cancer mainly. However tissue lesions were composed of diffuse small atypical lymphoid cells that is, centrocyte like cells-which were stained with positive for CD20, CD19, CD43 MUM-1 and Bcl-2, and negative for CD3, CD5, CD10, Bcl-6, and cyclin D1. Therefore it was diagnosed as mucosa associated lymphoid tissue lymphoma of the esophagus. For treatment, it is helpless by eradication of *H. pylori*, but useful by chemotherapy by R-CHOP.

Lymphoma arises not only from lymphoid tissue, such as in the tonsil or Peyer's patches, but also from the stomach that normally contains no lymphoid tissue. Extranodal lymphoma occurs most commonly in the gastrointestinal tract, which accounts up to 5% to 20% of all cases [1]. Mucosa-Associated Lymphoid Tissue (MALT) lymphoma is a subtype of extranodal lymphoma [2]. It is reported that MALT lymphomas occurring usually in other parts of the gastrointestinal tract, such as the gastric, duodenum, ileum, and colon, but infrequently in the esophagus [3,4]. Here we report a rare case of MALT lymphoma in the esophagus.

## Case Presentation

A 46 year man was referred to Hainan General Hospital in April 2019 for further examination and treatment of oesophageal neoplasm found by chest CT and upper gastrointestinal endoscopy in other hospital. His major discomfort was feeling of choking after swallowing solid food but not porridge or water for more than two months without fever, weight loss and night sweating. On admission, physical examination revealed that the right neck can be touched a mass about 8 cm × 8 cm in size, otherwise the liver, spleen, and superficial lymph nodes were not palpable. The patient's main laboratory data showed no abnormalities. No hepatitis B surface antigen, hepatitis C virus antibody, Epstein-Barr virus capsid antigen, or Syphilis antibody were detected. No elevation of tumour markers not autoimmune antibodies were observed. The level of anti-*Helicobacter Pylori* (HP) antibody was elevated.

Cervicothoracic & upper abdominal CT scan typed: Esophageal space occupying, was considered tumorous lesions, mainly for esophageal cancer, which showed tissue mass 10.2 cm × 6.5 cm in maximum cross section located in middle and upper esophagus, with multiple mediastinal lymph node enlargement. Ultrasound endoscopy was not used to check because of big oesophagus mass, so endoscopic examination revealed: A longitudinal irregular mass was located at approximately 17 cm to 40 cm away from the incisor teeth, sized in 1/2 of the ring lumen, showed smooth local surface, local nodular, surface ulceration, brittle texture (Figure 1).

To confirm a diagnosis of the lesions, tissue specimens were obtained by endoscopic mucosal resection of the esophagus using a cap fitted panendoscope (GIF-Q240; Olympus, Tokyo, Japan). Pathology showed squamous epithelial hyperplasia with obvious atypia, but subepithelial rich lymphoid tissue proliferation at fist (Figure 2). Immunostaining was positive for CD20, CD19, CD43 MUM-1 and Bcl-2, and negative for CD3, CD5, CD10, Bcl-6, and cyclin D1. The esophageal mass was diagnosed as MALT lymphoma. Mucosa Associated Lymphoid Tissue (MALT) lymphoma in the esophagus involving the parotid gland in Figure 3 and 4.

The patient refused to take further examination due to his financial cause. Because of a positive HP antibody test, a *Helicobacter pylori* eradication regimen was begun, which consisted of

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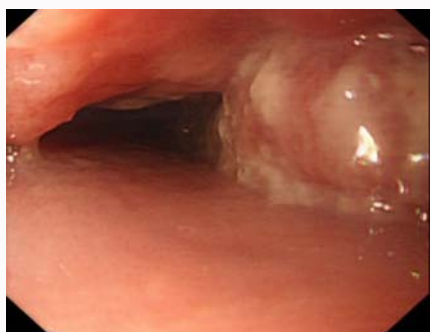
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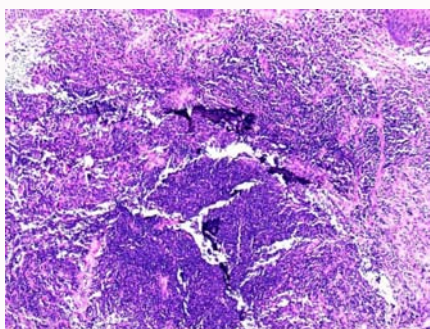
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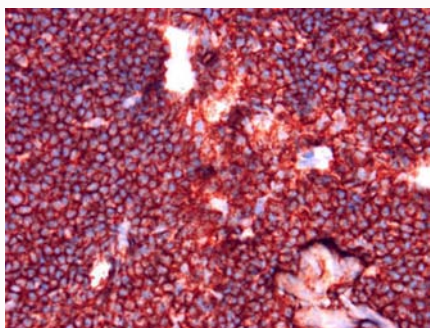
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**Figure 1:** The lesion in the Oesophagus under endoscopy Mass showed smooth local surface, local nodular, surface ulceration, brittle texture.



**Figure 2:** The pathological manifestation under light microscopy, HE staining, 40x magnification, squamous epithelial hyperplasia with obvious atypia, but subepithelial rich lymphoid tissue proliferation at fist.



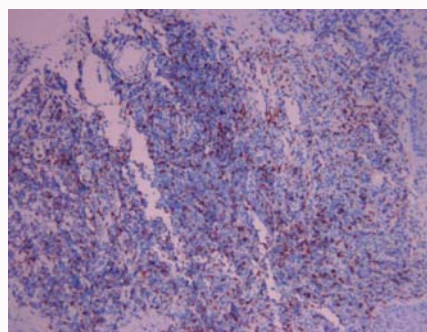
**Figure 3:** There is also a significant infiltrating of CD2-positive B cells, a characteristic feature of MALT lymphoma (immunoperoxidase).

clindamycin 500 mg twice a day, esomeprazole 20 mg twice a day, amoxicillin 1,000 mg twice a day and bismuth 150 mg quid a day for fourteen days. After two-week treatment, endoscopy that there was no change. Two months later, the patient was treated with R-CHOP chemotherapy (rituximab, cyclophosphamide, vincristine, adriamycin and dexamethasone). There was no evidence of tumor deterioration by CT.

## Discussion

Isaacson and Wright described a distinctive type of extranodal B-cell lymphoma, which was called “mucosa-associated lymphoid tissue lymphoma” in 1983 [2]. Primary extranodal malignant B cell MALT lymphomas involve mainly the gastrointestinal tract, as the largest group of all extranodal lymphomas [5].

Although MALT lymphoma may arise from any organ, the



**Figure 4:** CD3 staining of the mass. There is also a significant infiltrating of CD3-negative B cells, a characteristic feature of MALT lymphoma (immunoperoxidase).

esophagus is a very rare site of origin. Currently, there is no guideline for the treatment of MALT lymphoma in the esophagus. In current guidelines and reports, the first-line approach in gastric MALT lymphoma, regardless of clinical stage, is *H. pylori* eradication therapy [6]. More than 75% of patients can achieve regression after the successful eradication of *H. pylori*. However, some factors have been reported to be associated with eradication resistance, including: an endoscopic finding of non-superficial type

(mass-forming or diffuse in infiltrating type), the presence of t(11;18) (q21;q21) chromosomal translocation, location in proximal or multiple areas (non-antral involvement), the absence of *H. pylori* infection, and a Diffuse Large B-Cell Lymphoma (DLBCL) component [7]. In this case, eradication of *H. pylori* was helpless for treatment of MALT lymphoma in the esophagus. The patient accepted R-CHOP chemotherapy in another hospital, which was reported to be helpful for MALT lymphoma treatment.

The precise biological mechanism and clinical characteristics of MALT lymphoma in the esophagus remains unclear, thus further study is needed. It is necessary to accumulate information for further investigating patients with primary oesophageal MALT lymphoma.

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