

## Extremely rare case of primary esophageal mucous associated lymphoid tissue lymphoma

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### Abstract

SJ is a 37-year-old male who presented with one year history of dysphagia, odynophagia and 15 pounds weight loss. He underwent endoscopic evaluation which showed mid esophageal ulcers. It was thought that the cause of the ulcer was the multivitamins and the patient was asked to stop them. Furthermore Esomeprazole therapy was also initiated. Patient's symptoms persisted but he did not seek any medical attention until about one year later. Meanwhile the patient reported additional 15 pounds of weight loss. We repeated upper endoscopy again which showed evidence of two chronic non bleeding irregular friable ulcerations seen in the mid esophagus, 31 cm from the incisors. Biopsies and frozen section were taken and sent for assessment to the Pathology lab. Immunoperoxidase studies on frozen sections showed the presence of IgM and for

most plasma cells IgG. The microscopic and histologic findings were consistent with mucous associated lymphoid tissue lymphoma with plasmocytic differentiation. Computed tomographic scan done showed no evidence of spread to adjacent structures. The patient was referred to oncology and several cycles of radiation and Rituximab therapy were initiated which cured the disease. Subsequent endoscopies with blind biopsies were done which were negative for any neoplastic process.

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**Key words:** Lymphoma; Mucous associated lymphoid tissue; Esophagus

**Core tip:** This is a case of a 37-year-old gentleman presenting with chronic esophageal ulcers. Endoscopic biopsy samples were taken. Immunohistochemistry and hematoxylin and eosin staining of the biopsy samples were consistent with mucous associated lymphoid tissue (MALT) lymphoma. There was no sign of disease process in the stomach, and radiological studies revealed no evidence of metastasis. In consideration of these findings a diagnosis of primary esophageal MALT lymphoma was made, which is an extremely rare occurrence. On the basis of our experiences we recommend keeping primary esophageal MALT lymphoma in the differential diagnosis of chronic esophageal ulcers that are resistant to conservative management.

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### INTRODUCTION

We present a very rare case of primary esophageal B cell mucous associated lymphoid tissue (MALT) lymphoma.



**Figure 1** Photograph of esophagogastroduodenoscopy showing two ulcers at approximately 8 o'clock and 5 o'clock positions.

Gastrointestinal (GI) lymphoma is an uncommon disease but is the most frequently occurring extra nodal lymphoma and is almost exclusively Non-Hodgkin's type<sup>[1]</sup>. Esophageal lymphomas occur most often secondary to cervical and mediastinal lymph node invasion or contiguous spread from gastric lymphoma<sup>[1]</sup>. Primary esophageal lymphomas is a very rare condition accounting for less than one percent of GI lymphomas, with B cell lymphomas being the most common histological subtype<sup>[2]</sup>. The predominant presentation is that of sub mucosal infiltration but the tumor can also manifest as a polypoid mass into the lumen, ulceration or nodularity<sup>[3]</sup>.

We report a very rare case of primary esophageal MALT lymphoma in a middle aged patient.

## CASE REPORT

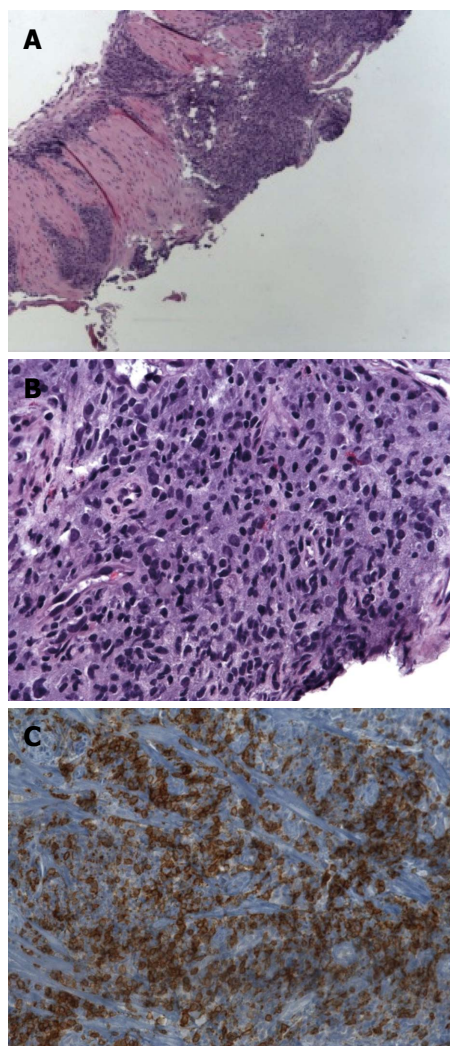
### Clinical presentation

The patient is a 37-year-old Indian gentleman, who presented with complains of difficulty swallowing, which started about a year ago. Shortly thereafter he started experiencing pain with swallowing as well. He was treated for three months with Esomeprazole and had resolution of his symptoms. However his symptoms recurred 6 mo later. An endoscopy was performed, and biopsies were taken. These slides were sent to be reviewed by pathology which indicated focally dense lymphoplasmacytic infiltrate with Kappa light chain excess, most consistent with a B cell MALT lymphoma with plasmacytic differentiation.

Previous medical history was significant for hypertension and recurrent bouts of sinusitis. Surgical history was significant for endoscopic surgery for sinusitis and appendectomy done several years ago. The patient had recently travelled to Pakistan, and denied any substance abuse. Medications included nifedipine (calcium channel blocker) and Losartan (angiotensin receptor blocker) for hypertension. Furthermore the patient's father had died from Non-Hodgkin's lymphoma of unknown histologic subtype. The rest of his immediate family members were alive and healthy.

### Endoscopic and microscopic findings

The endoscopy showed two superficial serpiginous fri-



**Figure 2** Photographs from the hematoxylin and eosin stain, and immunohistochemistry staining, of the biopsy samples. A: Photomicrograph shows atypical infiltrate under the mucosa of the esophagus  $\times 40$  original magnification hematoxylin and eosin (HE) stain; B: The tumor cells are medium sized lymphocytes and have a round or slightly constricted nuclei,  $\times 400$  original magnification, HE stain; C: Immunohistochemistry: CD20, CD3, CD43, Kappa and Lambda.

able chronic ulcers visualized 31 cm and 33 cm from the incisors, and measuring 1.2 cm and 1.5 cm respectively. Multiple core biopsies were taken from the affected region. The two ulcers seemed to be merged together with evidence of cicatrix formation between them. The gastro esophageal junction was at the level of 40 cm from the gums. The stomach, pylorus and the duodenum appeared to be normal. The endoscopic photograph of the ulcers is shown in Figure 1.

Hematoxylin and eosin (HE) staining of the cold biopsies, showed fragments of squamous epithelium with separate fragments of ulcer bed including acute and chronic inflammation with associated markedly atypical lymphoid infiltration. These findings were concerning for underlying lymphoma with superimposed ulcer bed. Photographs from the HE, and immunohistochemistry staining, of the biopsy samples are shown in Figure 2.

Additional immunoperoxidase staining showed small

aggregates of CD20 positive cells and a small number of CD3 positive cells within the nodules. Stains for immunoglobulin kappa and lambda light chains showed only very rare plasma cells positive for lambda, with the majority being positive for kappa. These microscopic and histological finding were most consistent with MALT lymphoma with plasmocytic differentiation.

A bone marrow biopsy and flowcytometry were also done, which were negative for any disease process.

### Radiology findings

Computed tomography (CT) scan showed mild thickening of the mid esophagus, most likely suggestive of a neoplasm. Except for an incidental finding of an 8 mm parenchymal density within the posteromedial right lung base, there were no other positive findings. A thoracic esophagram was also performed which showed adequate primary contractions observed fluoroscopically. The contrast was noted to flow freely into the stomach.

An endoscopic ultrasound was also done which showed diffuse obliteration of normal echo architecture of esophageal layers 1-3. No mediastinal, celiac axis or peripancreatic lymphadenopathy was observed.

### Diagnosis, treatment and follow up

The endoscopic findings were suggestive of esophageal ulcers with possible eosinophilic esophagitis. However Immunohistochemistry staining of the biopsy samples confirmed the diagnosis of MALT lymphoma and excluded eosinophilic esophagitis from the list of differentials. As there was no evidence of disease in the stomach on endoscopy, and in consideration of immunohistochemistry and HE staining a diagnosis of Primary Esophageal MALT lymphoma was made. EUS findings were in contrast to the usual presentation of esophageal lymphoma, that is a hypo echoic lesion<sup>[4]</sup>. A possible reason for this could be that the MALT lymphoma was diagnosed at an early stage.

On the basis of radiological, bone marrow biopsy and microscopic findings the tumor was staged as 1A. The patient received a 4 cycles of 36 Gy external beam radiotherapy. After radiation therapy he received four doses of Rituximab, as consolidative treatment.

After the treatment complete remission was achieved. The patient is seen at our institute regularly. Follow up endoscopies every 6 mo over last three years with targeted biopsies at the site of previous ulcers as evidenced by cicatrix formation have been negative.

### DISCUSSION

Lymphoid tissue neoplasms compromise a diverse yet closely related group of neoplasms, including hodgkin's lymphoma, non-hodgkin's lymphoma, multiple myeloma, MALT lymphomas and several other types.<sup>[5]</sup> MALT lymphomas can arise in various anatomic locations where lymphocytes are usually absent due to acquisition of MALT, including the gut, lung, thyroid, sali-

vary glands and liver<sup>[6]</sup>. Primary esophageal lymphoma is extremely rare<sup>[7]</sup>. Esophageal involvement by lymphoma is usually secondary to local spread from the stomach or the mediastinum<sup>[8]</sup>. Very few cases of primary esophageal lymphomas were reported in literature<sup>[6-11]</sup>. There are many morphological variants with most of the common being large B cell type and Non-Hodgkin's lymphoma<sup>[9]</sup>. We report a case of primary esophageal B cell MALT lymphoma in an immune competent patient. Only a few other cases of this particular morphological type of primary esophageal lymphoma have been reported in literature<sup>[7,12]</sup>.

Patients with acquired immunodeficiency syndrome (AIDS) are at an increased risk of developing malignant lymphomas, with the gastro intestinal tract being the most common site<sup>[13]</sup>. Chronic immunosuppression has also been suggested to be linked with the development of primary esophageal lymphomas<sup>[10]</sup>. MALT lymphomas of the stomach have closely been linked to Helicobacter Pylori infection, however no such relationship has been proven for primary esophageal MALT lymphoma<sup>[14]</sup>.

Esophageal cancer generally presents with symptoms of dysphagia and weight loss<sup>[15]</sup>. There has been a trend of increased incidence of esophageal cancers in population less than 55 years<sup>[15]</sup>. In the case that we report the patient presented with classical sign and symptoms but the rare nature of the histological classification of the disease made the diagnosis difficult to make.

In summation this case illustrates an instance of biopsy proven primary esophageal MALT lymphoma in a 37 years old man, with history of dysphagia, odynophagia and weight loss. The malignancy presented as persistent ulcers seen in mid esophagus, 31 cm from the incisors. Radiographic studies including CT scanning done at the time revealed localized disease without any spread to surrounding structures. The patient was referred to oncology where he received several cycles of radiation therapy. On subsequent follow ups the patient was found to be cured of the disease with normal endoscopic findings. Blind biopsy samples were taken from the esophagus which proved to be negative for any neoplastic process.

On the basis of our experience we suggest MALT lymphoma in the differential diagnosis of chronic esophageal ulcers that are resistant to conservative therapy.

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