



THE AGA KHAN UNIVERSITY

eCommons@AKU

Department of Surgery

Department of Surgery

February 2016

MALT lymphoma of the base of the tongue: a rare case entity

Haissan Iftikhar

Aga Khan University, haissan.iftikhar@aku.edu

Moghira Siddiqui

Aga Khan University, moghira.siddiqui@aku.edu

Khurram Minhas

Aga Khan University, khurram.minhas@aku.edu

Follow this and additional works at: http://ecommons.aku.edu/pakistan_fhs_mc_surg_surg

 Part of the [Otolaryngology Commons](#)

Recommended Citation

Iftikhar, H., Siddiqui, M., Minhas, K. (2016). MALT lymphoma of the base of the tongue: a rare case entity. *BMJ Case Rep*, 24(2), 1-3.

Available at: http://ecommons.aku.edu/pakistan_fhs_mc_surg_surg/217

CASE REPORT

MALT lymphoma of the base of the tongue: a rare case entity

Haissan Iftikhar,¹ Moghira Iqbaluddin Siddiqui,¹ Khurram Minhas²

¹Section of Otolaryngology, Department of Surgery, Aga Khan University Hospital, Karachi, Pakistan

²Department of Pathology and Laboratory Medicine, Aga Khan University, Karachi, Pakistan

Correspondence to

Dr Moghira Iqbaluddin Siddiqui,
moghira.siddiqui@aku.edu

Accepted 11 February 2016

SUMMARY

Lymphoma is a malignant tumour arising from lymphoid tissue, with the majority of cases being in the lymph nodes, however, in 1/4th of cases, these tumours are found in extralymphoid tissue. Lymphoid tissue is also found in organs having mucosa, such as the digestive tract, salivary gland and in tracheal tissue. This collection of lymphoid tissue is known as mucosa-associated lymphoid tissue (MALT), and non-Hodgkin lymphoma involving this extralymphoid lymph tissue is known as MALT lymphoma. It was first reported by Isaacson and Wright in 1983, however, it was not included as a working diagnosis in clinical use until it was reclassified as 'marginal zone B-cell lymphoma' in a 1994 Revised European American Lymphoma (REAL) classification. It is rarely seen in the head and neck region, and we report the sixth case of MALT lymphoma of the base of the tongue. A 61-year-old man presented with dysphagia and the feeling of a lump in his throat for 5 months.

BACKGROUND

When operating on a hypertrophic lingual tonsil one should keep in mind the differential diagnosis. We believe MALT lymphoma is a very rare entity that merits reporting.^{1,2}

CASE PRESENTATION

A 61-year-old man presented with difficulty in swallowing and the feeling of a lump in throat. He had been experiencing these progressively worsening symptoms for 5 months; he had no change in voice and no difficulty breathing. He had a 30-year history of smoking. Oral examination revealed enlarged lingual tonsils bilaterally. Other ear, nose and throat (ENT) examination was unremarkable.

INVESTIGATIONS

Fibreoptic laryngoscopy performed in the outpatient clinic showed two well-defined masses on the base of the tongue, pushing the epiglottis posteriorly, obscuring the glottis and making it difficult to visualise the airway. CT-scan showed a bilateral lingual tonsillar mass measuring 3.2×2.3 cm.

DIFFERENTIAL DIAGNOSIS

- ▶ Lingual thyroid
- ▶ Lingual dermoid
- ▶ Lingual tonsillar hypertrophy
- ▶ Minor salivary gland tumour.

TREATMENT

The patient was electively taken to surgery for excisional biopsy of the lesion, with a working

diagnosis of hypertrophic lingual tonsils. Endotracheal intubation was attempted, however, it was unsuccessful, and tracheostomy was performed to secure the airway. The lesion (**figure 1**) was soft and friable, pushing the epiglottis posteriorly and obscuring the glottis. Excision was attempted on the mass, but it was very friable, raising a clinical suspicion of malignancy. A frozen section was sent, which was inconclusive. Histopathology revealed extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma). Immunohistochemical stains showed diffuse positivity for Pan B (CD20), Pan T (CD3) and CD5, immunohistochemical stains highlighting admixed mature T lymphocytes and Cyclin-D1, CD10 and CD23 immunohistochemical stains were negative (**figures 2 and 3**).

OUTCOME AND FOLLOW-UP

The patient underwent eight cycles of CHOP therapy over 9 months and was then followed in ENT clinic for the next 10 months on a monthly bases in the outpatient clinic, with no evidence of recurrence in this follow-up period.

DISCUSSION

Lymphomas of the head and neck account for about 3% of all lymphomas.³ MALT lymphoma of the head and neck region is the second most common after gastrointestinal tract (GIT),¹ however, MALT lymphoma of the tongue is a rare entity and has only been reported in a few cases.¹ MALT lymphomas arise on a background of chronic pre-existing

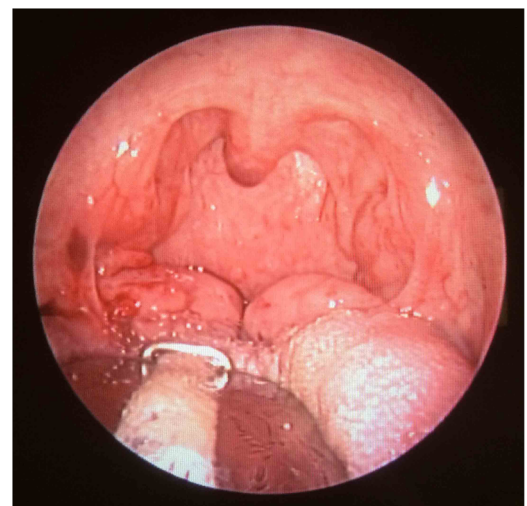


Figure 1 Intraoperative view of the lesion on both sides of the base of the tongue, as seen orally.



CrossMark

To cite: Iftikhar H, Siddiqui MI, Minhas K. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2015-213830

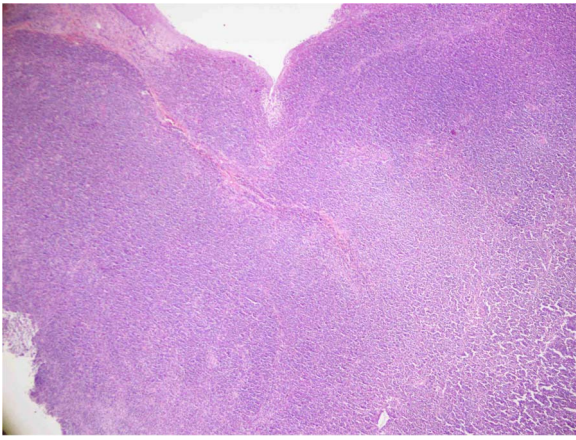


Figure 2 Lymphoid tissue exhibits effaced architecture with preserved surface squamous epithelium. Cells are arranged in diffuse sheets with a vague nodular pattern.

inflammatory disorders such as *Helicobacter pylori* infection of the stomach or an autoimmune disorder.³

MALT tissue specialises in protection of mucosa that is in direct contact with the external environment, unlike peripheral lymph nodes, which deal with antigens presented by afferent lymphatics.⁴ Peyer's patches in the terminal ileum are a good example of the true architecture of MALT tissue, which consists of a centre follicle surrounded by a marginal zone and clusters of marginal cell epithelium covering the epithelium.⁵ In the head and neck region, they are commonly seen in the Waldeyer's ring and mostly involving the Palatine tonsils.⁶ In the salivary glands, it is most commonly found in the parotid gland and is associated with Sjögren's syndrome.³

Two-thirds of cases are found in the GIT whereas the rest have been reported in various sites such as the lungs, pharynx, larynx, trachea, thyroid and salivary glands.⁶ MALT lymphomas have certain hallmark features such as (1) centrocyte-like tumour cells, (2) follicular colonisation, (3) lymph epithelial lesions and (4) plasma cells.^{7 8} No specific marker is linked with MALT lymphomas, however, they are known to express CD 20, CD 21, CD 32, CD 35, CD43 and bcl-2, and they are defined by the absence of CD 5, CD 10 and CD 23.⁶

Malt lymphomas have an inert natural course of disease,³ they tend to remain in remission but at the time of relapse they tend to occur in other extranodal sites.³ They have a better

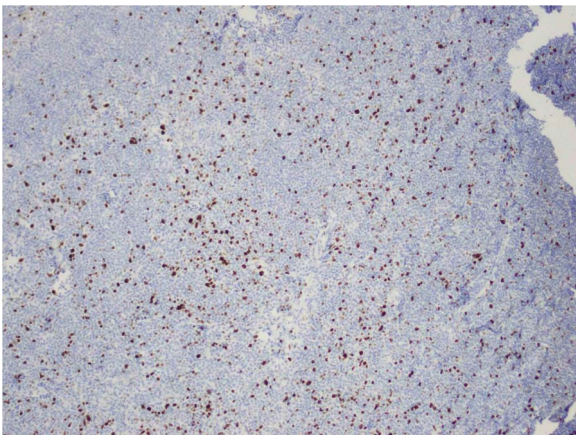


Figure 3 Sheets of cells show diffuse positivity for Pan B (CD20) immunohistochemical stain. Pan T (CD3) and CD5.

prognosis even when high grade, as compared to their nodal counterparts, since excision alone is suggested to be curative.^{9 10} Other authors have suggested radiotherapy to play the same role.¹¹ Deinbeck *et al*¹² reported to have cured 30 patients with marginal zone lymphoma with 40 Gy of radiation. Ueda *et al*¹³ reported extraintestinal MALT lymphoma to have a worse prognosis with multiorgan involvement than gastric MALT lymphoma, even after chemotherapy.

There have been five previously reported cases of MALT lymphoma of the tongue.^{1 3 14-16} One case was of a 62-year-old woman who underwent conservative treatment and developed systemic spread, and was then treated with chemotherapy 7 years after diagnosis,¹⁴ another was of a patient with a 1.9×1.0 cm tongue lymphoma who underwent surgical resection,¹⁶ two other cases were of 61-year-old and 80-year-old women who underwent only surgical resection,^{3 15} and the most recently reported case was of a 29-year-old woman with a history of T-cell lymphoma, developing MALT lymphoma at the base of her tongue 3 years later, successfully treated with radiotherapy.¹

There does not seem to be a consensus on the treatment of MALT lymphoma, especially when found on the tongue.⁶ In our case, the patient underwent surgical debulking followed by chemotherapy, and remained disease-free 10 months after completion of treatment.

To the best of our knowledge, ours is the sixth reported case of MALT lymphoma of the tongue.

Learning points

- ▶ Hypertrophic lingual tonsils may well be malignant, and one should consider mucosa-associated lymphoid tissue lymphoma in the differential diagnosis.
- ▶ Endotracheal intubation may be difficult with a mass at the base of the tongue, a retroflexed epiglottis, hence tracheostomy should be considered.
- ▶ We suggest a biopsy for suspicious cases of tonsillar hypertrophy.

Contributors HI contributed substantially to the conception and design of the work, and the acquisition, analysis and interpretation of data for the work. MIS critically revised the manuscript for important intellectual content and gave final approval of the version to be published. KM provided the histopathological diagnosis and the microscopic images.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Song JH, Sun DI, Hong YS, *et al*. MALT lymphoma at the base of tongue of a 29-year-old woman treated with radiation therapy alone. *J Cancer Res Ther* 2014;10:407.
- 2 Zucca E, Conconi A, Pedrinis E, *et al*. Nongastric marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue. *Blood* 2003;101:2489–95.
- 3 Goteri G, Ascani G, Filosa A, *et al*. Primary malt lymphoma of the tongue. *Med Oral Patol Oral Cir Bucal* 2004;9:461–3; 459–61.
- 4 Zucca E, Roggero E. Biology and treatment of MALT lymphoma: The state-of-the-art in 1996. *Ann Oncol* 1996;7:787–92.
- 5 Isaacson PG. Pathogenesis and early lesions in extranodal lymphoma. *Toxicol Lett* 1993;67:237–47.
- 6 Lee JT, Paquette R, Sercarz JA, *et al*. Mucosa-associated lymphoid tissue lymphoma of the lingual tonsil. *Am J Otolaryngol* 2000;21:271–6.
- 7 Mori N, Yatabs Y, Asai J. Mucosa associated lymphoid tissue (MALT) lymphoma. *Pathol Int* 1995;45:544–51.

- 8 Isaacson P, Spencer J. Malignant lymphoma of mucosa associated lymphoid tissue. *Histopathology* 1987;11:445–62.
- 9 Zinzani PL, Magagnoli M, Galieni P, *et al.* Nongastrointestinal low-grade mucosa-associated lymphoid tissue lymphoma: analysis of 75 patients. *J Clin Oncol* 1999;17:1254–1254.
- 10 Ferrer A, López-Guillermo A, Bosch F, *et al.* Linfomas del tejido linfoide asociado a mucosas (MALT) de localización extragástrica: análisis de 14 casos. *Med Clin (Barc)* 1999;112:577–80.
- 11 Tsang RW, Gospodarowicz MK, Pintilie M, *et al.* Localized mucosa-associated lymphoid tissue lymphoma treated with radiation therapy has excellent clinical outcome. *J Clin Oncol* 2003;21:4157–64.
- 12 Deinbeck K, Geinitz H, Haller B, *et al.* Radiotherapy in marginal zone lymphoma. *Radiat Oncol* 2013;8:2.
- 13 Ueda K, Terui Y, Yokoyama M, *et al.* Non-gastric advanced mucosa-associated lymphoid tissue (MALT) lymphoma has worse prognosis than gastric MALT lymphoma even when treated with rituximab-containing chemotherapy. *Leuk Lymphoma* 2013;54:1928–33.
- 14 Ferry JA, Yang WJ, Zukerberg LR, *et al.* CD5+extranodal marginal zone B-cell (MALT) lymphoma. A low grade neoplasm with a propensity for bone marrow involvement and relapse. *Am J Clin Pathol* 1996;105:31–7.
- 15 Sakabe H, Bamba M, Nomura K, *et al.* MALT lymphoma at the base of the tongue developing without any background of immunodeficiency or autoimmune disease. *Leuk Lymphoma* 2003;44:875–8.
- 16 Kuramoto T, Yao M, Ishikura Kano S. A case of mucosa-associated lymphoid tissue lymphoma at the tongue. *Kokubyo Gakkai Zasshi* 1995; 44:557.

Copyright 2016 BMJ Publishing Group. All rights reserved. For permission to reuse any of this content visit <http://group.bmj.com/group/rights-licensing/permissions>.
BMJ Case Report Fellows may re-use this article for personal use and teaching without any further permission.

Become a Fellow of BMJ Case Reports today and you can:

- ▶ Submit as many cases as you like
- ▶ Enjoy fast sympathetic peer review and rapid publication of accepted articles
- ▶ Access all the published articles
- ▶ Re-use any of the published material for personal use and teaching without further permission

For information on Institutional Fellowships contact consortiasales@bmjgroup.com

Visit casereports.bmj.com for more articles like this and to become a Fellow