*ejpmr, 2019,6(2), 561-565*

SJIF Impact Factor 4.897

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

# PRIMARY MUCOSA-ASSOCIATED LYMPHOIDE TISSUE LYMPHOMA OF THE TRACHEA, A VERY RARE DIAGNOSIS: ABOUT A CASE REPORT TREATED WITH RADIOTHERAPY

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**Article Received on 20/12/2018** **Article Revised on 10/01/2019** **Article Accepted on 31/01/2019**

## ABSTRACT

**Background:** Primary mucosa-associated lymphoid tissue (MALT) lymphoma of the trachea in a woman without any notable pathological history. **Methods:** Here we report the case of a 63-year-old woman without any pathological antecedents. Computed tomography of the chest showed a tumor process of the anterior wall of the trachea extended to the carina. Bronchoscopy was performed, and the results of a biopsy supported a diagnosis of mucosa-associated lymphoid tissue (MALT) lymphoma. No other lesions were detected in the systemic evaluation. The patient was fitted with an intra-tracheal prosthesis in Y and then treated with regional irradiation at a dose of 30Gy. **Results:** the patient presents a complete remission of her disease with the CT scan at 4 weeks of end of radiotherapy a disappearance of the tumor. **Conclusions:** external beam radiotherapy seems to be one of the most valuable strategies for treatment of MALT lymphomas of the central airway.

**KEYWORDS:** Mucosa-associated lymphoid tissue (MALT) lymphoma, trachea, radiotherapy.

## INTRODUCTION

Primary mucosa-associated lymphoid tissue (MALT) lymphoma is most commonly found in the stomach, lungs, orbital soft tissue, salivary glands and thyroid; involvement of the trachea is extremely rare.[1,2] it accounts for about 0.5% of all tracheal neoplasms.[3] Due to the paucity of lymphoid tissue found in the trachea, MALT rarely presents at this site. Common presenting symptoms include dyspnea on exertion, wheezing, cough or hemoptysis.[4] Current evidence suggests MALT's pathogenesis is likely related to either chronic immune reactions driven by external antigenic stimuli,[5] or indolent microbiologic infections.[6] There are no clear guidelines for the treatment of MALT lymphoma, and, with respect to lesions located in the trachea, a broad range of treatments have been shown to be effective, including surgical resection, radiotherapy, bronchoscopic therapy, chemotherapy, immunotherapy (rituximab) and immunochemotherapy.

## METHODS: CASE REPORT

A 63 year old woman with no history of smoking or chronic autoimmune disease, such as Sjögren's syndrome and Hashimoto's thyroiditis, or chronic infections, such as that with Helicobacter pylori. The patient complains of inspiratory, continuous, chronic and exertional dyspnoea without orthopnea, with a dry cough. Inspiratory and expiratory coarse rhonchi (stridor) were auscultated throughout over the lung, and large airway obstruction was suspected. On physical examination there was no clubbing, lymphadenopathy, or hepatosplenomegaly. Chest radiography and a laminagram showed narrowing of the thoracic trachea [fig 1], and a chest CT scan showed a mass protruding into the tracheal lumen extended to the carina [fig 2 (A and B)]. Bronchofibreoscopy identified a tracheal stenosis. Blood counts and chemistry were all within normal limits and electrophoretic analysis of the components of the serum proteins showed a normal pattern without any broad peaks in the c, b2, or a2 globulin regions. The bronchoscopic biopsy made it possible to establish the diagnosis of tracheal MALT, the histopathological and immunohistochemical analysis of the bronchoscopic biopsy specimen shows tumor cells expressing CD 20 and CD45, indicating an increase of B cells with a reactive response. CD5 and CD10 were negative, the final diagnosis was a low grade, B cell MALT lymphoma. Systemic screening including CT scans of the chest, abdomen and brain, a bone scan found no other abnormal lesions.

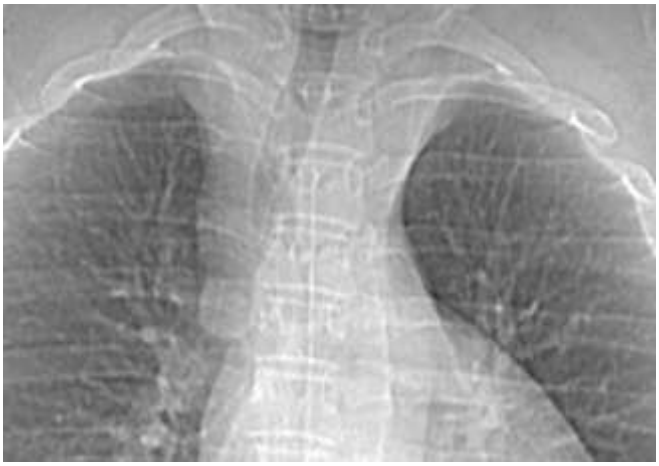
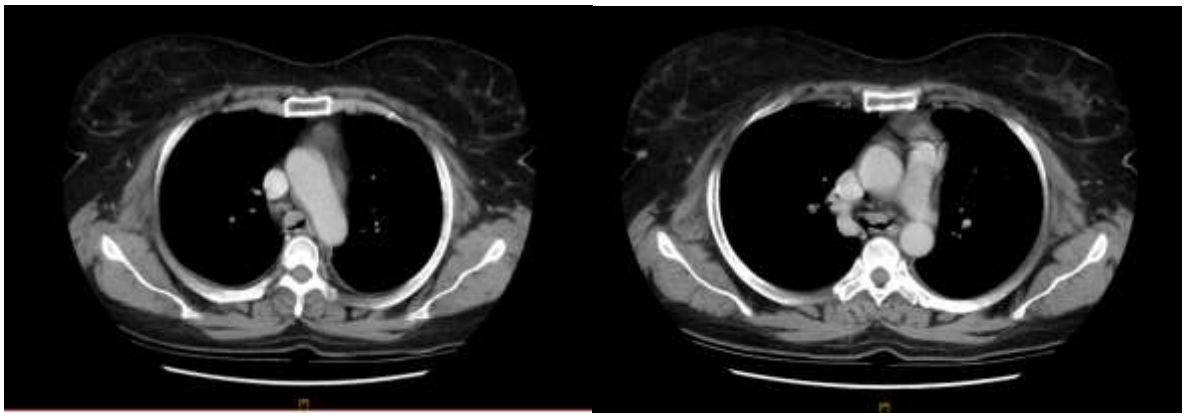


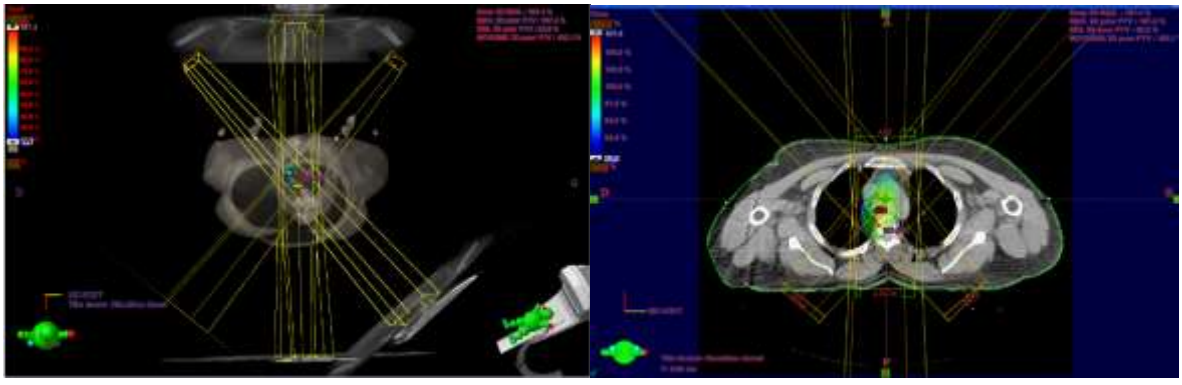
**Figure 1: Chest X-ray showing a mass protruding into the tracheal lumen.**



**A** **B**

**Figure 2: Chest computed tomography (CT) demonstrating irregularities of the tracheal wall and narrowing of the trachea.**

Because of the airway stenosis the patient benefited from the placement of a endotracheal prosthesis in Y, then she was transferred to our hospital where she received regional irradiation planned for a total dose of 30 Gy in 15 fractions, at a daily dose fraction of 2 Gy over 3 weeks using the conventional three-dimensional radiotherapy, with four irradiation fields (anterior, posterior and 2 posterior oblique fields) [fig 3 (A, B, C, D, E and F)]. the dose constraints to organs at risk have been respected, with optimal coverage and satisfactory target volumes [fig 4 (A, B, C and D)].

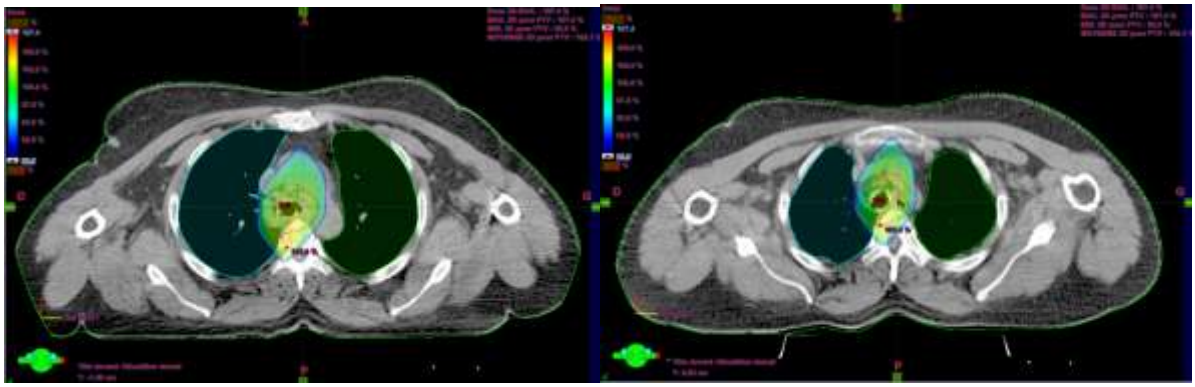


**A** **B**

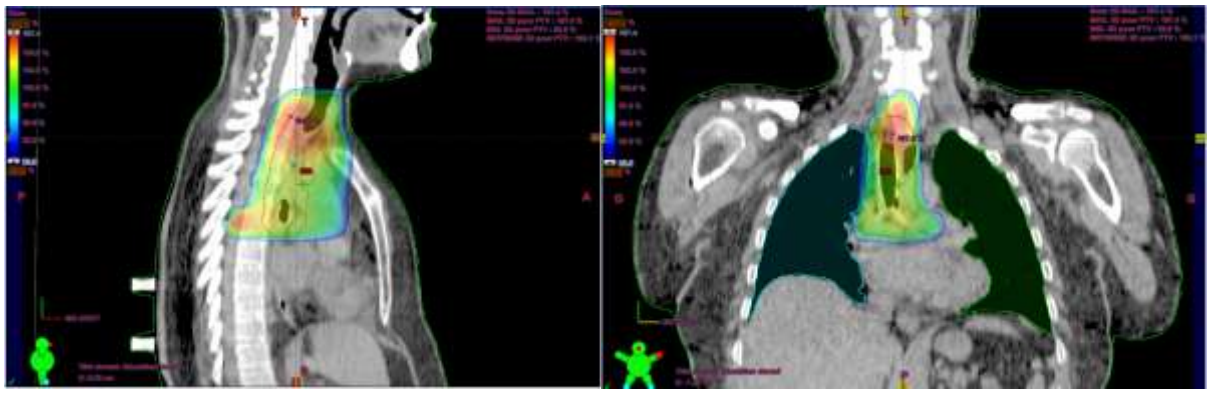
C D

E F

**Figure 3: Disposition of the four beams of irradiation**



A B

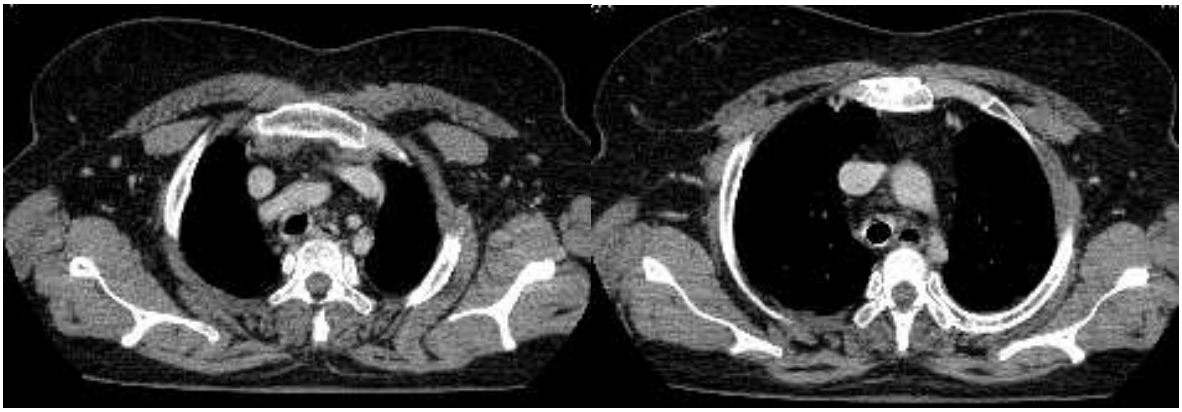


C D

**Figure 4: dose distribution on the three plans (axial, sagittal and coronal).**

**RESULTS**

The thoracic CT scan after the end of irradiation showed a disappearance of the tumor [fig 5 (A and B)], and the patient did not show signs of recurrence after 8 months.



**A** **B**

**Figure 5: CT scan sections of the thoracic CT control.**

**DISCUSSION**

MALT lymphoma was first described by Issacson and Wright in 1983 in a small series of patients with low-grade B cell gastrointestinal lymphoma,[7] and primary tracheal and bronchial tumors are rare. Gelder et al. reviewed 321 primary tracheal tumors and reported that 54.2% were squamous cell carcinoma (SCC), 10.6% were adenoid cystic carcinoma (ACC), and only 4 out of 321 (1.2%) were lymphoma.[8] Bronchus-associated lymphoid tissue (BALT) can be found in follicular bronchiolitis and can be associated with various autoimmune disorders, such as Sjögren's syndrome, but is not found in the normal lung.[9] As primary tracheal MALT lymphomas are extremely rare, most reports in the literature are case reports.[10-12]

The diagnosis of MALT lymphoma is based on the identification of morphologic, immunophenotypic, genotypic, and molecular features, including flow cytometry of the tumor (WHO recommendation). However, evaluating such features is difficult at times due to the small size of the specimens collected by bronchoscopy. In the case of Dincer et al., a second bronchoscopy was performed to obtain more specimens.[13] Because a molecular biological analysis uses fresh or frozen tissue samples, the freeze-preservation of biopsy tissue is important if MALT is suspected.

Previous reports have shown the effectiveness of a range of treatments, including surgical resection, radiotherapy, bronchoscopic therapy, chemotherapy, immunotherapy and immunochemotherapy, i.e., rituximab, cyclophosphamide, adriamycin, oncovin and prednisone (R-CHOP).[10-12] However, combination regimens have not been proven to be any more effective than single chemotherapy regimens.[14] Hitomi Y. and al. have treated the first case of MALT lymphoma of the trachea in a patient with breast cancer treated with prednisone alone. The details of this case suggest that prednisone monotherapy may be an effective strategy for treating MALT lymphoma.[15] Junji Tsurutani and al. have treated a patient with tracheal MALT by twice at an interval of 7 days with Nd-YAGlaser photoresection via a flexible fiberoptic bronchoscopeunder conscious sedation and local anesthesia in February 1996. The laser setting was 30 watts with a pulse duration of 1.0 second, with an overall total of 15,633 joules. The patient had no complications related to the laser treatment. Nd-YAGlaser photoresections were followed by two ethanol injections (1.6 ml in total) into a remnant of the tumor via a flexible bronchofiberscope in September 1996. The tumor disappeared completely, and a follow-up bronchoscopy one year later.[16] K Okubo and al. have treated a patient with tracheal MALT by a tracheal resection and reconstruction. The trachea was circumferentially divided proximal and distal to the tumour by median sternotomy of the upper region, and the affected portion of the trachea of six cartilaginous rings (3.0 cm) was resected. After frozen sections of the stumps were confirmed not to be malignant, the ends of the trachea were anastomosed with absorbable sutures (4-0 Vicryl). No paratracheal lymphadenopathy was identified. The postoperative course was uneventful and the patient has been well without any symptoms of recurrence for 53 months.[17] Takeshi Kawaguchi and al. reported two cases of patients with endobronchial MALT lymphoma; the first patient was asymptomatic and selected careful watchful waiting, the second was treated by radiotherapy, a total of 50 Gy was administered, resulting in a complete response of the tumor The patient was administered rituximab as consolidation therapy.[18]

MALT lymphoma of the trachea is an extremely rare and indolent disease and must be considered in the differential diagnosis of airway lesions. Tracheal tumors may mimic the features of asthma and chronic obstructive pulmonary disease (COPD) and should be

kept in mind as a rare cause of asthma and COPD-like symptoms. In this case, radiotherapy alone has been beneficial in the management of tracheal lymphoma in our patient.

**DECLARATIONS**

The authors state that they have no Conflict of Interest

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