A Review of Tracheal MALT-oma

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ABSTRACT:

Mucous associated lymphoid tissue lymphoma (MALT-oma) is a distinct group of non-Hodgkins lymphoma most commonly seen in the gastrointestinal tract, with head and neck manifestation being uncommon. We present the case of a 77 year old male with a three month history of increasing dyspnea misdiagnosed as asthma but subsequently found to have a MALT-oma of the distal trachea. Following tracheostomy and biopsy he was begun on treatment of his lymphoma. We discuss treatment options and present a literature review of this uncommon disorder.

INTRODUCTION:

Primary malignancies of the trachea are uncommon.[1,2] While the majority of these tumors are squamous cell or adenoid cystic carcinoma, lymphoma involving the trachea is distinctly unusual. [2] Extra-nodal lymphoma arising from mucous associated lymphoid tissue (MALT) was first described in 1984 by Isaacson and Wright.[3] These so called MALT-omas are a distinct group of non-Hodgkins lymphoma that often involve the gastrointestinal (GI) tract. In approximately 19% of cases, non-GI MALT-omas are found, in the breast, kidney, lung, and skin.[4] MALT-oma of the trachea is exceedingly rare with only 20 cases reported (scholar/google). We present the unusual presentation of such a case with a review of treatment options, and radiologic appearance.

CASE:

A 77 year old male presented to our tertiary medical center with a 3 month history of increasing stridor and dyspnea on exertion all of which were refractory to conservative asthma care. Of note is that he had a history of MALT lymphoma of the lacrimal duct treated in 1994

On flexible laryngoscopy, there was a large subglottic mass originating from the posterior tracheal wall.

The patient was taken to the operating theater for tracheostomy.

LITERATURE REVIEW:

Classically, MALT-omas exhibit an indolent course, with 5 year survival as high as 99% in low-risk groups. While non-gastric MALToma is well described in the literature, tracheal MALT-oma is exceedingly rare. In a review of seven series of tracheal tumors between 1930- 1989, only 1 case of non-Hodgkin's lymphoma was reported. Non-gastric MALToma is thought to occur along the various patches of mucous-associated lymphoid tissue scattered throughout the GI and aerodigestive tract.

CT Imaging

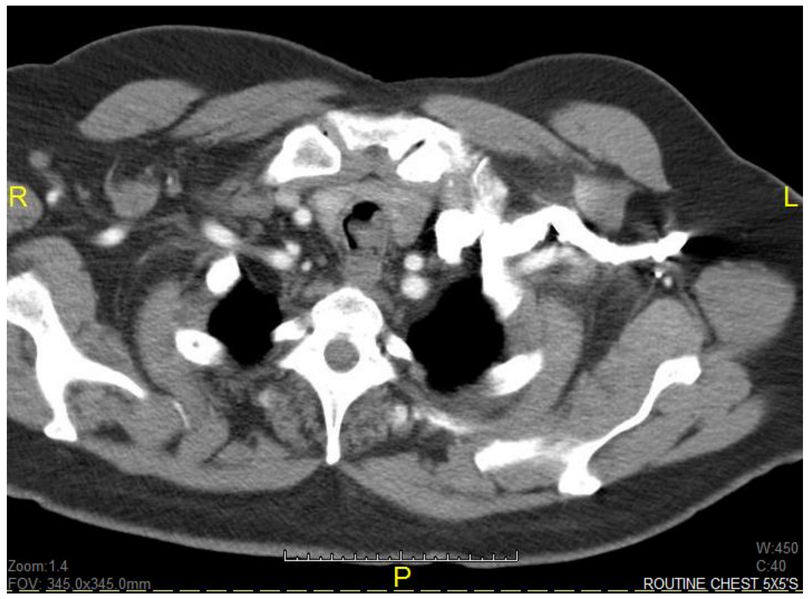

Figure 1: The above images represent the patient's chest CT significant for a 1.8 x 2.0 x 1.8 cm nodular density seen arising from the posterior wall of the trachea at the level of the thoracic inlet, with suspicion of a soft tissue tumor

Immunohistochemical Analysis

Antigen	Result	Strength
CD3	-	
CD5	+	Weak
CD10	-	
CD20	+	
CD23	-	
CD43	-	
PAX5	+	
BCL1	-	
BCL2	+	
BCL6	-	
Ki-67	+	

Karyotype of Tracheal Mass

48,XY,+3,+6,der(6;18)(p10;q10),del(8)(p21),del(9)(q21.2q22)[4]/46,XY

INTERPRETATION:

Chromosome analysis showed an abnormal male karyotype with numerical and structural abnormalities as described above in 4 of the 20 metaphase cells examined. The remaining cells were cytogenetically normal. The rearrangement (6;18) has led to an extra copy of the long arm of chromosome 18. Trisomy 3 and complete/partial trisomy 18 have been observed in marginal zone lymphomas of MALT type. Correlation with other clinical data is recommended.

DISCUSSION:

Non-Gastric MALToma is not associated with *H. pylori* and are treated with chemotherapy, radiation, and surgical debulking / resection. Tracheal MALT-oma typically presents with symptoms of airway obstruction, as seen in this case. To treat the obstructive symptoms, surgical intervention is usually required. This is either achieved by stenting, repeat bronchoscopy and debulking, or by tracheostomy. Acceptable pharmacologic treatment modalities for non-gastric MALT-oma includes: observation, radiotherapy, or chemotherapy. Once such chemotherapy regiment involved rituximab plus cyclophosphamide, doxorubicin-mitoxantrone, vincristine, and prednisolone irrespective of prior therapy.

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