Case Report

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Diffuse large B-cell lymphoma of thyroid gland with pyriform fossa extension presenting as emergency airway obstruction: a case report

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ABSTRACT

Primary thyroid lymphoma and laryngeal lymphoma are rare malignancies of the head and neck region. Airway obstruction caused by primary thyroid lymphoma is mainly by means of direct compression of trachea by huge thyroid mass. We present the first case report of diffuse large B cell lymphoma of thyroid gland with extension to pyriform fossa, in the absence of preexisting chronic thyroiditis. In this case, emergency airway obstruction was caused by direct extension of disease into pyriform fossa, occluding laryngeal inlet. We reviewed English literature between 1981 and 2018 for cases of primary thyroid lymphoma. There are no reported cases of primary thyroid lymphoma with direct extension into pyriform fossa. This presentation posed a great challenge in securing a patent airway by means of oral intubation and tracheostomy.

Keywords: Primary thyroid lymphoma, Pyriform fossa, Emergency airway obstruction

INTRODUCTION

Primary thyroid lymphoma is rare, comprising 1-2% of all extra-nodal lymphomas and 2-8% of all thyroid malignancies. Extra-nodal lymphoma of larynx is even rarer, accounting for less than 1% of all laryngeal tumors. We present the first case report of diffuse large B cell lymphoma of thyroid gland with extension to pyriform fossa, in the absence of pre-existing chronic thyroiditis. The diagnosis of diffuse large B-cell lymphoma was made by endoscopic biopsy of mass arising from right pyriform fossa as well as incisional biopsy of thyroid isthmus under general anaesthesia. The patient was treated successfully with chemotherapy.

CASE REPORT

A 50-year-old female presented with 3 -month history of anterior mid-line neck swelling which progressively

increase in size; worsening dyspnea for 2 months, odynophagia for 1 month and hoarseness of voice for 2 weeks. She did not seek professional medical advice previously but went to a district hospital when she was barely able to breathe. She was initially treated for bronchospasm in the aforementioned district hospital and her diagnosis was later revised as upper airway obstruction in tertiary hospital. Upon physical examination in tertiary hospital noted there was an anterior neck mass extending from level of hyoid bone suprasternal notch. Bedside flexible laryngoscope revealed huge mass over right pyriform fossa, obstructing laryngeal inlet and bilateral vocal cords.

Tracheostomy, direct laryngoscopy, esophagoscopy and biopsy was carried out the same day as emergency operation. Intra-operatively noted huge goitre obstructing surgical field (Figure 1 and 2). Upon thyroid

isthmectomy noted necrotic tissue and firm-to-hard consistency of isthmus. Airway study revealed fungating mass over right pyriform fossa (Figure 1 and 2), edematous right arytenoid and false cord. Unable to visualise bilateral true cords. Biopsies taken from right pyriform fossa and thyroid gland showed diffuse large B cell lymphoma, non-GC type.

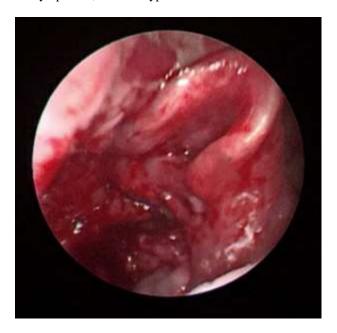


Figure 1: Intra-operative image showing mass over right pyriform fossa viewed during direct laryngoscope.



Figure 2: Closed up view of mass over right pyriform fossa.

Computer tomography of neck to abdomen revealed a well-defined homogenous enhancing right neck mass epicentre at level IV measuring 5.5×7.0×5.2 cm (APxWxCC). Direct infiltration into right thyroid gland

and this mass crosses midline to involve the contralateral thyroid. Involvement of right peritonsillar region, right pyriform fossa, aryepiglottic and right valeculla noted. Mass effect onto trachea and supraglottic region causing airway narrowing at the level of glottis, associated with regional cervical lymphadenopathy, Ann Arbor classification 1E.

Patient was subsequently transferred to hematology ward for chemotherapy. She received her first cycle of chemotherapy (R-CHOP) and responded well to treatment. On repeated indirect laryngoscopy three weeks later, no mass seen. Tracheostomy was successfully decannulated a week after the procedure.

DISCUSSION

Non-Hodgkin lymphomas usually present as extra-nodal disease. In head and neck region, Waldeyer's ring is the most common site for lymphoma, while para-nasal sinuses, salivary glands and thyroid gland (with underlying chronic thyroiditis) are common sites for extra-nodal lymphoma. Larynx is an uncommon site for non-Hodgkin lymphoma.

Primary thyroid lymphoma is rare, accounting for <5% of all thyroid malignancies and <2% of extra-nodal lymphomas. Females are most commonly affected.² Thyroid lymphomas are almost exclusively of the non-Hodgkin's, B-cell type, the most common type being diffuse large B-cell lymphoma (DLBCL), accounting for more than 50% of cases, followed by mucosa-associated lymphoid tissue (MALT) lymphoma, which represents about 10–23% of cases.^{3,4} A normal thyroid gland does not contain lymphoid tissue.⁵ Pre-existing chronic autoimmune thyroiditis (Hashimoto's thyroiditis) is a well recognized risk factor for development of the pathological intra-thyroid lymphoid tissue. 6-8 Although most thyroiditis cases do not proceed to lymphoma, most cases of lymphoma do arise in a background of thyroiditis, which is estimated to occur in approximately 60-90% of primary thyroid lymphoma cases. In our case, features of Hashimoto's thyroiditis are not detected clinically and histologically.

Primary thyroid lymphoma usually presents as painless, rapidly enlarging neck mass, with patients experiencing compressive symptoms such as dyspnea, dysphagia, stridor, and hoarseness of voice. B-symptoms such as weight loss, fever, and night sweats occur in up to 10% of patients.

Extra-nodal lymphoma of the larynx is even rarer, accounting for less than 1% of all laryngeal tumors. 9,10 The most common site of laryngeal lymphoma is supraglottis (47%) followed by glottis (25%). In our patient, the bulk of the lesion was found over thyroid gland and right pyriform fossa. This poses a dilemma in diagnosing diffuse large B cell lymphoma arising primarily in thyroid gland with extension to right pyriform fossa or its

contrary. We believe the former presentation is more probable based on patient's clerking history.

We reviewed English literature between 1981 and 2018 for cases of primary thyroid lymphoma. There are no reported cases of primary thyroid lymphoma with direct extension into pyriform fossa. Airway obstruction caused by primary thyroid lymphoma is mainly by means of direct compression of trachea by huge thyroid mass. ¹¹⁻¹⁷ There is no reported case of airway obstruction caused by direct extension of primary thyroid lymphoma into pyriform fossa, occluding laryngeal inlet.

Given the sensitivity of primary thyroid lymphoma to radiation and chemotherapy, these remain the mainstay of treatment. The conventional chemotherapeutic regimen for primary thyroid lymphoma includes cyclophosphamide, doxorubicin, vincristine and prednisone (CHOP). 18-20 Patients with primary thyroid lymphoma typically show good response to this regimen. The role of the surgeons in diagnosis and treatment of thyroid lymphoma has evolved from tumour-debulking to open biopsy. With the availability of radiotherapy and chemotherapy, the need for surgery has nearly disappeared as surgical excision of all disease is not possible or is associated with increased morbidity, except in those patients who present with significant airway compromise.

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