

Case Report

Delayed management of a huge thyroid mass, how to avoid a disaster! A case report

Suzan Al-Zaidi, Mahmoud Al-Reefi* and Saad Al-Muhayawi

Abstract

Demonstrator in the Otolaryngology
Department, King AbdulAziz University,
Rabigh, Saudi Arabia

*Corresponding Author's E-mail:
m_alreefi@yahoo.com
Phone: +955-562220699

In the following report, we discuss a rare case of a huge mass of the thyroid gland that developed in a 45 years old male patient presented with a diffuse neck mass. Imaging shows a large mass of the neck, with no extra thyroid extension. Preoperative diagnosis was made by fine needle aspiration as suspension of TB versus malignancy. Open surgical excision biopsy yielded a definitive diagnosis of thyroid mucosa-associated lymphoid tissue (MALT) lymphoma.

Keywords: Thyroid, Lymphoma, Tuberculosis, airway, obstruction, fine needle, aspiration, chemotherapy.

Case reports

46 years old male presented to our clinic with a midline neck mass which was first noticed by the patient himself around 6 months prior to presentation. It was increasing in size rapidly to involve the whole neck up to both ears causing him difficulties in breathing while lying down in the last 2 months, no stidor, voice change or swallowing problem.

No symptoms of hypothyroidism or hyperthyroidism, radiation exposure or weight loss. No family history of malignancy or thyroid disease. Patient is a smoker on half pack a day. Otherwise, history was unremarkable.

On Examination, the patient was breathing comfortably while in sitting position with no stidor, vitally stable.

Neck shows a huge midline mass extending from level of cricoids cartilage superiorly to 1 cm below the terna notch inferiorly and to posterior border of sternomastoid muscle laterally measuring 17cm in and 5 cm in height, hard in consistency, non tender, non mobile, no signs of inflammation, no palpable cervical lymph nodes and no other masses. Ear, nose and throat examinations were within normal limits.

Fibrooptic zero degree laryngoscopes was performed in the clinic. Pharynx was within normal limits, larynx examination showed sluggish movement of right vocal fold and normal left vocal fold, no masses were

appreciated.

The patient visited multiple head and neck clinics before presenting to us and a fine needle aspiration of the mass was done. It showed "numerous thyroid follicle cells with hurthle cells associated with occasional epithelioid granulomas and multinucleated giant cell in a back ground of inflammatory cells." Suggestive of tuberculosis of the thyroid gland. He was advised to seek medical care in a higher center to consider thyroidectomy since his airway was compromised and to avoid further progression and compression, and was referred to our center. No antituberculous medications were administered.

In our center, more investigations were done including thyroid function test which was normal. CT-scan of the neck with intravenous contrast shows: 17X14 cm hyperintense mass extending superiorly to the level of C1-C2 vertebral region with downward extension to level of superior clavicles (Figure 1). Erosion of inner cortex of thyroid cartilage, pushing great vessels laterally, encasing trachea (Figure 2). No retrosternal extension, patent airway (Figure 3) and multiple subcentimetric cervical lymphnodes.

Work up also included CT-scan of chest, abdomen and pelvis which were within normal limits and showed no distance metastasis or other organ involvement.



Figure 1. Coronal section CT-scan showing extension of the mass



Figure 2. Axial cut CT-scan showing thyroid encasing trachea



Figure 3. Sagittal section CT-scan showing the mass with patent airway



Figure 4. The excised thyroid on the left after sending it for frozen section. The extension lobe is seen on the right.

Due to the presence of shortness of breath, decision was made to take patient to operating room for total thyroidectomy with frozen section and possibility of neck dissection and tracheostomy if needed.

Intraoperatively, a successful easy intubation was made by the anesthesiologist with the guide of Glidoscope using a size 8 mm cuffed endotracheal tubes.

A collar incision was made 2 cm above sternal notch and classic total thyroidectomy was done. Bilateral recurrent laryngeal nerves were identified and preserved. An extension from left lobe was found (measuring around 7X5 cm) and removed successfully. The gland was found to be irregularly enlarged with well formed capsule and a firm nodular surface, it was not attached to surrounding tissues and no extra capsular lymph nodes were found.

Total gland was sent for frozen section Intra operatively and showed "lymphoproliferative lesion favoring the diagnosis of non Hodgkin lymphoma "(Figure 4) so no further neck dissection was done. Hemostasis was achieved and a large drain was inserted, then incision was closed in several layers. Patient was extubated successfully without complication.

Post operatively the patient did very well, no airway problems, calcium levels were within normal limits and started on thyroxin replacement therapy. Histopathological examination of the thyroid gland showed: "mixed population of lymphoid –plasma cell, lymphoid follicles with germinal center." Immunohistochemistry "a panel of markers were performed including CK PAN, LCA, CD20, CD56, Galectin 3 and CK19. Cytokeratin7 and LCA highlighted the lymphoepithelial lesions."

Final diagnosis was mucosa-associated lymphoid tissue lymphoma (MALT Lymphoma) in a background of Hashimoto's thyroiditis. Stage IE on Ann Arbor staging system.

Patient then was referred to oncology for further evaluation, his case was discussed in tumor board, a final decision was made that the tumor was of a low grade, so no need for further intervention and the patient will be followed up closely.

Thyroid lymphoma

Primary thyroid lymphoma is a rare disease that should not be forgotten when diagnosing a neck mass, since its management differs from other thyroid pathologies. In most of thyroid lymphoma, it is of the non-Hodgkin's type (Wang et al., 2005). It present less than 2% of all thyroid malignancies and it constitute 2% of extranodal lymphomas (Freeman et al., 1972). The mean and median ages of diagnosis are between 65-75 years with a female to male ratio of 8:1 (Logue et al., 1992).

The only known risk factor of developing thyroid lymphoma is the presence of chronic thyroiditis, precisely

Hashimoto's thyroiditis (Pedersen and Pedersen, 1996). Primary thyroid lymphoma is almost always of B-cell lineage. 60 to 80 percent of thyroid lymphomas are large B-cell lymphomas (Wolf et al., 1992). The next most common subtype (approximately 30 percent) is extranodal marginal zone lymphoma. The extranodal marginal zone lymphomas of mucosa-associated lymphoid tissue (MALT) type are generally associated with Hashimoto's thyroiditis.

The most characteristic presentation is that of a rapidly enlarging neck mass often associated with dysphagia (Singcr JA, 1988). The majority of patients are euthyroid and one third of patients have compressive symptoms. However distant metastasis is rare. The mass is usually fixed to surrounding tissues and half the patients have unilateral or bilateral cervical lymph node enlargement (Singcr JA, 1988).

As noted above, some patients may have hypothyroidism, indicative of Hashimoto's thyroiditis, but there is no laboratory abnormalities that are specific to thyroid lymphomas (Pedersen and Pedersen, 1996). Ultrasound of the thyroid gland and a needle biopsy is a good initial diagnostic test, however, differentiating thyroid lymphoma from Hashimoto's thyroiditis by thyroid cytology may be difficult. This difficulty can lead to open surgical biopsy to make the diagnosis (as the case presented). Open biopsy or core needle biopsy is adequate for a definitive diagnosis in all cases (Andeh et al., 1997). Given the frequent coexistence of Hashimoto's thyroiditis, small cell lymphomas are more difficult to diagnose cytologically, and immunohistochemical staining or flow cytometry may be necessary to establish monoclonality and characterize surface markers. CT scans and M.R.I are not helpful establishing the diagnosis of thyroid lymphoma. However, CT or M.R.I of neck, thorax, abdomen and pelvis are required for proper staging (Schlumberger and Caillon, 1996). For staging, the Ann Arbor staging classification is used (Table 1) (Carbone et al., 1971).

Surgery role in treating thyroid lymphoma has always been debatable; it ranges from open biopsy to debulking of the tumor in the presence of airway compression (Pasiaka, 2000). Effective treatment of thyroid lymphoma depends mainly on the extent of the disease and histological type. Patients with diffuse large B-cell lymphoma (DLBCL) of the thyroid should be treated in the same manner as employed in patients with DLBCL of any other site or extent. Thus, patients with localized, early stage disease can be treated with either three courses of combination therapy (e.g. CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) plus rituximab), followed by radiation therapy to the thyroid bed or six to eight cycles of CHOP plus rituximab without radiation (Matsuzuka et al., 1993). Patients with localized extranodal marginal zone lymphoma of the thyroid or other indolent histologies (eg, follicular lymphoma, small cell lymphoma) can be effectively

Table 1. Ann Arbor classification for staging Hodgkin's and non-Hodgkin's lymphomas.

Stage I	Involvement of a single lymph node region (I) or of a single extralymphatic organ or site (IE)*
Stage II	Involvement of two or more lymph node regions or lymphatic structures on the same side of the diaphragm alone (II) or with involvement of limited, contiguous extralymphatic organ or tissue (IIE)
Stage III	Involvement of lymph node regions on both sides of the diaphragm (III), which may include the spleen (IIIS) or limited, contiguous extralymphatic organ or site (IIIE) or both (IIIES)
Stage IV	Diffuse or disseminated foci of involvement of one or more extralymphatic organs or tissues, with or without associated lymphatic involvement

treated with radiation therapy alone. Those with advanced stage indolent histologies are usually treated with chemotherapy alone (Sun et al., 2013).

Teaching points

Although rare, thyroid lymphoma is pathology of the thyroid gland that should not be underestimated or forgotten. The proper diagnostic approach would be core biopsy or, in case of air way disturbance, complete excision of the mass. Fine needle biopsy will not be helpful in this condition and it can cause misdiagnosis or delay of reaching the proper diagnosis.

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