

# Delayed management of a huge thyroid mass: how to avoid a disaster

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## Abstract

A rare case of a huge mass in the thyroid gland that developed in a 45-year-old man who presented with a diffuse neck mass is discussed. Imaging showed a large mass in the neck, with no extra thyroid extension. A preoperative diagnosis was made by fine-needle aspiration as suspicious for tuberculosis or malignancy. Open surgical excision biopsy yielded a definitive diagnosis of thyroid mucosa-associated lymphoid tissue (MALT) lymphoma.

## Keywords

Neck; thyroid; mass; tuberculosis; lymphoma; MALT; fine-needle aspiration; core biopsy.

## Case report

A 46-year-old male presented to our clinic with a midline neck mass, which was first noticed by the patient about 6 months before presentation. It was increasing in size rapidly to involve the neck up to both ears causing him difficulties in breathing while lying down over the last 2 months; there was no stridor, voice changes or swallowing problems. No symptoms of hypothyroidism or hyperthyroidism, radiation exposure or weight loss were present and there was no family history of malignancy or thyroid disease. The patient was a smoker on half pack a day. Otherwise, his history was unremarkable.

On examination, the patient was breathing comfortably while in the sitting position with no stridor; he was vitally stable. A huge midline mass was visible in his neck extending from the level of the cricoids cartilage superiorly to 1 cm below the sternal notch inferiorly and to the posterior border of the sternomastoid muscle laterally. The mass was 17 cm wide and 5 cm high, of hard consistency, non-tender, non-mobile, with no signs of inflammation, no palpable cervical lymph nodes and no other masses. Ear, nose and throat examinations were within normal limits.

Fiber optic zero degree laryngoscopy was performed in the clinic. The pharynx was examined and found to be within normal limits; examination of the larynx showed sluggish movement of right vocal fold and normal left vocal fold; no masses were found.

The patient visited several head and neck clinics before presenting to us. A fine-needle aspiration of the mass showed numerous thyroid follicle cells with Hurthle cells associated with

occasional epithelioid granulomas and multinucleated giant cells in a background of inflammatory cells. These findings were suggestive of tuberculosis of the thyroid gland. He was advised to seek medical care in a higher center to consider thyroidectomy because 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 a thyroid function test, which was normal. A computed tomography (CT) scan of the neck with intravenous contrast showed a  $17 \times 14$  cm hyperintense mass extending superiorly to the level of the C1-C2 vertebral region with downward extension to the level of the superior clavicles (Fig. 1). Erosion of the inner cortex of the thyroid cartilage was present, the mass was pushing the great vessels laterally, encasing the trachea (Fig. 2). There was no retrosternal extension, the airway was patent (Fig. 3) and multiple subcentimeter cervical lymph nodes were found. Work up also included a CT scan of the chest, abdomen and pelvis which were within normal limits and showed no distant metastasis or other organ involvement.

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

Intraoperatively, successful easy intubation was performed by the anesthesiologist guided by a GlideScope using an 8-mm cuffed endotracheal tube. A collar incision was made 2 cm above the sternal notch and a classic total thyroidectomy was carried out. Bilateral recurrent laryngeal nerves were identified and preserved. An extension from the left lobe was found (measuring approx.  $7 \times 5$  cm) and removed successfully. The gland was found to be irregularly enlarged with a well-formed capsule and a firm nodular surface; it was not attached to surrounding tissues and no extracapsular lymph nodes were found.

The whole gland was sent for frozen section intraoperatively and showed a lymphoproliferative lesion favoring the diagnosis of non-Hodgkin lymphoma (Fig. 4) so no further neck dissection was done. Hemostasis was achieved and a large drain was inserted before the incision was closed in several layers. The patient was extubated successfully without complications. Postoperatively, the patient did very well; he had no airway problems, his calcium levels were within normal limits and he was started on thyroxin replacement therapy.

Histopathologic examination of the thyroid gland showed a mixed population of lymphoid and plasma cells, lymphoid follicles with germinal center. A panel of immunohistochemistry markers was performed including pan-cytokeratin, leukocyte common antigen (LCA), CD20, CD56, Galectin 3 and CK19. Cytokeratin7 and LCA highlighted the lymphoepithelial lesions.

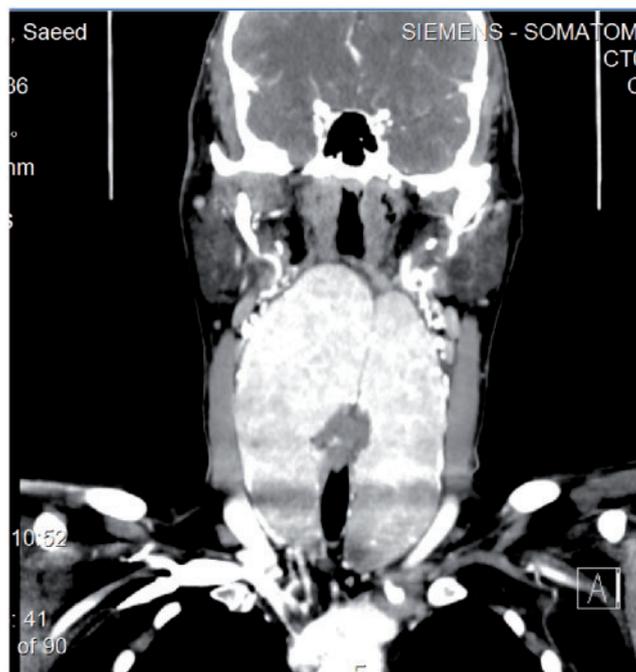


Fig. 1. Coronal section CT scan showing the extension of the mass.



Fig. 2. Axial cut CT scan showing thyroid encasing the trachea.

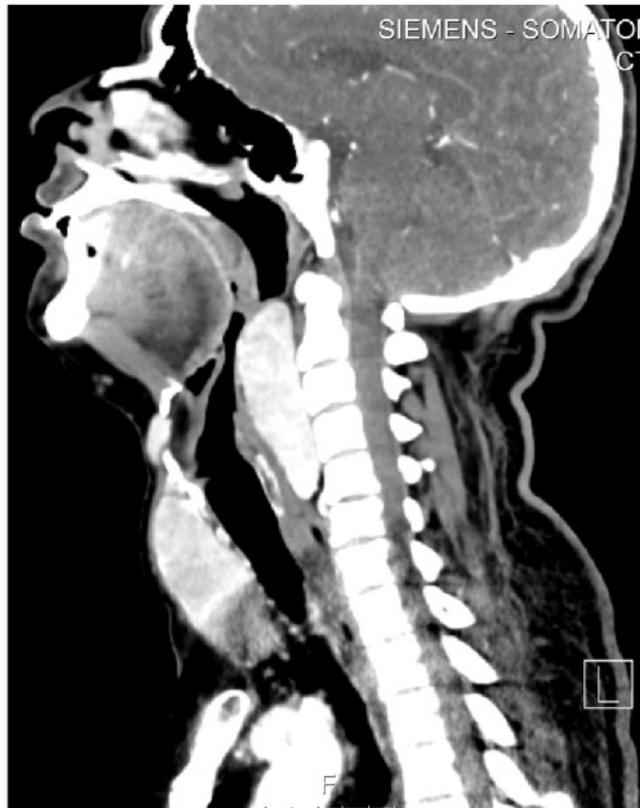


Fig. 3. Sagittal section CT scan showing the mass.

The final diagnosis was mucosa-associated lymphoid tissue lymphoma (MALT lymphoma) on a background of Hashimoto thyroiditis (stage IE on the Ann Arbor staging system).

The patient was referred to oncology for further evaluation; his case was discussed by the tumor board, and the final decision was that the tumor was low grade and there was no need for further intervention other than close follow-up.

## Discussion

Primary thyroid lymphoma is a rare disease that should not be forgotten when diagnosing a neck mass, because its management differs from other thyroid pathologies. Thyroid lymphomas are usually non-Hodgkin type<sup>[1]</sup>. Occurrence is less than 2% of all thyroid malignancies and 2% of



Fig. 4. The excised thyroid and the extension.

**Table 1.** The Ann Arbor classification for staging lymphoma

Stage I	Involvement of a single lymph node region (I) or of a single extralymphatic organ or site (IE) <sup>a</sup>
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

All cases are subclassified to indicate the absence (A) or presence (B) of the systemic (B) symptoms of significant unexplained fever, night sweats, or unexplained weight loss exceeding 10% of body weight during the 6 months prior to diagnosis. Clinical stage refers to the extent of disease determined by diagnostic tests following a single diagnostic biopsy. If a second biopsy of any kind is obtained, even if negative, the term pathologic stage is used.

<sup>a</sup>The designation E generally refers to extranodal contiguous extension (i.e., proximal or contiguous extranodal disease) that can be encompassed within an irradiation field appropriate for nodal disease of the same anatomic extent. A single extralymphatic site as the only site of disease should be classified as IE, rather than stage IV.

extranodal lymphomas<sup>[2]</sup>. The mean and median age at diagnosis is between 65 and 75 years with a female to male ratio of 8:1<sup>[3]</sup>. The only known risk factor for developing thyroid lymphoma is the presence of chronic thyroiditis such as Hashimoto thyroiditis<sup>[4]</sup>. Primary thyroid lymphoma is almost always of B-cell lineage and 60–80% of thyroid lymphomas are large B-cell lymphomas<sup>[5]</sup>. The next most common subtype (approx. 30%) is extranodal marginal zone lymphoma. The extranodal marginal zone lymphomas of MALT type are generally associated with Hashimoto thyroiditis.

The most characteristic presentation is that of a rapidly enlarging neck mass often associated with dysphagia<sup>[6]</sup>. Most 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<sup>[6]</sup>.

As noted above, some patients may have hypothyroidism, indicative of Hashimoto thyroiditis, but there are no laboratory abnormalities that are specific to thyroid lymphomas<sup>[4]</sup>. Ultrasonography of the thyroid gland and a needle biopsy are a good initial diagnostic tests, however, differentiating thyroid lymphoma from Hashimoto thyroiditis by thyroid cytology may

be difficult. This difficulty can lead to open surgical biopsy to make the diagnosis (as in this case). Open biopsy or core-needle biopsy is adequate for a definitive diagnosis in all cases<sup>[7]</sup>.

Given the frequent coexistence of Hashimoto 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 magnetic resonance imaging (MRI) are not helpful for establishing the diagnosis of thyroid lymphoma. However, CT or MRI of the neck, thorax, abdomen and pelvis are required for proper staging<sup>[8]</sup>. The Ann Arbor staging classification is used (Table 1)<sup>[9]</sup>.

The role of surgery in treating thyroid lymphoma has always been debatable; surgery ranges from open biopsy to debulking of the tumor in the presence of airway compression<sup>[10]</sup>. 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 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<sup>[11]</sup>. Patients with localized extranodal marginal zone lymphoma of the thyroid or other indolent histologies (e.g., follicular lymphoma, small cell lymphoma) can be treated effectively with radiation therapy alone. Those with advanced stage indolent histologies are usually treated with chemotherapy alone<sup>[12]</sup>.

## Teaching points

Although rare, thyroid lymphoma is a pathology of the thyroid gland that should not be underestimated or forgotten. The proper diagnostic approach is core biopsy or, in the case of airway disturbance, complete excision of the mass for a better histopathologic evaluation. Fine-needle biopsy is not helpful for this condition because the tissue obtained is inadequate and it can cause misdiagnosis or delay in reaching the proper diagnosis.

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