

Case Report

Primary thyroid lymphoma: A rare malignancy in the head and neck

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Abstract Primary thyroid lymphoma is a relatively uncommon pathology of the thyroid gland that mainly occurs in elderly females. We describe a rare case of B-cell thyroid lymphoma in a young healthy male. It is an important diagnosis to be considered in patients presenting with a rapidly enlarging neck mass as its management is different from other differentiated thyroid carcinoma which require total thyroidectomy plus adjuvant radioactive iodine ablation. Our report emphasizes the need for clinical awareness leading to early detection, followed by early multidisciplinary management.

Keywords: Chemotherapy; lymphoma; neck mass; radiotherapy; thyroid gland.

Introduction

Primary thyroid lymphoma (PTL) is diagnosed whenever the pathology of lymphoma involves only the thyroid gland and lymph nodes of the neck, without contiguous spread or distant metastases from other areas of involvement at diagnosis (Hedhili *et al.*, 2015). It is an uncommon thyroid pathology and constitutes only 1%-5% of thyroid malignancies and 1%-2% of extranodal lymphomas (Alzouebi *et al.*, 2012; Agarwaf *et al.*, 2013). It is important to diagnose and to differentiate it with other thyroid malignancies such as papillary or follicular thyroid carcinoma which require surgery and adjuvant radioactive iodine treatment. Most PTL are B-cell non-Hodgkin lymphoma and usually treated by chemoradiotherapy (Soni *et al.*, 2017).

Case report

A 48-year-old male with no known medical illness presented with anterior neck swelling, which progressively increased in

size in the last four months. It was associated with hoarseness for one-week duration. He denied any noisy breathing at that time. There was no history of dysphagia, shortness of breath or nasal symptoms. He had no fever, night sweats or weight loss. The patient was an ex-smoker and had no previous neck radiation exposure. There was no family history of malignancy. Clinically there was a diffuse non-tender anterior neck mass measuring 8 cm x 8 cm which extended laterally until medial border of sternocleidomastoid muscle. There was no palpable lymphadenopathy (Fig. 1). Laryngoscopy revealed right vocal cord paresis. Thyroid function test and other routine blood parameters were normal. The patient underwent a contrast-enhanced computer tomography (CT) of the neck and it showed grossly enlarged (6.2 cm x 7.2 cm x 9.7cm) mass in the right thyroid gland with retrotracheal extension, pushing the trachea to the left (Fig. 2). There were no cervical lymph nodes seen. CT thorax and abdomen showed no distant metastasis.



Fig. 1 Diffuse anterior neck mass (post incisional biopsy).

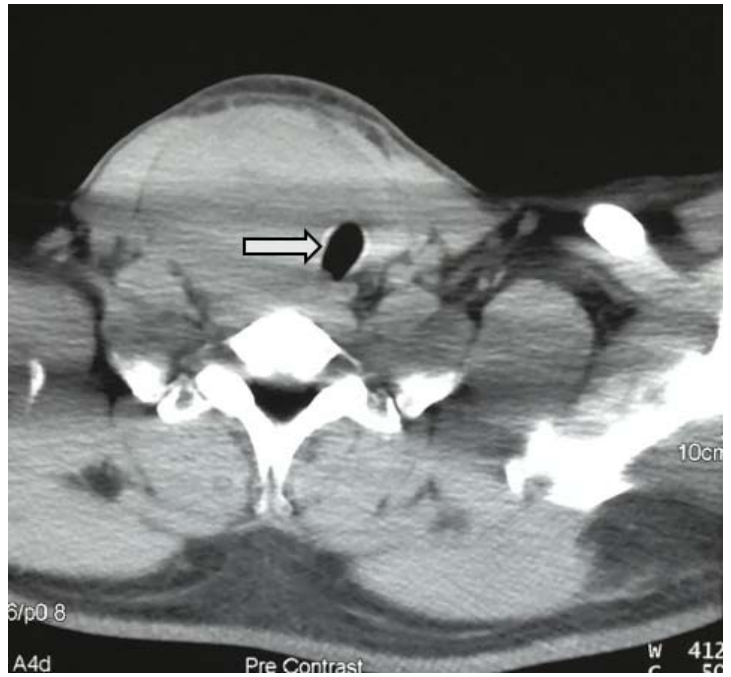


Fig. 2 CT showing a large heterogeneous mass of the right thyroid gland pushing the trachea to the left (arrow).

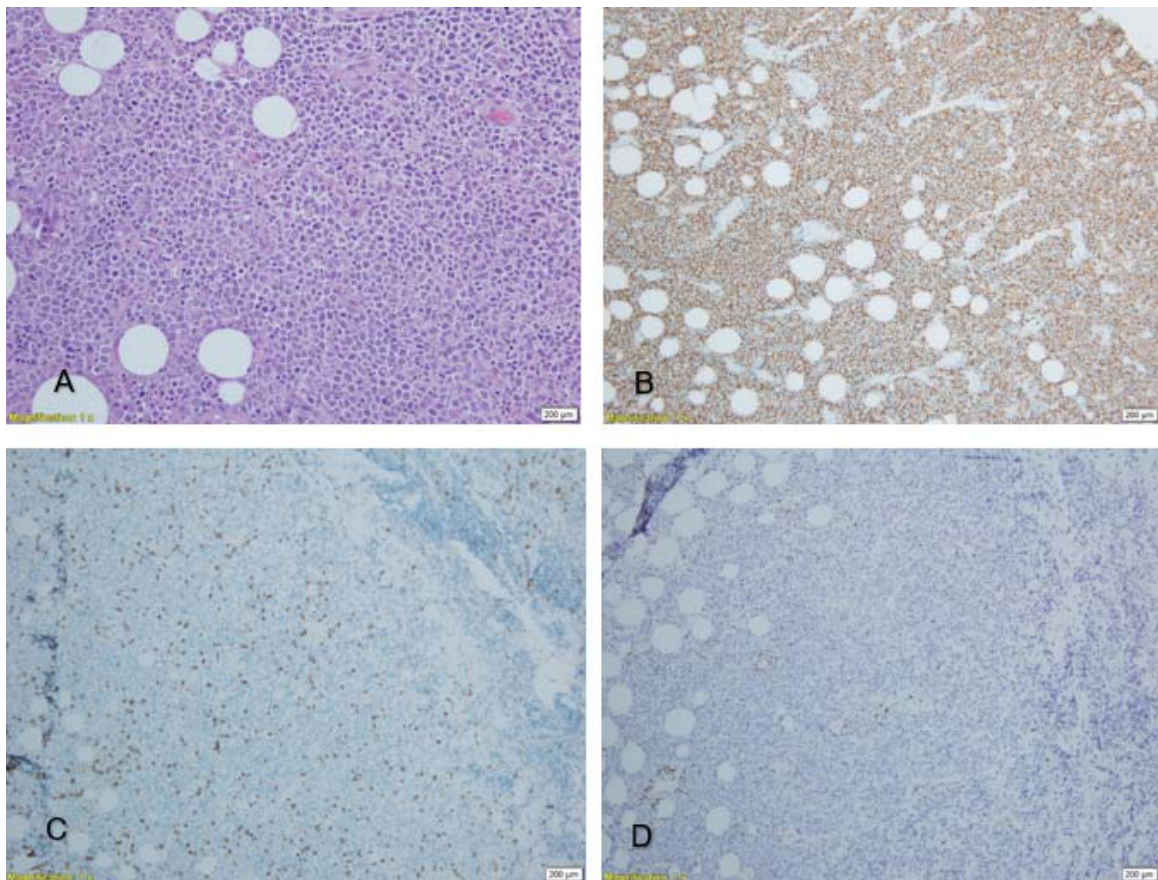


Fig. 3 Histological and immunohistochemical photographs of the thyroid mass. (A) Microscopic Examination (H&E x200 magnification); (B) CD20 and CD79a are positive (x 100 magnification); (C) CD3 is negative (x 100 magnification); (D) BCL2 is negative (x 100 magnification).

Fine needle aspiration cytology (FNAC) of the mass was obtained which showed suspicion of lymphoproliferative disease thus incisional biopsy was done for further tissue diagnosis. Microscopically, there was diffuse sheets of malignant lymphoid cells composed of medium to large-sized cells. These cells exhibited pleomorphic, fine chromatin clumps and are admixtures of centroblasts and occasional immunoblasts. Immunohistochemical analysis was reported as positive for CD20, CD79a and MUM1 with high proliferative index (ki67=70%). They are negative for CD3, CD10 and BCL2 (Fig. 3).

With histological and immunohistological analyses, the diagnosis of thyroid Diffuse Large B-cell lymphoma was reached. The patient was urgently referred to the haematology and oncology teams for further management. At this time of presentation, patient clinically showed no evidence of airway obstruction. One week later, just prior to his haematology and oncology appointment, unfortunately, the patient passed away, most likely due to airway compromise.

Discussion

PTL is a rare entity, making less than 5% of thyroid malignancies. From all lymphomas, it accounts not more than 2.5% (Hedhili *et al.*, 2015). Most PTL are B cell non-Hodgkin lymphoma (Chen *et al.*, 2014). About 50-80% of the PTL are diffuse large B cell lymphoma (DLBCL) and extra nodal mucosa associated lymphoid tissue (MALT) lymphomas constitute the remaining 20-30%. Other rare subtypes include follicular lymphoma (12%) and Hodgkin disease (7%) (Chen *et al.*, 2014). Contradictory to the present case, PTL commonly occurs in the 60-70 years old, and frequently in females with the ratio of 3:1 (Hedhili *et al.*, 2015).

The most common clinical presentation is a rapidly growing, painless mass in thyroid region, either in the form of goitre or discrete nodule. Compression by the mass over adjacent structures will produce symptoms of hoarseness, dysphagia and stridor in 30% of patients (Katna *et al.*, 2013). About 20% of patient will have fever, sweating and weight loss

(Katna *et al.*, 2013). These clinical presentations mimic anaplastic thyroid carcinoma (ATC). Our patient presented with a rapidly expanding neck swelling, so it is important to diagnose it promptly as PTL which is highly curable without the need of thyroidectomy.

Although ultrasonography (USG) is the usual modality of choice for imaging of the thyroid lesion, it is less effective to evaluate the larger thyroid masses as in the present case. To evaluate the mass extension, assessment of local invasion, lymph node metastasis, evaluation of oro-naso-laryngopharynx and oesophagus; CT is much preferred (King, 2008). The positron emission tomography (PET) scan is useful for detection of distant metastasis or staging of the disease, radiotherapy treatment planning as well as treatment response evaluation (Soni *et al.*, 2017). In our patient, the lesion only localised at thyroid gland without nodal and distant metastasis, staged as T3N0M0. When FNAC is unsatisfactory or not diagnostic, a tissue diagnosis is achieved through core biopsy, incisional biopsy or even a thyroidectomy (Sarinah and Hisham, 2010).

In view of the similar presentation, it is very important to differentiate between ATC and PTL as the latter has excellent response with chemoradiotherapy (Soni *et al.*, 2017). While ATC is the most feared thyroid malignancies with mean survival of six months, PTL has excellent prognosis with early treatment (Kitamura *et al.*, 1999). The five-year life expectancy after the treatment of PTL is 75% (Graff-Baker *et al.*, 2009). Owing to its high curability, early detection and referral to haematology team will benefit the patient.

Chemotherapy, such as cyclophosphamide followed by radiotherapy is the mainstay of the treatment for PTL, not thyroidectomy (Katna *et al.*, 2013). With the relapse rate of 7.7% following chemoradiotherapy compared to 37.1% and 43% after radiotherapy and chemotherapy alone, respectively, combination of chemoradiotherapy has shown superior results (Doria *et al.*, 1994).

In the present case, although patient did not have any comorbidity, but late presentation with aggressive nature of

tumour may contribute to the mortality of patient. In the event of upper airway obstruction, the airway is best secured via orotracheal intubation. Opening the neck for tracheostomy may not be a good choice until the tissue diagnosis is obtained in order to avoid tumour seeding. Besides, it is technically very difficult as the trachea is encased and pushed by the enlarged mass. Unless the neck is opened for tissue biopsy, hemithyroidectomy can be performed as part of impending airway obstruction management until definitive plan is given (Lawal *et al.*, 2017). Addition of steroid can be instituted to reduce airway oedema as steroid is part of non-Hodgkin lymphoma treatment as well (King, 2008).

Conclusion

Lymphoma of thyroid is uncommon, especially in a healthy young male patient as in our case. Tissue biopsy is required for the diagnosis and to differentiate with other thyroid malignancy particularly ATC. Thyroidectomy is not indicated in PTL as chemoradiotherapy has excellent treatment outcomes.

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