

Primary Thyroid Lymphoma: A Report of Two Cases

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We present two cases of primary thyroid lymphoma suggesting the possibility of anaplastic carcinoma. Fine needle aspiration (FNA) revealed lymphocytes that are interpreted as a thyroiditis. Open surgical biopsy yielded a definitive diagnosis as a thyroid lymphoma in both cases.

Key words: Thyroid, lymphoma, chemotherapy

Introduction

Primary lymphomas of the thyroid are uncommon tumours, representing approximately %5 of the thyroid neoplasms and %2 of extranodal lymphomas (1,2,3). The peak incidence is in the 5th to 7th decades and the male/female ratio is 1/3 (2,4). The most characteristic presentation is that of a rapidly enlarging neck mass often associated with dysphagia (5). 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 (2,5,6). Most thyroid lymphomas are nonhodgkin lymphomas and its differentiation from small cell anaplastic carcinoma of thyroid gland may be difficult with FNA (2,7). Lymphoma is eminently treatable, whereas anaplastic carcinoma has a very poor prognosis. Incidence of lymphocytic thyroiditis (Hashimoto's thyroiditis) in patients

diagnosed with primary lymphoma of the thyroid is 25 to 100 percent range (5,8,9,10). However, the actual relationship between Hashimoto's thyroiditis and thyroid lymphomas remains obscure (2).

There is no non invasive diagnostic test. Chest radiographs may demonstrate deviation of the trachea, ultrasound shows characteristic asymmetrical pseudocystic pattern, CT scans and magnetic resonance imaging may confirm the presence of an enlarged thyroid mass (1,4,11,12,13). Gallium 67 scanning may have a role in diagnosing nonhodgkin lymphomas (8,14). FNA can help to distinguish lymphoid proliferation from epithelial tumours.

However, differentiating thyroid lymphoma from Hashimoto's thyroiditis by thyroid cytology may be difficult (2,15,16). This difficulty can lead to open surgical biopsy to make the diagnosis. Open biopsy or core needle biopsy is adequate for a definitive diagnosis in all cases (1). Accurate diagnosis and staging are important for planning treatment.

Staging investigations include physical examination, blood count, serum lactate dehydrogenase and B2-microglobulin measurements, liver function tests, bone marrow biopsy, CT or MRI of neck, thorax, abdomen and pelvis and appropriate biopsies (2).

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CASE REPORT

By the Ann Arbor Classification; Stage IE (E indicates extranodal disease) involves localised disease with in the thyroid, Stage IIE is disease confined to the thyroid and regional lymph nodes. Stage IIIE involves disease on both sides of the diaphragm. Stage IVE is disseminated disease (2). Proper place for the various treatment modalities are still controversial. Despite the fact that nonhodghin's thyroid lymphoma responds to both chemotherapy and radiotherapy, there are still proponents of radical surgical resection. Therefore on the basis of a review of the literature, the role of surgery is limited to obtaining tissue for diagnosis and to limited resection when the airway is compromised (2,4,5, 17,18).

In the present paper, we reported two thyroid lymphomas which were diagnosed by surgical biopsy and successfully treated with chemotherapy.

Case 1

36 year old 12 weeks pregnant female presented with four weeks history of respiratory problems, difficulty in speaking, cough and swelling on the left side of her neck. Physical examination was normal apart from a hard mass on left side of her thyroid approximately 4 cm in diameter and decreased pulmonary sounds on lower lobe of left lung. Ultrasound showed asymmetrical diffuse enlargement of left thyroid lobe lasting left thoracal area and a cervical lymph node. Chest x-ray revealed pleural effusion on left lung. CT scan showed a mass on the left side of the neck (Figure 1) associated with a thoracal mass (Figure 2). Biochemical profile and thyroid function tests were normal. FNA showed lymphoid cell infiltration, pleural tap revealed no atypical cells in pleural fluid. Diagnosis of non-Hodgkin's lymphoma was accomplished with open surgical biopsy. Her pregnancy was terminated.

CT scans of thorax abdomen and pelvis were normal except left pleural effusion. Bone marrow aspiration and biopsy were normal. With these findings patient was staged as IV and commenced on chemotherapy. She received 6 cycles of etoposide, adriamycin, cyclophosphamide and prednisolon chemotherapy which produced a complete regression.

Figure 3a., 3b., 4a., 4b. Show regression of tumour after first and second cycles of chemotherapy. She is alive and well 3 months after completing chemotherapy.

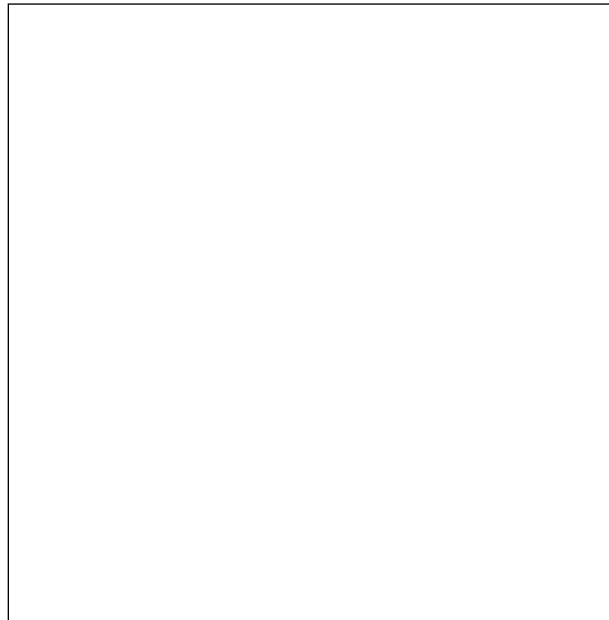


Figure 1. Case 1. Cervical mass in CT scan.

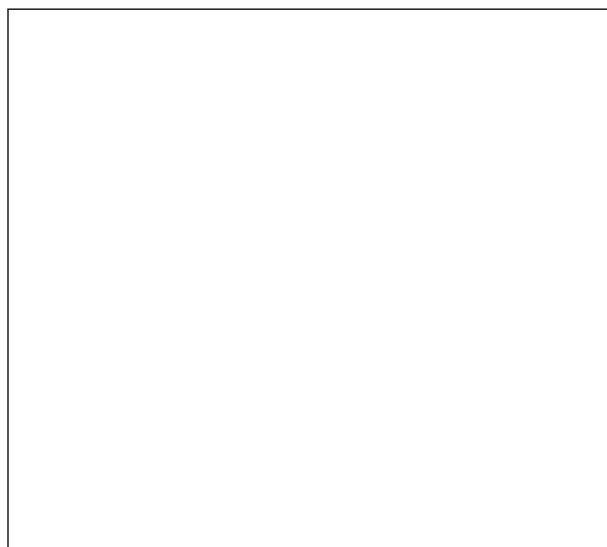


Figure 2. Case 1. Thoracal mass in CT scan.



Figure 3a. Case 1. One month after chemotherapy.

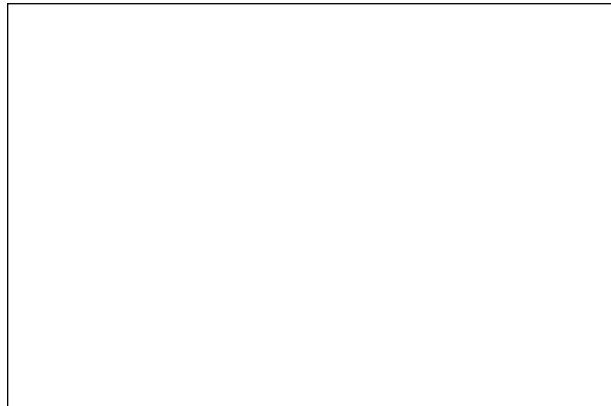


Figure 3b. Case 1. Two months after chemotherapy..

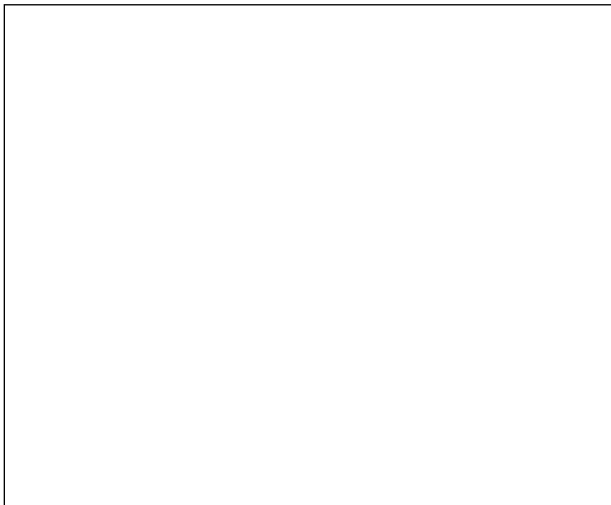


Figure 4a. Case 1. One month after chemotherapy.

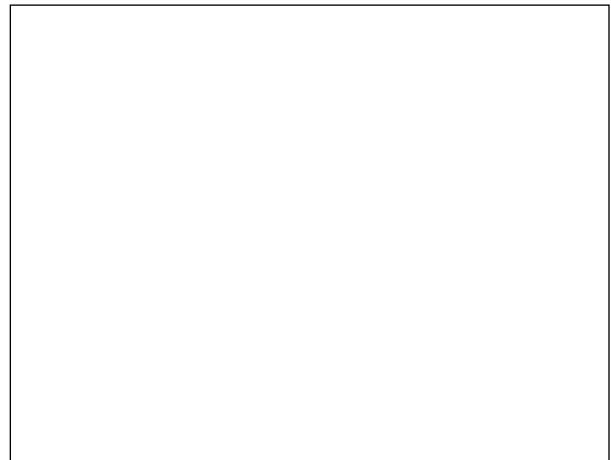


Figure 4b. Case 1. Two months after chemotherapy.

Case 2

70 year old male presented with 3 months history of swelling on the left side of his neck. Physical examination was normal except a hard mass occupying left lobe of his thyroid. Biochemical profile and thyroid function tests were normal. Ultrasound showed a diffuse enlargement of the left lobe of thyroid, encompassing carotid artery (Figure 5). MRI revealed a mass containing necrotic components extending into the mediastinum and compressing on trachea (Figure 6). FNA revealed suspected thyroiditis. Low grade non Hodgkin's lymphoma -diffuse centrocytic centroblastic lymphoma (Kiel)- with open surgical biopsy.

Staging procedures showed no disseminated disease. Treatment consisted of 6 cycles of CHOP (adriamycin, cyclophosphamide, vincristine, prednisolone) chemotherapy.

His treatment is still continuing. A complete response was obtained after the second cycle of chemotherapy.

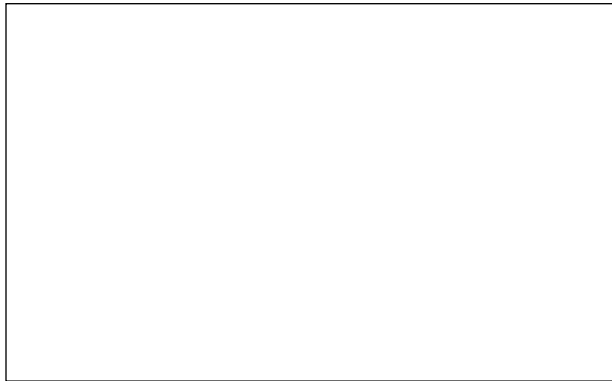


Figure 5. Case 2. Ultrasonographic appearance of the mass.

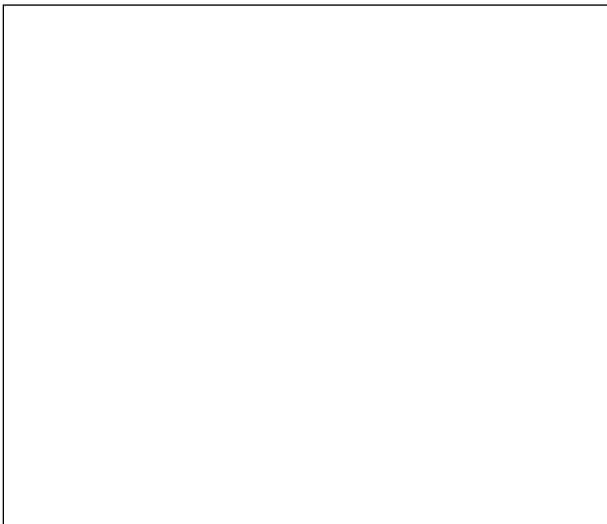


Figure 6. Case 2. MRI showing compression on trachea.

Discussion

It may be difficult to distinguish thyroid lymphoma from anaplastic carcinoma and thyroiditis. On the other hand the treatment for patients with primary thyroid lymphoma differs from other primary malignant tumours of the thyroid gland (5,16). The majority of thyroid lymphomas are diffuse, large cell tumours of B cell origin with a minority of the small cell variety. Present studies recommend an ultrasound scan of the thyroid gland and fine needle aspiration biopsy as initial diagnostic procedures when ever thyroid lymphoma is suspected (3). The overall sensitivity of FNA is % 60 (19). FNA was not diagnostic in neither of our patients.

According to clinical and imaging studies, we suspected possibility of thyroid lymphoma in both cases and then open surgical biopsy were done to

make diagnosis and for specific typing of lymphomas.

Several authors discuss both clinical and pathologic indicators of prognosis. Importance of histological type is controversial (5). Several authors found no correlation between histological type and prognosis (5). On the other hand, some others stated that nodular lymphomas have a better prognosis than diffuse tumours (5,20,21).

Most authors agree that extension outside of the gland, bulky tumours and lymph node metastases decrease the survival (5). Reported 5 year survival rates for primary lymphoma of the thyroid ranging 40 to 60 percent, with the percentage survival increasing to 70 to 80 percent for stage I disease. Incontrast patients for stage III and IV disease the survival rate drops to 30 to 40 percent (5). There is no agreement about the role of the surgeon in the treatment of thyroid lymphomas. Surgery has evolved from surgical debulking to open biopsy (18). Initially, surgery was used extensively for the treatment of this disease. More recently, surgical removal has been shown to have a limited benefit. Aggressive surgery to debulk large thyroid lymphomas, is neither feasible nor necessary. The risks of aggressive thyroid surgery include damage to the recurrent laryngeal nerves, the trachea, the esophagus, the large vessels, and the parathyroid glands (15). It's well known that there is no statistically significant differences in survival rates among patients who underwent surgery followed radiation therapy and combinations of radiation and chemotherapy without extensive surgery (19).

Others have suggested that the addition of surgical debulking is necessary because the amount of residual disease in the neck correlated to the relapse. In spite of this surgery followed by local radiation therapy is indicated only if the malignancy is completely intrathyroidal (20). Thyroid lymphomas have been shown to be both chemosensitive and radiosensitive (21). If it is necessary external radiotherapy should be given in a total dose of 40 Gy (4000 rads) to the neck and mediastinum. About one third of the patients will have systemic relapse when treated with local radiation alone (22). Use of combination chemotherapy may decrease the change of distant relapse. The chemotherapy

should be an anthracycline based regimen. it usually consists of three to six cycles of the CHOP regimen (doxorubicin 50 mg/m², cyclophosphamide 750 mg/m², vincristine 1.4 mg/m², prednisone 40 mg/m²) (2,5,23). Some authors reported that the long term survival rate of combined chemo and radiotherapy was nearly % 100 (6). This treatment may not be suitable for patients in poor general health conditions. We used surgery just for diagnosis in both cases, and as a treatment we prefer to use combination chemotherapy.

in conclusion, lymphoma of the thyroid is a relatively unusual tumour. it should be remembered if there is a mass in neck which is enlarging rapidly, ultrasound guided FNA is an appropriate initial step when there is a high index of suspicion of nonhodgkins lymphoma. But surgeons will often need to perform an open biopsy. Definitive treatment of localised nonhodgkin lymphoma should be consists of chemotherapy with or without radiotherapy. There have been no prospective randomised studies evaluating the role of surgery in this tumour. Although resection of lymphoma is not suggested.

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