中文題目：不典型的雙側腎上腺及甲狀腺淋巴瘤：個案報告

英文題目：Bilateral adrenal glands plus thyroid lymphoma with atypical presentation

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Introduction :

Bilateral adrenal glands plus thyroid lymphoma is a rare condition. To our knowledge, most cases' pathology were diffuse large B cell lymphoma. Here we presented a case of bilateral adrenal glands and thyroid Maltoma.

Case Presentation :

This 69 y/o, Taiwanese female had experienced intermittent low grade fever to 38*C since 2 weeks ago, accompanied with cough, exertional dyspnea, general weakness, night sweating, abdomen distension, poor appetite, decreased body weight. She had hypertension before under life style control, but her family found that her blood pressure was unstable at home (high and low intermittently), sometimes accompanied with vertigo. Because of sudden onset vertigo, she was sent to Emergency room for help.

At ER, Her vital signs were T: 38.8*C, P: 103/min, RR: 25/min, SPO2: 99% under room air, BP: 82/40. PE found conscious clear, slight tachypnea, stridor, irregular heart bear, soft abdomen, no legs edema. Lab data documented WBC: 9900/ul, Hb:13.1g/dl, platelet: 191000/ul, no band form cell, crp: 6.2 mg/dl, creatinine: 1.31 mg/dl, BUN: 19 mg/dl, Na:132 meq/L, K: 4.7 meq/L, normal liver function, random glucose:114 mg/dl, NTpro-BNP: 1937 pg/ml, troponin I < 0.01 ng/ml, TSH:1.324 uIU/ml, free T4:1.330 ng/dl. Blood gas analysis showed PH: 7.537, PCO2:30mmHg, PO2:213.8mmHg, HCO3:25.7meq/L. EKG documented atrial fibrillation with rapid ventricular response. CXR revealed no abnormality over both lung zones and heart figure is not enlarged.

Whole body CT were then arranged to find infection source and other cause of cough which revealed 1. Left thyroid tumor mass, 7.1 cm in diameter with focal central necrosis 2.Bilateral adrenal mass lesion, 13.8 cm on the right and 8.1 cm on the left, both with some central necrosis (The inhomogeneous part with central hypodensity has Hounsfield unit around 30 and other part around 60 to 80) 3.small lymph nodes at mesentaric root and aortocaval region. Based on the finding of left thyroid and bilateral adrenal mass, she was admitted.

On 2021/06/22, echo guide biopsy of left thyroid mass was done. Further lab data for bilateral adrenal mass showed normal urine 24 Hr-VMA, nomal early morning cortisol, high ACTH: 159 pg/ml (normal range 7.9-47.1 pg/ml), aldosterone: 3.19 ng/dl ↓, renin:

3.15 ng/ml/hr, low testosterone: 0.08 ng/ml. Her urine norepinephrine, epinephrine and dopamine were all normal. Thyroid mass pathology finally showed mucosa associated lymphoid tissue lymphoma (low grade small B cell lymphoma). PET/CT revealed thyroid and adrenal B-cell lymphoma with both-sided diaphragmatic nodal involvement (right parotid, bilateral neck, superior diaphragmatic, gastric, hepatic, precaval, aortocaval, and mesenteric lymph nodes) and suggestive of bone marrow involvement. Bone marrow biopsy was then performed which showed no bone marrow involvement.

The patient was diagnosed with MALToma, Ann Arbor stage IV. She received pre-phase Dexamethasone (2021/6/23-6/26), and received chemotherapy with R-CHOP (Cyclophosphamide, Doxorubicin, Oncovin, Prednisolone), C1 on 2021/6/30. We planned to arrange 6-8 cycle of therapy. The follow up PET/CT found both thyroid and adrenal mass significant regression and resolution. Now the patient was regularly followed up at OPD under stable condition.

Discussion :

Non-Hodgkin’s lymphomas (NHL) mostly arise in lymph nodes or other lymphatic tissues such as the thymus and spleen. After staging examination, involvement of extranodal organs is a common finding. Sometimes, a substantial part of NHL even arises in these sites, also named as primary extranodal NHL. Thyroid, GI tract, intra-abdominal solid organ, pleural, bone and CNS involvement have been reported.

The most common clinical presentation of thyroid lymphoma is a rapidly growing neck mass, as in our case. Thyroid dysfunction can be seen in 7-59% of patients due to tumor involvement and most of them have evidence of Hashimoto's thyroiditis. Paper reported that tests for the serum anti-thyroid antibodies were positive in 83% of the patients with thyroid lymphoma in Japan. Our patient showed no evidence of thyroid dysfunction or Hashimoto's thyroiditis.

Report suggested that secondary adrenal tumors occurred frequently. Secondary involvement of the adrenal gland has been reported in 25% patients with NHL at autopsy. Nevertheless, primary adrenal lymphoma is extremely rare. There have been a few case reports of adrenal insufficiency caused by bilateral adrenal lymphomas, but in our patient there was only a slight elevation of plasma ACTH, despite massive bilateral involvement. On the follow- up examination, adrenal function was convinced to be normal by one-hour standard ACTH stimulation test before the second cycle of chemotherapy.

There are only a few cases with simultaneous involvement of the thyroid and adrenal gland by extranodal NHL. Mostly the cases were Diffuse large B cell lymphomas. Baskal

et al. reported the first case whose initial presentations were symptoms of adrenal insufficiency due to involvement of both adrenal and thyroid glands by high grade immunoblastic lymphoma. More recently Nasu et al. reported a case of diffuse large B cell lymphoma presenting with adrenal insufficiency and hypothyroidism. Dae Ho Lee reported a case with presentation of rapidly enlarged neck mass without thyroid dysfunction. The case was also diagnosed with diffuse large B cell lymphoma and has good response after R-CHOP therapy.

In our case, it is hard to differentiate where the primary site of lymphoma was. Also, the definite pathology of thyroid mass was challenging due to the biopsy was done by needle guide biopsy (not surgery biopsy) so the tissue was small and limited to observe. The final pathology revealed Maltoma by multiple confirmation, however, we still concerned about the possibility of diffuse large B cell lymphoma transformation based on the clinical presentation which was highly aggressive. After R-Chop therapy, we found rapid improvement of both thyroid and adrenal mass during PET/CT follow up so we did not obtain further adrenal biopsy.

Conclusion :

Here we presented a case of bilateral adrenal glands and thyroid Maltoma. It was a indolent type of lymphoma but have aggressive presnetation.