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# Primary adrenal insufficiency secondary to bilateral adrenal lymphoma

## Abstract

Lymphoma is a common malignancy, but involvement of adrenal is less frequent where it can present with spectrum of symptoms ranging from non specific constitutional symptoms to primary adrenal insufficiency which is a very rare presentation. We are reporting a 62 years old lady who presented with adrenal insufficiency in the form of postural drop of blood pressure, weight loss, nausea and vomiting of one month duration, diagnosis was supported by low am cortisol and high ACTH and was found to have bilateral adrenal enlargement, CT guided biopsy after rolling out pheochromocytoma confirmed type-B non Hodgkin lymphoma. She was started on steroid replacement and referd for chemotherapy but the condetion had deteriorated rabidly and she passed.

**Keywords:** primary adrenal insufficiency, Non-Hodgkin lymphoma, bilateral disease

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## Introduction

When Thomas Addison described the disease that now bears his name, bilateral adrenal destruction by tuberculosis was its most common cause. Now tuberculosis accounts for only 7-20 % of cases; autoimmune disease is responsible for 70-90 %, with the remainder being caused by other infectious diseases, adrenal hemorrhage, infarction drugs or replacement by metastatic cancer lymphoma.[1,2]

Lymphomas are cancers that arise from the white blood cells and have been traditionally divided into two large subtypes: Hodgkin and non-Hodgkin lymphoma. B-cell lymphoma is the most common subtype of non-Hodgkin lymphoma; almost 85% of patients with lymphoma have this variant. Lymphomas can potentially arise from any lymphoid tissue located in the body; however, primary adrenal non-Hodgkin lymphoma is extremely rare.[5]

Primary adrenal lymphoma represents 3% of extra nodallymphomas,[3] the symptoms of the disease and response to treatment are variable depending on the type of lymphoma, tumor size, and presence of adrenal insufficiency. Presentation is variable between nonspecific symptoms to primary adrenal insufficiency in bilateral disease.

Most common bilateral adrenal masses in an area which is not endemic for tuberculosis are metastases from malignant tumors arising from lungs, breast, or colon; these are not usually associated with adrenal insufficiency because 90% of the adrenal glands have to be destroyed before adrenal insufficiency becomes clinically apparent [4]. Bilateral cases constitute 70% of the total adrenal lymphomas, the most common type of adrenal Non-Hodgkin lymphoma is diffuse large B cell lymphoma phenotype( DLBCL), comprising 70% of cases.[3]The initial diagnosis can usually be established on the basis of image-guided FNA/ biopsy; however, it is still considered inadequate for diagnosis by many clinicians. Histological diagnosis is a gold standard in the evaluation of lymphoma.

We are reporting a case of 62 years old lady who presented with signs and symptoms of primary adrenal insufficiency secondary to diffuse bilateral involvement of adrenal glands with Non- Hodgkin lymphoma.

## Case history

62 years old Saudi female patient who is known case of long standing diabetes mellitus type 2 requiring insulin and hypertension presented with 3 weeks history of postural dizziness, fatigue, body ache, frequent loose bowel motions with significant weight loss .The family noticed gradual decline in insulin requirement and recurrent hypoglycemia in the previous 1-2 months, they also noticed darkening of sun exposed areas over the face and hands. The history was significant of vague subjective complaints over the preceding 6 months in the form of fatigue, poor appetite, body ache and was prescribed antidepressant with no improvement. On examination, she looked depressed and dehydrated with postural drop of blood pressure (110/72 mmHg supine and 85/60 mmHg setting), hyperpigmentation over face; hands and oral mucosa (Figure 1) (Figure 2), she had no clinically palpable lymph nodes or organometallic. Initial laboratory findings showed: Na 115 mg/dl, K 5.6 mg/dl, $HCO_3$ 22 meq/l, hemoglobin 11 g/l with normal calcium level, hormonal study revealed normal thyroid function test and 8 Am cortisol of 8.8 ng/dl, ACTH 190 ng/dl. Based on these findings the patient was diagnosed to have primary adrenal insufficiency and started on stress dose IV hydrocortisone 100 mg every 6 hours after which she had significant clinical improvement with stabilization of blood pressure and normalization of serum electrolytes, the treatment was shifted to oral steroid replacement but she was requiring supra-physiologic dose to achieve clinical stability, she required hydrocortisone 20 mg Am, 10 mg at lunch time and 10 mg Pm and fludrocortisones 0.05 mg daily. Adrenal CAT -scan with and without contrast showed bilateral heterogeneous adrenal enlargement that keeping with adrenal contour and few enlarged par adrenal lymph nodes (Figure 3) (Figure 4). Adrenal biopsy under CT-guidance was done, histopathology from right adrenal tissue confirmed: non germinal center (activated) b cell like, diffuse large b cell lymphoma. Immunohistochemical markers was done: CD20 strongly positive in neoplastic cells, CD5 positive in background T-cells, CD10 and BCI-6 both are negative, MUM1 strongly positive in neoplastic cells (Figure 5). The patient then was referred to hemato-oncology for treatment.

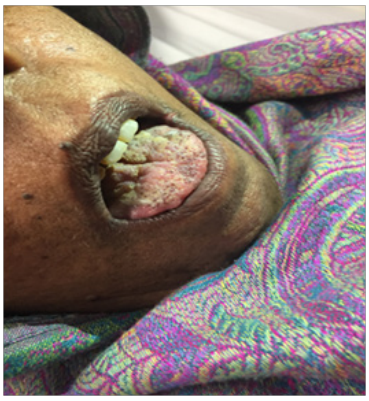


**Figure 1** Hyperpigmentation of tongue and oral mucosa.

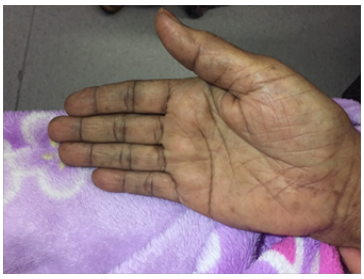


**Figure 2** hyperpigmentation of palmer creases.

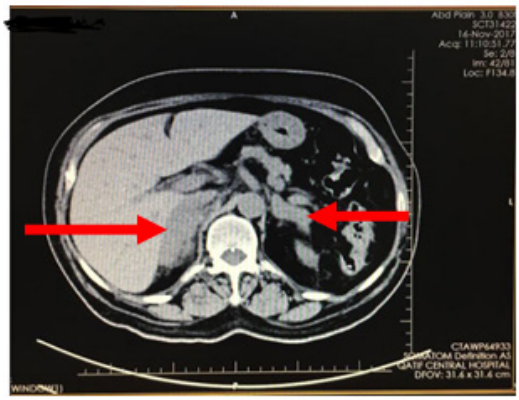


**Figure 3** Non contrast adrenal CT, shows bilateral adrenal enlargement, which are hypo dense maintaining overall shape.

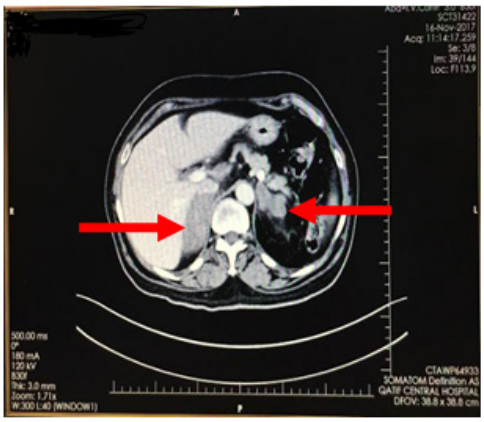
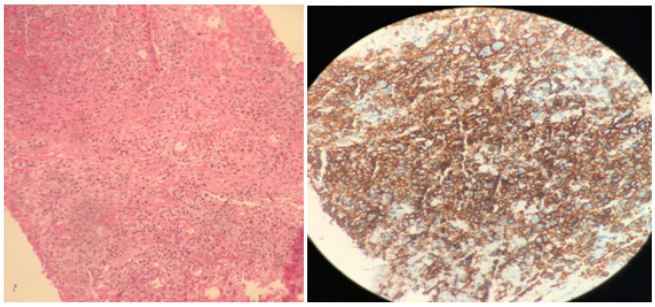
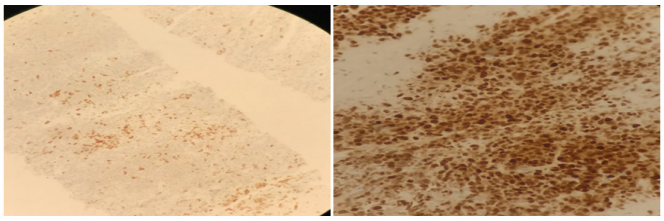


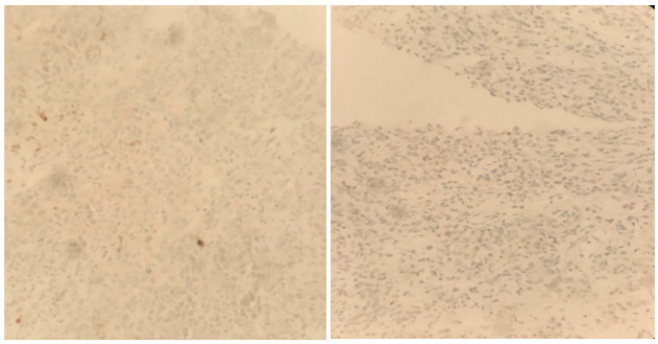
**Figure 4** CT adrenal with contrast, shows heterogeneous enhancement of both adrenal with no calcification and few Para-aortic lymph node enlargements, largest was 1.5 cm indicating an infiltrative process.



H&E stain: diffuse large b cell lymphoma CD20 strongly positive in the neoplastic cells



CD5: positive in the back ground of T-cells. MUM1: strongly positive in neoplastic cells.



CD10: Negative in neoplastic cells BCL6: Negative in neoplastic cells

**Figure 5** Histopathology with immunohistochemistry markers.

## Conclusion

Adrenal lymphoma is an extremely rare but rapidly progressive disease. The presentation is variable , between nonspecific symptoms to varying defects in adrenal function specially if both bilateral glands are involved and should be suspected in patients presenting with bilateral adrenal infiltration and a picture of primary adrenal insufficiency.

## Acknowledgments

None.

## Conflicts of interest

The author declares there is no conflict of interest.

## References

1. Irvine WJ, Barnes EW. Adrenocortical insufficiency. *Clin Endocrinol Metab.* 1972;1(2):549–594.
2. Zelissen PM, Bast EJ, Croughs RJ. Associated autoimmunity in Addison's disease. *J Autoimmun.* 1995;8(1):121–130.

3. Ozimek A, Diebold J, Linke R, et al. Bilateral primary adrenal non-Hodgkin's lymphoma and primary adrenocortical carcinoma--review of the literature preoperative differentiation of adrenal tumors. *Endocr J.* 2008;55(4):625–638.

4. Simpson WG, Babbar P, Payne LF. Bilateral adrenal non-Hodgkin's lymphoma with adrenal insufficiency. *Urol Ann.* 2015;7(2):259–261.

5. Ram N, Rashid O, Farooq S, et al. Primary adrenal non-Hodgkin lymphoma: a case report and review of the literature. *J Med Case Rep.* 2017;11(1):108.

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