Case Report

ACTH-independent Cushing’s syndrome due to bilateral adrenocortical adenoma: A case report

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ABSTRACT

The chronic excess of glucocorticoids results in Cushing's syndrome. Cushing's syndrome presents with a variety of signs and symptoms including: central obesity, proximal muscle weakness, fatigue striae, poor wound healing, amenorrhea, and others.

ACTH-independent Cushing’s syndrome is usually due to unilateral adenoma. A rare cause of it is bilateral adrenal adenomas.

In this paper we report a case of a 43-year-old woman with Cushing’s syndrome due to bilateral adrenal adenoma.

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Introduction

Cushing's syndrome (CS) is caused by chronic excess glucocorticoids [1]. It is associated with increased morbidity and mortality due to hypertension, diabetes, coagulopathy, cardiovascular disease, infections, and fractures. [2]. The excess of glucocorticoid can be endogenous or exogenous. Endogenous CS can be divided into adrenocorticotropin hormone (ACTH)-independent and ACTH-dependent forms [3]. ACTH-independent CS accounts for 15-20% of endogenous CS cases, mostly caused by unilateral adrenocortical adenomas [4]. Bilateral adrenocortical adenoma (BAA) is a rare cause of ACTH-independent CS [3].

In this paper, we report a case of a 43-year-old woman suffering from Cushing's syndrome due to bilateral adrenocortical adenoma (BAA).

Case presentation

A 43-year-old female patient presented to endocrinology clinic at Al-Mouwasat University Hospital complaining of fatigue, lower back pain, increased weight, muscle weakness, and amenorrhea. She is a smoker but consumes no alcohol.

The patient's history included left hip joint replacement 1 year ago (fracture after trauma) and hypertension since

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3 years which was not well-controlled by medication. There is no family history of hypertension but a history of diabetes mellitus was found. She also mentioned a history of irregular steroid use.

Her vital signs at admission were as follows: blood pressure 150/90 mm Hg, heart rate = 77 beats per minute, body temperature = 36.5°C, respiratory rate = 20 breathes per minute, oxygen saturation = 98% on room air.

On physical examination, we found ecchymosis, thinning of the skin, purple striae, buffalo hump, lower limbs edema, and proximal muscle weakness with no evidence of acne or hirsutism.

Biochemical and radiological assessment were done. Laboratory work-up found a high level of fasting blood glucose (135 mg/dL; normal range: 70-99 mg/dL), slightly elevation of HbA1c level (6.5%; normal range: 5.7%-6.4%), normal level of cholesterol and triglycerides (total cholesterol = 176 mg/dL; normal range: <200 mg/dL, triglycerides = 145 mg/dL; normal range: <150 mg/dL), and low level of 25(OH) vitD (15.6 ng/mL; normal range: 30-50 ng/mL). A high level of midnight serum cortisol was observed (37.8 mcg/dL; normal range: <7.5 mcg/dL) and low level of ACTH (2 pg/mL; normal range: 10-60 pg/mL) that was followed by cortisol suppression test which was abnormal (37.1 mcg/dL). Plasma metanephrine and plasma normetanephrine were normal (61 pg/mL; normal range: 0-140 pg/mL and 151 pg/mL; normal range: 70-1700 pg/mL, respectively).

Magnetic resonance imaging (MRI) of pituitary gland revealed evidence of a partial empty sella turcica, however, Multi-slice Computed Tomography (MSCT) of the abdomen showed bilateral, heterogeneous, well-demarcated adrenal masses with moderate contrast enhancement with no evidence of hemorrhage or calcification (Fig. 1).

Laparoscopic bilateral adrenalectomy was performed and sent to pathology lab for examination. The level of serum cortisol has been significantly reduced after surgery and the pathological evaluation confirmed the diagnosis of bilateral adrenocortical adenomas (Figs. 2 and Figs. 3).

She was administered with oral prednisolone 10 mg BID (5 mg at morning + 5 mg at night), 0.2 mg of fludrocortisone, 1000 mg of metformin and supplemental calcium with vitamin D3. Clinical and laboratory follow-up showed improvement of her fatigue, blood pressure 110/80, muscle strength, and fasting blood glucose (80 mg/dL).

**Discussion**

Adrenocortical adenoma is the most common benign tumor of the adrenal gland, it accounts for approximately 52% of adrenal tumor cases [5]. The prevalence of adenoma varied according to age, it is 0.14% in patients with age of 20-29 years and 7% in those with older than 70 years [6].

Adenomas are mostly unilateral [4] and the frequency of bilaterality is only 20% of cases in one series [7].

Adrenocortical adenoma may secret glucocorticoids, aldosterone or sex hormones causing different clinical manifestations diseases [8]. Glucocorticoid-producing adenoma
The radiological findings of patient with bilateral adrenal mass vary based on the underlying pathology. In the case of a low level of ACTH, the differential diagnosis of ACTH-independent CS includes: BAAs, primary pigmented nodular adrenocortical disease, ACTH-independent macronodular adrenal hyperplasia (AIMAH) [3].

The definitive diagnosis of adrenocortical adenoma puts according to pathology evaluation. Tumors associated with CS present as sharply circumscribed mass that usually measure 3 to 4 cm in average diameter. Adenomas are most often composed of small nests, cords, or alveolar arrangements of vacuolated (clear) cells that most closely resemble those of the normal fasciculate [13]. Our case has typical findings.

Treatment is laparoscopic unilateral or bilateral adrenalectomy. Bilateral adrenalectomy leads to rapid resolution of hypercortisolemia and related morbidity but patients need lifelong glucocorticoid and mineralocorticoid replacement [1].

Conclusion

Although Cushing’s syndrome due to bilateral adrenal adenoma is rare, it should be considered in mid-aged females with typical symptoms and radiological assessment finds bilateral well-encapsulated adenomas of 2.0-3.5 cm in diameter, with the presence of single or multiple nodules.

Patient consent

Patient consent has been obtained.

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