BILATERAL LARGE ADRENAL LESIONS IN A PATIENT WITH UNDERTREATED CONGENITAL ADRENAL HYPERPLASIA

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INTRODUCTION

- Congenital adrenal hyperplasia (CAH) is a group of autosomal recessive diseases characterized by enzyme deficiencies in cortisol secretion.
- The most common form is 21-alpha hydroxylase deficiency.
- Adrenal cortex hyperplasia can be detected in inadequately treated CAH cases. However, adrenal incidentalomas are reported rarely and when present, tumor size and adrenal size are positively correlated with the 17-OH progesterone levels.
- Here, we report a patient with undertreated CAH and bilateral large adrenal masses

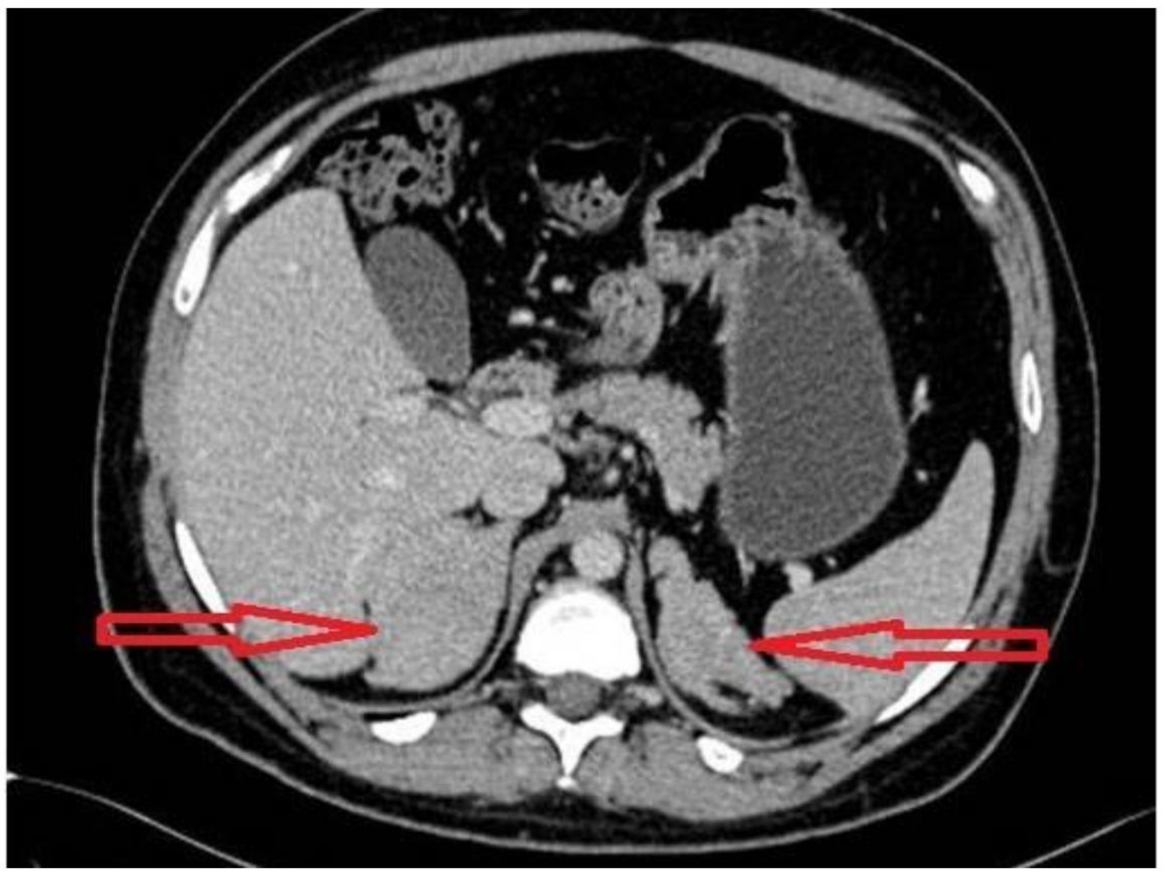
CASE REPORT

- 34 years old male patient diagnosed with CAH and testicular anorchia at the age of 7 admitted to our clinic for general weakness.
- Hydrocortisone treatment was started at the diagnosis but he never used it regularly and was not taking glucocorticoid replacement for 10 years.
- In physical examination, blood pressure was 100/70 mmHg, there was diffuse hyperpigmentation, and no testicular tissue could be palpated.
- In laboratory examination, fasting blood glucose was 97 mg/dL (74-106), sodium 143 mmol/L (136-145), K 4.5 mmol/L (3.5-5.1) and renal, liver and thyroid functions were normal. His serum cortisol, adrenocorticotrophin hormone (ACTH) and 17-OH progesteron levels were 4.8 mcg/dL (6.24-18), 366 pg/mL (0-60) and 217 ng/mL (0-1.39), respectively. Serum aldosteron was low and and renin was high (Table-1).
- In abdominal CT (computed tomography), there were hypertrophied adrenal glands with solid nodular lesions of 47x44 mm in right and 22x24 mm in left glands (Figure -1)
- Pheochromocytoma was excluded by normal 24 hour urinary catecholamines.
- 17-OH progesteron decreased to 40.8 ng/mL after 2 months of 0.5 mg/day dexamethasone treatment.

Table 1. Laboratory data of the patient

	Laboratory results	Normal range
Glucose	97 mg/dl	74-106
Sodium (Na)	143 mmol/L	136-145
Potassium (K)	4.5 mmol/L	3.5-5.1
Creatine	0.96 mg/dl	0.7-1.2
Thyrotrophin (TSH)	3.3 uIU/ml	0.27-4,2
Cortisol	4.8 ug/dl	6.24-18
ACTH	366 pg/Ml	0-60
17-OH progesteron	217 ng/mL	0-1.39
Renin	117.7 uIU/ml	5.3-99.1
Aldosteron	7.1 ng/dl	3.7-43.2

Figure -1. Abdominal computerized tomography



CONCLUSION

- Although, adrenal enlargement is an expected finding in untreated CAH due to excessive ACTH secretion, adrenal lesions are rarely observed.
- CAH should be included in the differential diagnosis of large bilateral adrenal lesions to avoid unnecessary adrenalectomies.