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***P-12 ACTH-INDEPENDENT CUSHING SYNDROME
WITH INCONCLUSIVE ADRENAL CT SCAN
RESULTS***

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Clinical Case: We present the case of a 21-year-old lady who visited the Faiha Specialized Diabetes, Endocrine, and Metabolism Center (FDEMC) in Basrah, south of Iraq in 2022. She presented for evaluation of gradually increased hirsutism and secondary amenorrhea. Menarche started at the age of 11 years old; it was regular then, at the age of 13, the patient began to have menstrual irregularities with cycles every 2-3 months and in the same time she noticed gradually increased hair growth in her upper chin, abdomen, and back until the age of 19 years when she visited our center for medical advice. By this time, the patient had secondary amenorrhea; the last cycle was more than 6 months ago. Clinical examination

revealed moon face, facial plethora, acanthosis nigricans, buffalo hump, lemon-on-stick appearance, and central obesity with wide purple striae more than 1 cm in diameter. The Ferriman-Gallwey score (FG score) was 30/36, with no frontal baldness, BP 130/90, her height was 148 cm and her BMI was 28.7 kg/m². When Carney's complex features were inspected, she had no skin lesions and her Echocardiogram was normal. Drug history was negative.

Family history revealed and older sister diagnosed with ACTH-independent Cushing syndrome, who died 10 years ago, on day 4 post-bilateral adrenalectomy, due to complications.

Laboratory results revealed cortisol: 23 µg/dL, ACTH <1 pg/mL, DHEAS: 400 µg/dL, total testosterone: 163ng/dL, estradiol: 39.7 pg/mL, and 1-mg overnight dexamethasone suppression test: 21.8 µg/dL. TSH, FSH, LH were normal. Pelvic ultrasonography was normal, apart from mild PCOS changes.

The Initial CT-scan of the adrenal gland was normal and pituitary MRI was also normal. ACTH-independent Cushing syndrome was diagnosed, and due to the unfortunate death of her sister, the patient pursued medical treatment for now, ketoconazole tab 200 mg TID was started with interval CT of the adrenals to assess any future changes. A DEXA scan was done with a total Z-score of -1.5. Serum cortisol basal and 2-hours postprandial was done twice on two different occasions and didn't show any significant difference. After 2 months of ketoconazole treatment, the menstrual cycle resumed but was irregular, being every 14 days (polymenorrhea) and she developed hypertension. Her serum potassium levels did not change from the baseline and stayed around 4.5 mmol/L, and her HbA1c was 5.7. **Conclusion:** The patient is now on ketoconazole tab 200 mg four times a day; her FG score is 26/36; regular cycle and the BP is controlled with spironolactone tab 50mg OD. We don't have genetic studies, and the only available drug that is suitable for her condition is ketoconazole, which is being up-titrated until the maximum dose is reached. The patient is open to the surgical option if offered any in the future.

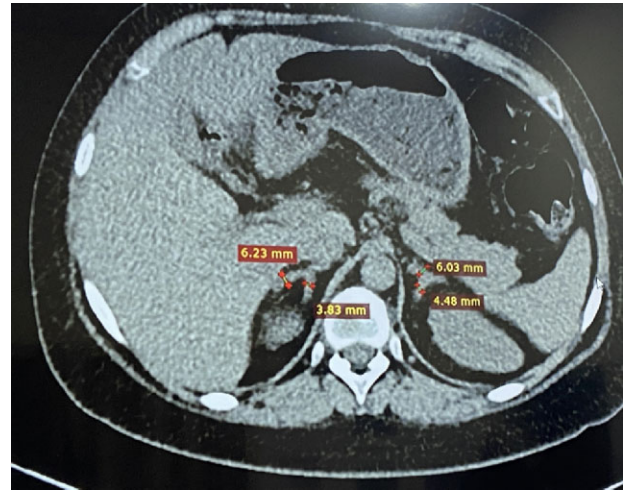


Figure 1: CT scan of adrenal glands
This is the second CT scan of adrenal showing thickening of the adrenals, this was taken in November 2023 after 6 months from the initial CT scan.

Table 1: Biochemical evaluation after starting ketoconazole tab 200mg
Biochemical evaluation after starting ketoconazole tab 200mg three times daily

Date of visit	Cortisol (µg/dL)	ACTH (pg/mL)	DHEAS (µg/dL)
25-9-2023	7.6	<1	63
21-11-2023	25.6	<1	343
7-2-2024	5.3	<1.5	274
23-4-2024	25	3.4	174

Table 2: Initial biochemical evaluation done to the patient at the FDEMC
This table shows the initial visits results of the patient done at FDEMC, which supports the diagnosis of Cushing syndrome. ONDST: 1 mg overnight dexamethasone suppression test

Date of visit	Cortisol (µg/dL)	ACTH (pg/mL)	DHEAS (µg/dL)	ONDST (µg/dL)
15-5-2023	23.5	<1	400	
24-5-2023				21.8
19-6-2023	40.3	<1	262	
20-6-2023				45.6