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CUSHING'S SYNDROME SECONDARY TO ABERRANT HORMONE RECEPTORS IN A PATIENT WITH MACRONODULAR ADRENAL HYPERPLASIA ACTH-INDEPENDENT

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INTRODUCTION

Cushing's syndrome (CS) due to macronodular adrenal hyperplasia ACTH-independent (MAHAI) is rare. Regulation of cortisol secretion by G-protein-coupled receptors (GPCRs) aberrantly expressed is frequently found in MAHAI. Various aberrant receptors have been reported, mostly: GIP, vasopressin, beta-adrenergic, LH/ hCG and serotonin.

CASE REPORT

61 years old woman.

Medical history: type 2 diabetes, hypertension, obesity, dyslipidemia and severe sleep apnea. Treatment: metformin, insulin, lyxisenatide, irbesartan-hydrochlorothiazide, torasemide, diltiazem, acetyl salicylic acid and atorvastatin.

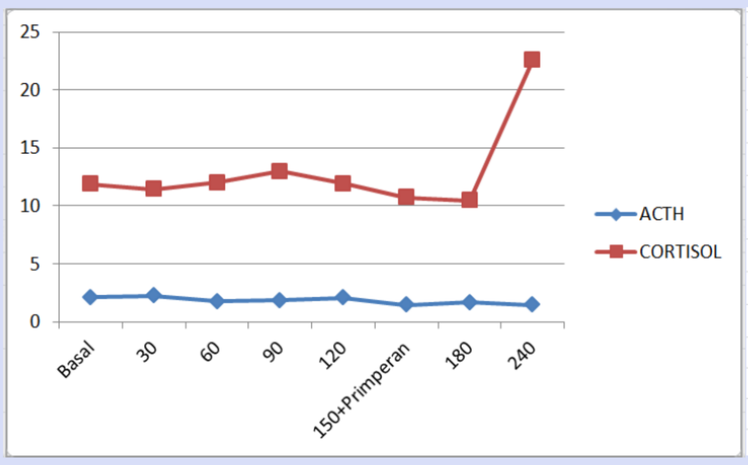
Physical examination: moon face, centripetal obesity, abdominal striaes and thin extremities.

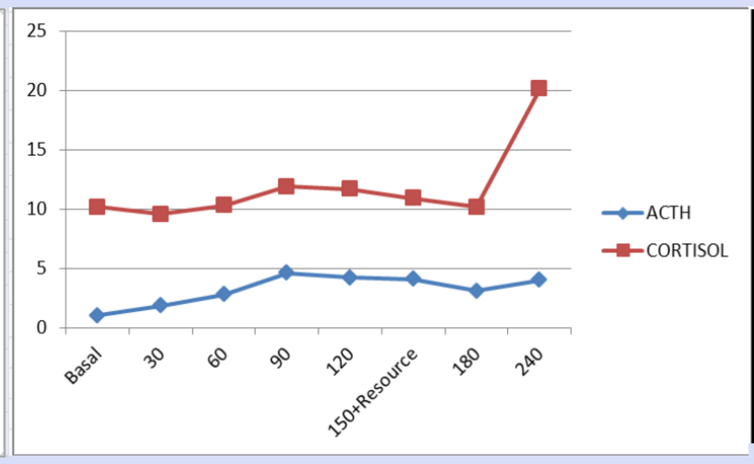
Laboratory evaluation: HbA_{1c} 8.4%, TSH 2.06 µIU/ml, serum cortisol: at 8 a.m.:17.2µg/dl (5-25), overnight dexamethasone suppression (1mg): 5.3µg/dl, after 0.5 mg dexamethasone every 6 h for 48 h suppression: 5.03µg/dl, after nocturnal 8 mg dexamethasone suppression: 5.7 µg/dl. Urinary free cortisol repeatedly normal (49.4 and 24 µg/24 h), salivary cortisol repeteadly reased (0.382 and 0.292 ng/ml). ACTH 2.83, others adrenal hormonal profile normal.

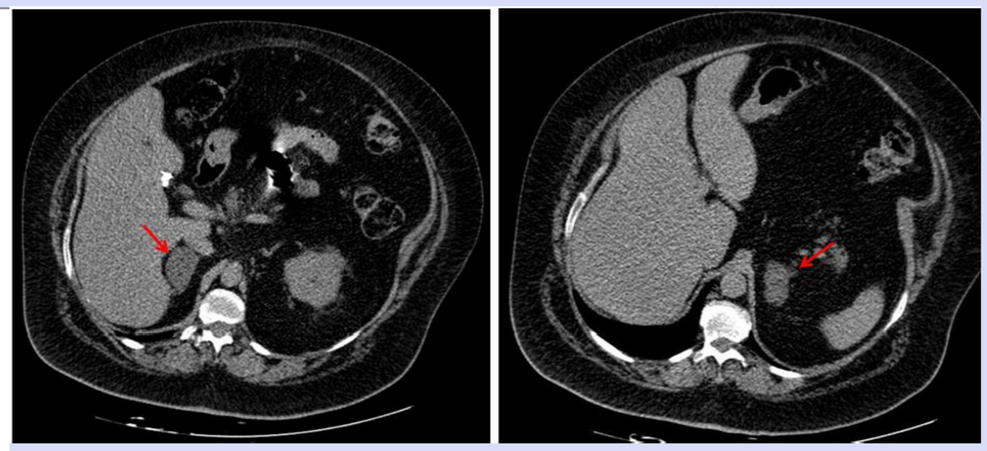
With these findings, the suspected diagnosis was Cushing's syndrome.

Because of the known association between MAHAI and aberrant GPCRs some tests were performed: posture test, standard mixed meal, LHRH (100 mg iv), metroclopramide (10 mg orally), glucagon (1 mg iv) and AVP (10 IU im).

Right adrenalectomy is planned.







Positive response (50% increase in plasma cortisol levels) in the standard mixed test and metroclopramide test (indicating serotonin and GIP aberrant receptors).

CT: adrenal glands enlarged with homogeneous hypodense nodules (3.3 x 2.6 cm right; 3.2 x 1.9 cm left).

CONCLUSION

In patients with MAHAI aberrant GPCRs can be identified. The detection of such aberrant receptors is necessary in all patients with MAHAI. Identification of these receptors can provide specific pharmacological treatment with or without adrenalectomy

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