

Adrenocortical Malignancy and Hypercortisolism Manifested as Uncontrolled Hypertension : A Case Report and Review of Literature

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Abstract

Adrenocortical carcinoma is one of the rare malignancies with worst prognosis. It presents with mass effects and less commonly with features of hormone excess (About 60% of all adrenocortical carcinoma's). Hormonally active adrenocortical carcinomas most commonly secrete cortisol while the co-secretion of multiple steroid hormones is rare. We report the case of a 47- year-old woman presenting with weakness of lower limbs, truncal obesity and growth of terminal hair on upper lip for one and half months. She was diagnosed Cushing Syndrome due to malignant adrenal tumour by abdominal CT scan and serum cortisol and ACTH. This case was inoperable and finally she received palliative chemotherapy. In cases of adrenocortical malignancy hormonal status should be investigated as the pattern of hormone secretion may be a clue to the malignancy of an adrenal lesion. In addition, more data are needed to clarify the clinical and prognostic significance of the combined production of all adrenocortical hormones affecting either the survival or the quality of life of patients with adrenocortical malignancy.

Key words: Cushing Syndrome, Adrenocortical Malignancy, Hypercortisolism

Introduction

Adrenocortical malignancy accounting for 0.05-0.2% of all cancers with worst outcome depending on the stage of the disease and the completeness of the resection^{1,2}. About 60% of adrenocortical malignancies are hormone-secreting and the steroid profile often displays a wide variety of steroids in adrenocortical malignancies, which can be used as tumour markers. Most commonly adrenocortical malignancies secrete cortisol while the combined secretion of all adrenocortical hormones is quite rare with unknown either clinical or prognostic significance³⁻⁵.

Case presentation

In March 2017, a 45 year old woman was admitted in the department of Internal Medicine, Bangabandhu Sheikh Mujib Medical University (BSMMU) with weakness of both lower limbs which makes her unable to stand from squatting, heaviness of central abdomen and growth of terminal hair on upper lip for one and half months. She had anorexia with subjective complaints of weight gain without any documentation. She had no history of steroid use in any form. On examination her face was puffy, plethoric, terminal hair on right side of upper lip, mildly anaemic. All limbs were thinner with abdominal swelling. No striae or skin colour change. She was hypertensive (BP 150/90mm Hg), bedside urine was positive for sugar.

Nervous system examination revealed wasting of both upper and lower limbs with proximal weakness (Upper limb-Proximal MRC grade 3, distal 5, lower limb-proximal 2, distal 4). No other abnormality. Other system normal. Endocrino-logical investigation, revealed potassium level was at lower limit (serum potassium: 3.6 mmol/L, Na⁺ 138 mmol/L), random blood sugar was 14.5 mmol/L, ACTH was <5 pg/ml (ND- 46pg/ml), cortisol >2069.25nmol/L (138-690nmol/L). But 24 hrs urinary cortisol was within range- 164.3 nmol/24hrs (11.8-485nmol/24hrs) as there was difficulty in sample collection. Aldosterone level was not measured. Ultrasonogram of whole abdomen revealed right supra renal mass with bilateral nephrocalcinosis and hepatomegaly (16.1cm). Subsequently CT scan of abdomen with contrast was done that revealed right adrenal malignant mass (8.6 × 7.5 cm) with radiological characteristic of malignancy, bilateral nephrocalcinosis mild ascites and intra abdominal lymphadenopathy.

The patient was started on losartan potassium treatment, with gradual improvement of her blood pressure, metformin for high blood sugar. After consultation surgery and oncology department it was decided to give palliative chemotherapy without further invasive diagnostic procedure and she was admitted to the department of oncology for palliative chemotherapy.

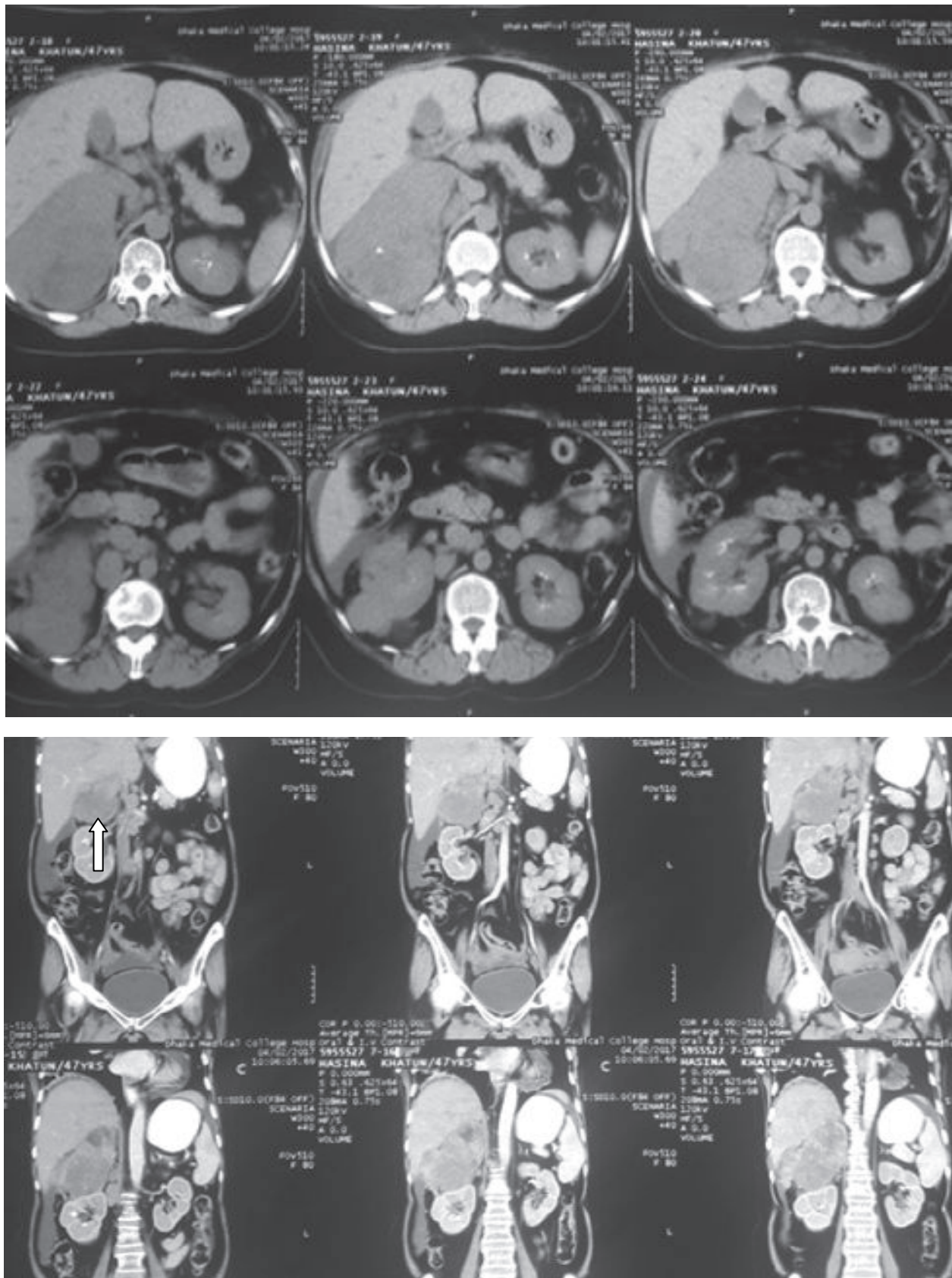


Fig.-1: Contrast enhanced CT scan of abdomen showing right adrenal mass (13.5×7.5cm)

Discussion

We report a patient with adrenocortical malignancy with hypercortisolism. She presented with features of Cushing syndrome (central obesity, proximal weakness, growth of terminal hair, hyperglycaemia and new onset hypertension with hypercortisolism and low ACTH) within very short time of one and half month without any history of steroid use. Serum electrolytes level showed potassium level on lower limit with normal sodium level. There was hyperglycaemia. ACTH level was low and cortisol level was very high (>2069.25 nmol/L). Her 24 hrs urinary free cortisol was within range (164.3 nmol/24hrs) possibly due to difficulty in sample collection (She could not use pan without assistance). Ultrasonogram of abdomen showed right supra renal mass (6×5.5 cm) with bilateral nephrocalcinosis and hepatomegaly (16.1 cm). CT scan of abdomen with contrast showed large right adrenal malignant mass (13.5×7.5 cm) with radiological characteristic of malignancy, bilateral nephrocalcinosis mild ascites and intra abdominal lymphadenopathy. Tumour morphology was inhomogeneous with irregular margin, there was mild contrast enhancement. Besides these findings, there was mild ascites and abdominal lymphadenopathy. These findings on CT scan are confidently suggesting an adrenal mass of malignant origin. Moreover, correlation with clinical features, she had anorexia and symptoms developed over very short time (one and half months) which best fits with a malignant adrenal tumour. As there was advanced stage (Ascites and abdominal lymphadenopathy) further tissue sampling was not done. Adrenal tumour secretes hormones in 60% cases and common hormone is cortisol⁴.

It has been reported that adrenocortical malignancies may produce hormones and the steroid profile often displays a wide variety of steroids, which may be used as tumour markers to

detect metastatic disease. Less frequently, adrenocortical malignancies produce combined hormonal syndromes most commonly cortisol with androgens (up to 46.7% of cases) while the combined hypersecretion of all adrenocortical hormones occurs rarely³⁻⁵. Increased cortisol production has been characterized as a poor prognostic factor for adrenocortical malignancies.⁶

Conclusions

In conclusion, the hormonal status should be carefully investigated in all cases of suspected adrenocortical malignancies, as the pattern of hormone secretion may be a clue to the malignancy of an adrenal lesion⁷. In addition, further data are needed to clarify the growth rate of the adrenocortical malignancies and the clinical and prognostic significance of the combined production of all adrenocortical hormones affecting either the survival or the quality of life of patients with adrenocortical malignancies.

Abbreviations:

MRC, medical research council; ACTH, adrenocorticotropic hormone; CT, computed tomography.

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