中文題目：以庫欣氏症為表現之腎上腺皮質癌

英文題目：A Case of Adrenal Cortical Carcinoma Presented with Cushing's Syndrome

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Background:

The incidence of adrenal tumors is estimated to be 3 to 10%. About 36 to 94% of them are reported to be non-functional benign adrenal adenomas. On the other hand, a minority of adrenal tumors is functional with hyper-secretion of adrenergic hormone, cortisol, aldosterone or sex hormone. Although functional adrenal tumors are often benign, malignancies are still reported with the incidence less than 1%. We reported a rare case of adrenal cortical carcinoma with initial presentations of hypercortisolism.

Case Report:

A 44-year-old female, with dizziness, lower limb weakness, and amenorrhea for one year, visited our outpatient department for survey. Physical examination showed high blood pressure (180/100 mmHg), moon face, buffalo hump, purple striae, and central obesity. Hypokalemia (serum potassium 1.5mmol/L) and metabolic alkalosis were noted. Abdominal sonography revealed a huge left intra-abdominal tumor (15.7x12.4x16.5cm) over the area of left adrenal gland, adjacent to stomach and liver. The exams of hormone levels showed cortisol 65.20μg/dL and adrenocorticotropic hormone (ACTH) 8.01pg/mL. In addition, the level of aldosterone was 9.74ng/dL with aldosterone-renin ratio of 4.27, and the urine vanillylmandelic acid was 5.98mg/day. The pathology of excisional biopsy of the tumor was adrenal cortical carcinoma. Left adrenalectomy was then performed, and the surgical specimen also proved adrenal cortical carcinoma. The patient received adjuvant chemotherapy with etoposide, doxorubicin, and cisplatin. After eight courses of the chemotherapy, her Cushing appearance gradually resolved, and body weight decreased from 76Kg to 69Kg. Laboratory data showed improvement with Cortisol 1.30μg/dL, ACTH 22.94pg/mL, and serum potassium 3.7mmol/L.

Discussion:

Functional malignant adrenal tumors are rare but still existed. Clinicians should always have a detailed history taking and survey for the adrenergic hormone levels when managing adrenal tumors with malignant characteristics seen on image study. Adrenal cortical carcinomas most frequently present as sporadic tumors, and recent studies about pathogenesis have focus on the role of genetic mutations including various oncosuppressor genes and oncogenes. Further genetic studies should be carried out to elucidate the possible causative gene leading to adrenal cortical carcinoma in our case.