

Esra Tatal<sup>1</sup>, Demet Yilmazer<sup>1</sup>, Taner Demirci<sup>1</sup>, Evrim Cakır<sup>1</sup>, Salih Sinan Gültekin<sup>1</sup>, Bahadır Celep<sup>1</sup>, Oya Topaloğlu<sup>1</sup>, Erman Çakal<sup>1</sup>, Özlem Özmen<sup>2</sup>, Tuncay Delibaşı<sup>3</sup>

<sup>1</sup>Diskapi Yildirim Beyazit Teaching and Research Hospital, Ankara, Turkey

<sup>2</sup>Ataturk Chest Diseases and Thoracic Surgery Training and Research Hospital, Ankara, Turkey

<sup>3</sup>Hacettepe University (Kastamonu), School of Medicine, Ankara, Turkey

## Introduction

Ectopic adrenocorticotrophic hormone (ACTH) syndrome is characterized by hypercortisolism due to the hypersecretion of non-pituitary ACTH-secreting tumor leads to the Cushing's syndrome. Herein, we present a case with Cushing's syndrome, which is diagnosed ACTH-secreting renal malignant paraganglioma.

## Case Report

A 40-year-old woman presented with a five month history of newly diagnosed hypertension and diabetes, weakness, hyperpigmentation, oligomenorrhea, hirsutism and acneiform lesions. She showed cushingoid features including moon face, facial hirsutism, facial and truncal acne, hyperpigmentation and severe muscle weakness of the limbs. She had not the findings including striae, supraclavicular fat accumulation and buffalo hump. Laboratory examination showed the presence of hypopotasemia, hyperglycemia, hyperthyroidism and leukocytosis. Serum levels of ACTH, cortisol and the urine free cortisol were markedly elevated. Cortisol value after an overnight 2 mg dexamethasone suppression test was 46.1 mcg/dL and there were no suppression after 2 day 8 mg dexamethasone administration. Magnetic resonance imaging (MRI) of the pituitary gland indicated two microadenomas. An abdominal MRI scan revealed horseshoe kidney, bilateral adrenal hyperplasia and a mass with dimensions of 35x31 mm in the left kidney. Inferior petrosal sinus sampling showed no evidence for a central to peripheral gradient of ACTH. A positron emission tomography/computed tomography scan was showed intense increased activity in the lower pole of left kidney. Left adrenalectomy and left partial nephrectomy were performed. The resected tumor were diagnosed as the ACTH-secreting paraganglioma in the pathological examination that was confirmed by immunohistochemical studies with chromogranin, synaptophysin and ACTH. The periadrenal lymph node was evaluated as a metastatic lymph node.

## Conclusions

Only few cases of paragangliomas as the cause of the ectopic ACTH syndrome have been reported. To the best of our knowlege, this is the first case of renal malignant paraganglioma resulting in Cushing's syndrome due to the ectopic ACTH hypersecretion.

