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A Rare Case of Aldosterone-Producing Adrenocortical Carcinoma with Co-Secretion of Cortisol and Estradiol

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Introduction:
Adrenocortical carcinoma is a rare malignancy with an incidence of one to two per million population per year. Aldosterone-producing adrenocortical carcinoma comprises less than 7% of all functioning adrenocortical carcinomas. We report a rare case of adrenocortical carcinoma with a clinical picture of primary aldosteronism and subclinical Cushing's syndrome and feminization.

Case Presentation:
An 18-year old male presented with uncontrolled hypertension, recurrent bilateral leg weakness and hypokalemia. He had gynecomastia on physical examination without signs of hypercortisolism. Plasma aldosterone concentration and aldosterone/renin ratio were elevated. Adrenal CT scan revealed a 4.7 x 4.1 x 4.8 cm left adrenal mass with enhancement features suggestive of a lipid-poor adenoma. Hormonal evaluation showed hypercortisolism and elevated estradiol. He underwent laparoscopic adrenalectomy. Histopathologic diagnosis was adrenocortical carcinoma based on Weiss criteria and immunohistomorphology with Ki67 index of 5-10%. After complete surgical resection, his blood pressure normalized and aldosterone, cortisol and estradiol levels returned to normal.

Conclusion:
In the work up of suspected adrenal carcinoma, complete hormonal evaluation is necessary even if clinical symptoms are absent. The pattern of tumor secretion and tumor characteristics on CT scan may point to the malignant potential of the tumor. Complete surgical resection is the cornerstone of treatment and Ki67 index is the most powerful prognostic marker and used to guide treatment decisions. Long-term monitoring is recommended with imaging and hormonal evaluation used as tumor markers for recurrence.