

MALIGNANT HYPERTENSION LEADING TO A DIAGNOSIS OF AN ALDOSTERONE AND CORTISOL CO-SECRETING ADRENAL TUMOR

Dereje Desta, MD; Leigh M. Eck, MD; Becky N. Lowry, MD

INTRODUCTION

Only 10 to 15% of adrenal incidentalomas are classified as hyper-functioning. Of functional tumors, the majority are subclinical Cushing's syndromes, followed by pheochromocytomas and aldosterone producing adenomas. Aldosterone and cortisol co-secreting tumors are rare. Current clinical practice guidelines have limited recommendations regarding the evaluation and management of co-secreting adenomas.

CASE PRESENTATION:

An 86 year old female with long standing, resistant hypertension despite a five drug regimen, was transferred for management of hypertensive emergency. Due to notable hypokalemia on presentation, a work up for primary aldosteronism (PA) was pursued with a suggestive screening aldosterone-to-renin ratio of 38. PA was confirmed with an elevated aldosterone level on a salt loaded 24-hour urine collection. Adrenal imaging revealed a right adrenal mass, likely an adrenal adenoma. In light of this finding, screening for Cushing's syndrome and pheochromocytoma was undertaken to complete the hormonal evaluation. Notably, a 24-hour urine free cortisol was elevated at 121µg/24hrs with a subsequent abnormal overnight dexamethasone suppression test confirming a diagnosis of subclinical Cushing's syndrome. This was ACTH independent in etiology confirming an adrenal source. Laparoscopic adrenalectomy was pursued with pathology revealing a 4.2 x 3.1 x 3.0 cm adrenal nodule. The patient did well post-operatively requiring only one blood pressure agent.

Test	Result	Normal Value
Aldosterone	23	<22 ng/dl
Renin	0.6	2.9-10.8 ng/ml/hr
PAC/PRA	38	<30
24 hr UFC	121	3.5-45 µg/ml/hr
1mg DST	9	< 1.8 µg/dl
24 hr urinary catecholamine and metanephrine	Normal	
UFC : Urinary free cortisol		
DST: Dexamethasone suppression test		

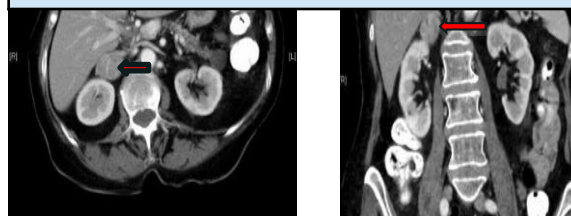


Figure 1: CT scan of the abdomen without intravenous contrast revealing a right adrenal mass, left transverse view and right coronal view

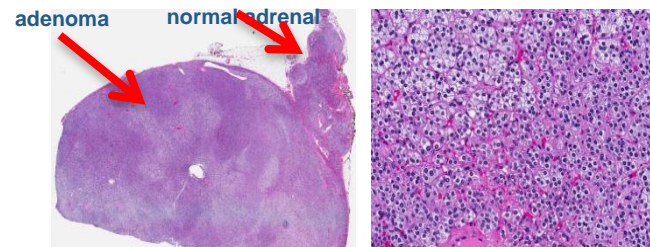


Figure 2: Pathology slides, low power (left) and high power (right), showing the similarity of the adenoma cells to the non-neoplastic cells of the adrenal cortex; the cells are bland, without nuclear pleomorphism.

DISCUSSION

Primary aldosteronism is a common cause of secondary hypertension. Although screening for pheochromocytoma, Cushing's syndrome and PA is recommended for the work up of an adrenal incidentaloma, expert guidelines for the evaluation of PA do not mandate the same comprehensive hormonal evaluation. Had this screening not been undertaken in our patient, a diagnosis of an aldosterone and cortisol co-secreting adrenal tumor would have been missed.

1. Nieman LK, Biller BM, Findling JW, et al. The diagnosis of Cushing's syndrome: an Endocrine Society Clinical Practice Guideline. *J Clin Endocrinol Metab* 2008; 93:1526-1540
2. John W. Funder, Robert M. Carey, et al. Case detection, diagnosis, and treatment of patients with primary aldosteronism: an endocrine society Clinical Practice Guideline. *J Clin Endocrinol Metab* 2008; 93: 3266-3281
3. Mantero, F, Terzolo, M, Arnaldi, G, et al. A survey on adrenal incidentaloma in Italy Study group on adrenal tumors of the Italian society of endocrinology and metabolism 2000 ; 85,637-644.
4. Piaditis GP, Kaltsas GA, Androulakis II, et al. (2009) High prevalence of autonomous cortisol and aldosterone secretion from adrenal adenomas. *Clin Endocrinol (Oxf)* 71: 772-778.
5. NIH Consensus Panel. NIH state of the science conference on the management of the clinically in apparent adrenal mass. 2002.