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# Adrenocortical Carcinoma: A Stressful Diagnosis

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# ADRENOCORTICAL CARCINOMA, A STRESSFUL DIAGNOSIS

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## Introduction

Adrenocortical carcinoma (ACC) is a rare and biologically aggressive malignancy. The incidence of ACC is between 0.7-2 cases per million, accounting for 0.05-2% of all malignant tumors[1].

Cortisol-producing ACC is the most common type, occurring in 30-40% of all ACCs[1].

Patients classically present with features of elevated cortisol: refractory hypertension, glucose intolerance, and central obesity[2].

We present a case of a 75-year-old male with a cortisol-secreting variant of adrenocortical carcinoma.

## Case Report

A 75-year-old male, with a prior medical history significant for hypertension, type two diabetes, and severe, untreated obstructive sleep apnea, presented to the emergency department with a blood pressure of 259/123 and respiratory distress.

Computerized tomography angiography (CTA) revealed a heterogeneous slightly lobulated, macroscopic fat-containing mass in the left adrenal gland, measuring up to 4 cm (image A and B). Initial biochemical work up was unremarkable for a hypertensive etiology.

A year later, imaging revealed the same left adrenal mass, now measuring 7.1 cm x 5.8 cm with extensive mediastinal and hilar lymphadenopathy (image C and D).

Left adrenal mass core biopsies revealed benign adrenocortical neoplasm, favoring adenoma, negative for metastatic carcinoma.

Hospital course was complicated by bacteremia, septic shock requiring stress-dose steroids, and respiratory failure.

Subcarinal lymph node fine needle aspiration revealed malignant cells, consistent with metastatic adrenocortical carcinoma.

Further work up was deferred, as the patient pursued hospice.

## Images

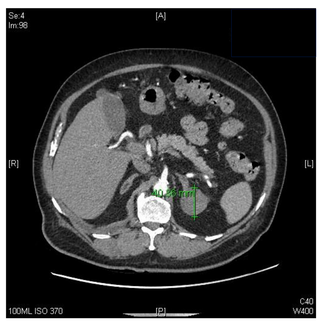


Image A: Initial, transverse view of the adrenal mass

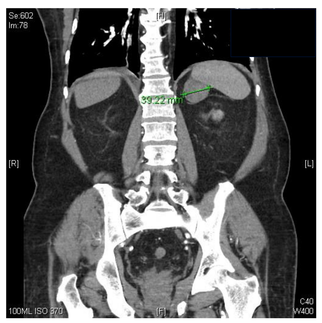


Image B: Initial, coronal view of the adrenal mass

**Initial Biochemical Work Up**

| Test | Result | Normal Range |
|---|---|---|
| Urine Total Catecholamines | <60 mcg/g | 9-74 mcg/g |
| Urine Epinephrine | <20 mcg/g | 2-16 mcg/g |
| Urine Norepinephrine | <40 mcg/g | 7-65 mcg/g |
| Urine Total Metanephrines | 505 mcg/24h | 149-603 mcg/24h |
| Urine Fractionated Metanephrines | 56 mcg/g | 21-153 mcg/g |
| Urine Dopamine | 106 mcg/g | 40-390 mcg/g |
| Serum Aldosterone | 11 ng/dL | < or = 28 ng/dL |
| Serum Renin Activity | 0.47 ng/mL/h | 0.25-5.82 ng/mL/h |

Initial work up did not include dehydroepiandrosterone sulfate testing.

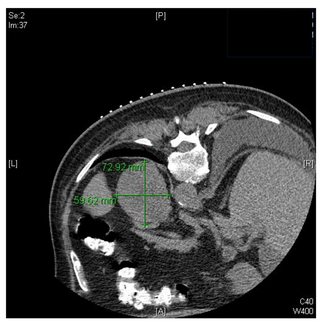


Image C: Transverse view of the adrenal mass during a prone, CT-guided adrenal biopsy

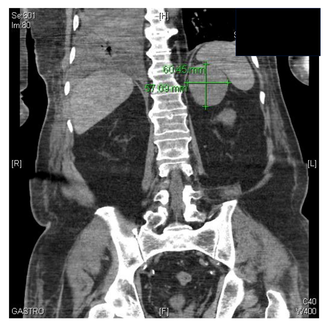


Image D: Coronal view of the adrenal mass during a CT-guided adrenal biopsy

## Discussion

Approximately 80% of patients with adrenal incidentalomas have a non-functional adenoma; less than 5% of patients with adrenal incidentalomas will have ACC[3].

Patients with adrenal incidentalomas should undergo evaluation for signs and symptoms of hypercortisolism, hyperaldosteronism, presence of pheochromocytoma, or a malignant tumor[3]. Evaluation for hypercortisolism includes the measurement of serum dehydroepiandrosterone sulfate and low-dose dexamethasone suppression testing[4]. If the patient is hypertensive, evaluation for hyperaldosteronism should include the measurement of plasma aldosterone and plasma renin[4].

For adrenal incidentalomas measuring >4 cm, open adrenalectomy is gold standard of treatment. For those <4 cm, repeat imaging should be performed at 3 and 6 months, and then annual for 1-2 years. Biochemical follow up should be performed annual for up to five years. Interval enlargement of more than 1 cm warrants consideration of resection[3].

This case illustrates the importance of longitudinal radiographic monitoring and frequent, complete biochemical evaluation of an adrenal incidentaloma. Although the patient's biochemical evaluation was unremarkable at the time of incidentaloma discovery, the rate of mass growth revealed on imaging should highlight the importance of clinical suspicion of an adrenocortical carcinoma.

## References

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