Adrenal Incidentaloma in Young – A Gray Zone?

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Adrenal Incidentaloma in Young - A Gray Zone?

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BACKGROUND

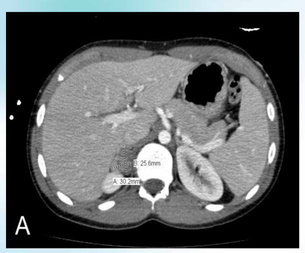
- Adrenal incidentalomas are masses 1cm or greater, identified incidentally on imaging performed for indications other than for an evaluation of adrenal disease.
- The incidence of adrenal incidentalomas ranges between 0.5-4 % and the frequency of adrenal cancers among incidentalomas is extremely small with an estimated prevalence of 4–12 per million population.[1-2]
- Adrenocortical carcinomas are extremely aggressive tumors and are mostly fatal if not identified early in the disease course.

CASE PRESENTATION

- A 24-year-old male presented with 4-month history of facial flushing, weight gain, abdominal pain, increasing abdominal girth and significant hypertension.
- Physical exam showed cushingoid facies with facial plethora, abdominal distention and associated tenderness.
- Patient noted a history of hospitalization 7-months ago for a motor vehicle accident. CT scan ruled out internal trauma but revealed a right adrenal incidentaloma measuring 3cm x 2.6cm x 3cm (Figure A). The patient was discharged and unfortunately lost to follow up.
- A CT scan done during current admission showed the right adrenal mass increased in size to 8.9cm x9.1cmx 6cm (Figure B).
- A comprehensive endocrine work up showed the mass to be functional with overall findings consistant with cushing syndrome (Table 1).
- He further underwent right adrenalectomy with pathology confirming high grade adrenocortical carcinoma with clinical stage II, T2 N0 M0.
- Despite surgery and aggressive adjuvant chemotherapy, cancer progressed rapidly in 6 months to reach a massive size (Figure C and D).
- The patient survived only 7 months after initial chemotherapy.

Table 1.

Lab	Result	Reference Values
Cortisol level	33 ug / dL	4.3 - 22.4 ug / dL
ACTH	<10 pg / mL	10 - 60 pg / mL
24 hour urine cortisol	4796 ug / 24 hours	3.5 - 45 ug / 24 hr
Plasma metanephrine	Normal	
Plasma normetanephrine	Normal	
Serum aldosterone level	Normal	



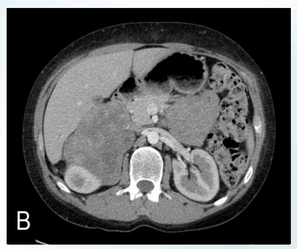
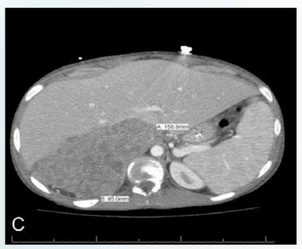


Figure A: Right adrenal incidentaloma measuring 3cm x 2.6cm x 3cm.

Figure B: Heterogeneously enhancing mass in the right adrenal fossa increased in size to 8.9cmx9.1cmx6cm.



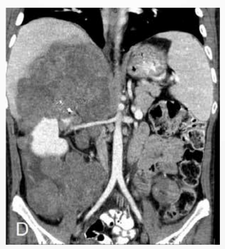


Figure C and D: Adrenal cortical carcinoma measuring 27cmx15cmx 11cm occupying the entire hemi-abdomen abutting the liver, extending into retroperitoneum, compressing the inferior vena cava and encasing the renal vein.

DISCUSSION

- The diagnosis of adrenal carcinomas can easily be overlooked in incidentalomas of a young patient population, partly due to its rarity and partly due to the subclinical nature of the disease at early stages.
- There is lack of data on the true incidence of adrenal carcinoma in incidentalomas of young.
- There is high prevalence of benign adrenal cortical adenomas in incidentalomas with increasing age; hence the finding of an adrenal mass of any size in a young patient should increase the suspicion for malignant potential of the mass.[2]
- Features suggesting malignancy include irregular or unclear margins, heterogeneity with mixed densities, attenuation (density) > 10 Hounsfield unit (HU) on unenhanced CT scan with rapidity of washout of contrast medium < 50 % at 10 minutes and hyper intense lesion in relation to liver on T2 weighted imaging on MRI.
- Though most authors suspect malignancy in masses above 4 cm and recommend resection, the validity of that threshold is questionable in a much younger population. Adrenal cancer and metastatic cancers are the most difficult to diagnose especially at an early stage as they can often mimic adenomas in appearance at smaller sizes.[6,7]
- As per AACE and AAES Guidelines, patients with adrenal incidentalomas smaller than 4 cm and radiologic characteristics consistent with a benign adenoma need to have radiographic reevaluation at 3 to 6 months and then annually for 1 to 2 years. Hormonal evaluation should be performed at the time of diagnosis and then annually for up to 5 years. AACE - American Association of Clinical Endocrinologists, AAES - American Association of Endocrine Surgeons.
- These guidelines are largely based on a patient population with a median age of 60 years (range 34–79).[10,11]
- 5% of the patient population with adrenal incidentaloma can have subclinical cushing syndrome, which would require work up with 1-mg dexamethasone suppression test or urine free cortisol in addition to other routine hormonal work up.
- In a much younger population, a lower size threshold and a prompt initial diagnostic work up with a multidisciplinary approach involving an experienced radiologist, endocrinologist, a surgeon and a pathologist is warranted.

CONCLUSION

- Smaller an adrenocortical carcinoma at the time of diagnosis, the lower the tumor stage and the better the overall prognosis.
- Adrenal carcinomas are extremely aggressive and are invariably fatal if not diagnosed early. They have tendency to attain massive size as in our patient and causes significant debilitation.
- All adrenal incidentalomas especially in young age group require an immediate and an optimal diagnostic approach to rule out malignancy or secretory function, close biochemical and radiographic follow up.
- Adrenalectomy should be a consideration in the presence of autonomous hormonal secretion or if the mass enlarges by 1 cm or more during the period of observation.[1]

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