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 1 cm 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 a 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 3 cm x 2.6 cm x 3 cm (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 8.9 cm x 9.1 cm x 6 cm (Figure B).
- A comprehensive endocrine work up showed the mass to be functional with overall findings consistent with cushing syndrome (Table 1).
- He further underwent right adrenalectomy with pathology confirming high grade adrenocortical carcinoma with clinical stage II, T2 N0 Mo.
- 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.

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.3,4
- Features suggesting malignancy include irregular or unclear margins, heterogeneity with mixed densities, attenuation (density) > 10 Hounsfield unit (HU) on unenhanced CT scan with rapidly 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.5,6
- 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.

Table 1.

<table>
<thead>
<tr>
<th>Lab</th>
<th>Result</th>
<th>Reference Values</th>
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</thead>
<tbody>
<tr>
<td>Cortisol level</td>
<td>0.2 µg/dL</td>
<td>3.6–11 µg/dL</td>
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<tr>
<td>ACTH</td>
<td>&lt;10 pg/mL</td>
<td>10–40 pg/mL</td>
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<tr>
<td>24-hour urine cortisol</td>
<td>1760 µg/24hr</td>
<td>15–45 µg/24hr</td>
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<tr>
<td>Plasma metanephrine</td>
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<td>normal</td>
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<tr>
<td>Plasma normetanephrine</td>
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<td>normal</td>
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<tr>
<td>Plasma 17-OHCS</td>
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<td>normal</td>
</tr>
<tr>
<td>Serum aldosterone level</td>
<td>normal</td>
<td>normal</td>
</tr>
</tbody>
</table>

References: