Pheochromocytoma A Salty Diagnosis

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Introduction

Pheochromocytomas are rare catecholamine-secreting tumors that arise from chromaffin tissue within the adrenal medulla and extra-adrenal sites. The most common clinical sign of pheochromocytoma is sustained or paroxysmal hypertension (90%), and the most common symptoms are headache, excessive truncal sweating, and palpitation. In some cases, the clinical symptoms are not clear. Roughly 70% of adrenal incidentalomas are non-functional. A small group of 5–7% of the functional ones (30%) may exist as pheochromocytoma. The literature indicates that incidental pheochromocytoma cases that are smaller than 1 cm have no clinical symptoms.

Case Report

We present the case of a 47 year old female with a history of uncontrolled hypertension, status post stroke with left sided hemiplegia who was referred to our hospital for evaluation of a left sided adrenal mass. In spite of being on multiple antihypertensive medications her blood pressure (BP) was ranging from 160/90 to 180/95 mmHg. Abdominal CT and MRI revealed a 4.7 x 5.4 cm mass in the left adrenal gland. Elevated 24-hour urine levels of VMA, metanephrines and catecholamines confirmed the diagnosis of pheochromocytoma. Histopathological data correlated with pheochromocytoma as well.

In addition, the patient was noted to have hypercalcemia with elevated PTH which raised the possibility of MEN 2A syndrome. A thyroid and parathyroid ultrasound was negative along with the RET protooncogene. The patient underwent explorative laparotomy with left adrenalectomy and was discharged one week later. Unfortunately, the patient was lost to follow up due to her living out of state.

Evaluation & Treatment

<table>
<thead>
<tr>
<th>Test</th>
<th>SEN (%)</th>
<th>SPEC (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ucathols</td>
<td>83%</td>
<td>88%</td>
</tr>
<tr>
<td>Utotal metanephrines</td>
<td>76%</td>
<td>94%</td>
</tr>
<tr>
<td>Ucathols+metaneph</td>
<td>90%</td>
<td>98%</td>
</tr>
<tr>
<td>Uvma</td>
<td>63%</td>
<td>94%</td>
</tr>
<tr>
<td>Plasma catecholamines</td>
<td>85%</td>
<td>80%</td>
</tr>
<tr>
<td>Plasma metanephrines</td>
<td>99%</td>
<td>89%</td>
</tr>
</tbody>
</table>

Discussion

Pheochromocytomas are found in 0.1% of patients tested for hypertension and in 4% of patients with incidental adrenal adenomas. According to the Mayo Clinic, 24 hour fractionated urine metanephrines and catecholamines are the most reliable method to diagnose pheochromocytomas. This method provides greater than 90 percent sensitivity and specificity. If one has a high clinical suspicion then fractionated plasma metanephrines should also be measured. Surgical excision remains the only cure with laparoscopic adrenalectomy being the procedure of choice. Our case is unique in that the patient was misdiagnosed with essential hypertension for many years and had end organ damage, a stroke.

In summary, clinicians should perform a thorough evaluation of hypertension in all patients, especially in patients with refractory hypertension.

References