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Case Report

A Rare Case of Coexisting Pheochromocytoma and Hyperaldosteronism Discovered as an Adrenal Incidentaloma

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Abstract

Adrenal incidentalomas are becoming a frequent finding as imaging technologies are readily available and more accurate. These lesions rarely represent hormonally active tumors. We report the case of a 53-year-old woman who presented with an adrenal incidentaloma compatible with pheochromocytoma and hyperaldesteronism. This association has only been reported in few cases in the literature. She was asymptomatic, except for mild hypertension and non-specific symptom such as sweating, headaches and palpitations. Her biochemical abnormalities resolved post-adrenalectomy.

ABBREVIATIONS

CT: Computed Tomography; MRI: Magnetic Resonance Imaging; HU: Hounsfield Unit; PET: Positron Emission Tomography

INTRODUCTION

Adrenal incidentalomas are becoming an increasingly common finding due to the widespread use of more advanced imaging technology. Fortunately, these lesions are malignant or secreting in less than 5% and 10% of cases, respectively [1]. The coexistence of pheochromocytoma and hyperaldosteronism has only been reported in few case reports of patients presenting with symptoms of either disease [2-6]. We describe a unique presentation of this association in a case of adrenal incidentaloma.

CASE PRESENTATION

A 53-year-old woman presented to the emergency room with a history of abdominal pain. A non-contrast enhanced CT scan was ordered to exclude nephrolithiasis. Although no cause of abdominal pain could be identified, adrenal incidentalomas were found: two left nodules of benign appearance, measuring 2 cm and 7 mm, and a single 3cm mass on the right gland with a density of 35 HU. She was otherwise in good health; her past medical history was relevant for mild asthma, hypothyroidism and depression. She was an active smoker and occasional drinker, but did not use any illicit drugs. She was taking thyroid hormones, calcium supplements and a selective serotonin reuptake inhibitor on a regular basis. She reported occasional non-specific symptoms such as headaches, sweating and palpitations. The clinical exam was unremarkable, with a maximum blood pressure of 150/100mm Hg without criteria for orthostatic hypotension. Her routine laboratories were normal, including a potassium level of 4.6 mmol/L.

The two left nodules were considered as adenomas because of rapid 15-minute washout on a contrast enhanced CT and homogeneous lipid-rich appearance on MRI. On the other hand, the larger right mass had delayed contrast washout time (absolute 56%, relative 36%). It was lipid-poor and mildly hyper intense on T2-weighted MRI images. This lesion was also strongly hyper metabolic on PET scan and demonstrated intense capitation on ^{131}I-metaiodobenzylguanide scintigraphy. A routine hormone-secretion panel was also ordered (Table 1). The fractionated plasmatic and 24-hour urinary metanephrine levels were repeatedly abnormal. Although selective serotonin reuptake inhibitors could have falsely elevated catecholamine levels, combined radiologic and biochemical evidence strongly supported the diagnosis of right adrenal pheochromocytoma and surgery was planned. Remarkably, morning plasma aldosterone-renin ratio was also repeatedly abnormal, suggesting concurrent subclinical hyperaldosteronism. This finding was not investigated with further confirmation tests or adrenal venous sampling.

Laparoscopic adrenalectomy was performed after

Table 1: Laboratory findings.

Test		Pre-adrenalectomy	Post-adrenalectomy	Normal
8 am cortisol, post 1 mg dexamethasone (nmol/L)		59		(<50)
Aldosterone (pmol/L)				
Supine	1445	155	(<520)	
Erect	925		(<520)	
Renin (pg/mL)				
	Supine	2.6	2.5	(<41.4)
	Erect	5		(<70.1)
Aldosterone(pmol/L)/Renin(pg/mL)		556	62	
Fractionated plasma metanephrines (pmol/L)				
	Catecholamines:			
	Epinephrine	460		(55-820)
	Norepinephrine	3290		(1180-3550)
	Metanephrines:			
	Normetanephrines	1.69	0.41	(<0.9)
	Metanephrines	2.14	0.22	(<0.5)
Fractionated 24-hour urinarymetanephrines (nmol/d)				
	Catecholamines:			
	Adrenaline	87		(<110)
	Noradrenaline	258		(<475)
	Dopamine	2267		(<2600)
	Metanephrines:			
	Normetanephrine	425		(<522)
	Methoxytyramine	357		(<350)
	Metanephrines	2202		(<266)

pharmacologic preparation with prazosin and propranolol for dual alpha-adrenergic and beta-adrenergic blockade. There were no perioperative complications and the anti-hypertensive medication was quickly tapered off. The final pathology report confirmed the diagnosis of a 4 cm pheochromocytoma. Free plasmatic metanephrine levels and plasmatic aldosterone-renin rationormalized post operatively (Table 1). These findings support the diagnosis of residual non-functioning adenomas. These lesions will be followed with imaging andhormone-secretion panel six months post-op, and according to incidentaloma guidelines subsequently.

DISCUSSION

Adrenal incidentalomas rarely represent secreting tumors. Indeed, previous reports suggested prevalence of 3.1%for pheochromocytomas, 0.6% for aldosteronomas and 6.4% for Cushing's disease.[1]The simultaneous occurrence of pheochromocytoma and primary hyperaldosteronism is highly unlikely. Yet, a small number of those cases has been reported [2-6]. Whether there is a pathophysiological relationship between these two conditions remains unclear. Pheochromocytomas secrete a large array of hormones, some of which could promote adrenocortical hyperplasia and aldosterone secretion [2]. This could be enhanced by increased contact between adrenal structures caused by architecture distortion triggered by the tumor itself [4]. Secondary hyperaldosteronism has also been reported. It has been hypothesized that catecholamines may induce renin secretion, perhaps through a direct action on renal cells or a reduction in renal perfusion pressure [2,6,7]. Nonetheless, the extremely low incidence does not favour such a pathophysiological association.

Unlike our case, most patients diagnosed with concurrent disease presented with suggestive symptoms, such as poorly controlled hypertension and hypokalemia. Adrenalectomy resulted in complete resolution of related symptoms and biochemical abnormalities in all cases [2-4,6]. Pathology revealed confirmation of both diseases in other reports. Unfortunately, pathologic confirmation of primary hyperaldosteronism was not obtained in our case, perhaps due to a small sampling from a heterogeneous adrenal gland. However, biochemical findings in our patient were strongly in favour of concurrent hyperaldosteronism. Plasma renin-aldosterone ratio was repeatedly abnormal without any interfering medication, and normalized after surgery. Optimal follow-up for these rare patients, who could be considered at higher risk, is unclear.

In conclusion, we observed the concomitant occurrence of pheochromocytoma and hyperaldosteronism, both cured by unilateral adrenalectomy. Although extremely unlikely, this suggests that multiple pathologies may coexist in an adrenal incidentaloma. As more cases are reported, one can raise the question whether this is truly an accidental finding.

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