Pheochromocytoma Presenting with Adrenal Necrosis due to Spontaneous Hemorrhage: A Case Report

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Pheochromocytomas are seldom and vascular neuroendocrine tumors-secreting catecholamines. They may be adrenal or extra-adrenal in origin. Symptoms are due to periodic catecholamine discharge into the vasculature, the mass effect of the lesion or adrenal insufficiency. There are various etiologies causing adrenal hemorrhage of which a mass lesion is one of the rarest. Adrenal hemorrhage occurring during pheochromocytoma is an extremely rare but life-threatening complication especially when it extends to the retroperitoneum. Case reports of adrenal hemorrhage associated with a mass lesion are not frequent in the literature. We report a case of adrenal hemorrhage causing necrosis of the mass which was biochemically documented to be pheochromocytoma and fully recovered after surgery.

Keywords: Adrenal, Hemorrhage, Necrosis, Pheochromocytoma

INTRODUCTION

Pheochromocytoma is a rare and catecholamine-secreting neuroendocrine tumor of the adrenal medulla.¹ It is a rare cause of secondary hypertension with a prevalence of 0.1-0.6%.² Adrenal hemorrhage may be secondary to sepsis, anticoagulation, bleeding disorders, trauma, or a result of spontaneous bleeding associated with mass.³ The literature on adrenal hemorrhage with an associated mass has been limited to small series and case reports. We present a case of pheochromocytoma in which necrosis developed following spontaneous adrenal hemorrhage and fully recovered after surgery.

CASE REPORT

A 56-year-old woman with known comorbidities of hypertension and Type 2 diabetes mellitus presented to internal medicine outpatient clinic with complaints of nausea, vomiting, headache, recurrent abdominal pain on the left lower quadrant, and weight loss of 15 kg in the last month. On detection of a hypochromic microcytic anemia and very high erythrocyte sedimentation rate (90 mm/h) she was internalized for further evaluation. Episodes of palpitation, diaphoresis, and hypertension reaching 200/110 mmHg were observed during an early hospital stay. Abdominal ultrasonography reported a suspicious mass lesion in the left renal-adrenal region and computerized tomography (CT) was recommended for exact diagnosis. Abdominal CT reported a heterogeneous left adrenal mass lesion with 8 cm × 8 cm × 9 cm of size and intrastructural portions of the high density of hemorrhage. In the arterial phase, there were areas with density equal to the artery, and these areas were increasing in diameter in progressing phases. These structures were thought to be hemorrhage relevant to pseudoaneurisms. Any possible mass lesion liable could not be differentiated among areas of hemorrhage, and magnetic resonance imaging (MRI) was recommended. MRI, when interpreted together with CT, confirmed spontaneous adrenal hemorrhage but could not rule out hemorrhage due to a mass lesion (Figures 1 and 2).

To evaluate the nature of the tumor (hormonally active or not), a series of laboratory tests were performed. Basal cortisol level at 8 am, adrenocorticotropic hormone (ACTH) level, dehydroepiandrosterone level, dexamethasone suppression test, and ACTH stimulation test were all

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detected to be normal. The analysis of 24 h – urine was as follows: Metanephrine - 18642.81 µg/24 h, normetanephrine - 6547.2 µg/24 h, dopamine - 163.88 µg/24 h, epinephrine - 561.33 µg/24 h, norepinephrine - 332.03 µg/24 h, vanilmandelic acid (VMA) - 51.13 mg/24 h, homovanillic acid (HVA) - 10.24 mg/24 h, 5-hydroxy indole acetic acid (HIAA) - 5.07 mg/24 h (Table 1). After interpreting the test results, pheochromocytoma was diagnosed. To differentiate in between a sole adrenal tumor and any other accompanying masses (paraganglioma); metaiodobenzylguanidine labeled iodine-123 scintigraphic scanning study was performed which detected no other mass excluding the left adrenal gland.

The patient was operated for the left adrenal mass with intraoperative close hemodynamic monitorization. The pathological evaluation reported macroscopically an encapsulated mass lesion resembling tumor and microscopically the mass in the adrenal gland consisted entirely of necrosis with minimal residual adrenal cortex (Figures 3 and 4).

The patient maintained normotensive and had no remaining complaints in the post-operative period. A control 24-urine study for catecholamines, metanephrine, normetanephrine, VMA, HVA, HIAA reported normal results (Table 1). The rest of the hospital course of this patient was unremarkable, and she fully recovered.

### DISCUSSION

Pheochromocytomas are rare, catecholamine-secreting, vascular, neuroendocrine tumors arising from chromaffin cells of the adrenal medulla. About 15-20% of such tumors are extra-adrenal in origin and is termed paraganglioma or extra-adrenal pheochromocytoma.1 Both adrenal and extra-
adrenal forms have similar clinical symptoms due to excess secretion of catecholamines. Pheochromocytoma should be suspected in patients exhibiting classic adrenergic spells consisting of episodic palpitations, diaphoresis, pallor, and tremors. In addition, those with resistant hypertension at a young age (<20 years), abnormal blood pressure response during anesthesia, surgery, or angiography and those within incidentally discovered adrenal masses should be evaluated as well.

Pheochromocytoma is diagnosed via a biochemical confirmation of hormonal excess, followed by anatomical localization of the catecholamine-secreting tumor. The biochemical diagnosis of catecholamine excess is established by measuring levels of catecholamines (dopamine, norepinephrine, and epinephrine) or their metabolites (normetanephrine and metanephrine, metabolites of norepinephrine and epinephrine, respectively) in the plasma and urine. The present case demonstrated high levels of especially epinephrine and norepinephrine and their metabolites.

Adrenal hemorrhage usually arises within the medulla as in our case. Various etiologies may present with adrenal hemorrhage of the most frequent being anticoagulation, bleeding disorders, and trauma. Other less frequent disease states are sepsis or spontaneous bleeding associated with mass. In the differential diagnosis of adrenal hemorrhage, it is important to consider ruptured abdominal aortic aneurysm, renal cell carcinoma, angiomylolipoma, or renal artery aneurysm. Adrenal hemorrhage is usually first diagnosed via CT or MRI. Hematoma on CT is heterogeneous and of high density. MRI has been reported to be more accurate than other imaging modalities for diagnosing adrenal hematoma, with high signal intensity on T1-weighted images. MRI also may differentiate subacute hemorrhage from chronic hemorrhage.

Presenting symptoms of adrenal hemorrhage are typically non-specific and include sudden abdominal, chest, flank, or back pain, nausea and vomiting, hypotension/shock, tachycardia, and fever. Thus, adrenal hemorrhage is a challenging diagnosis because symptoms can mimic many other abdominal pathologies. Fortunately, the hemorrhage in our patient was limited in the adrenal mass and did not extend to retroperitoneum which would inevitably cause a state of shock.

CONCLUSION

Physicians should consider adrenal hemorrhage in the differential diagnosis of any cancer patient presenting with acute abdominal pain, especially if accompanied by significant anemia, hypotension suggesting possible massive hemorrhage, or signs of adrenal insufficiency. Early recognition can reduce morbidity and in some cases may lead to life-saving interventions for these patients.

REFERENCES