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Permalink
https://escholarship.org/uc/item/72f1438m

Journal
Journal of Education and Teaching in Emergency Medicine, 4(4)

ISSN
2474-1949

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Publication Date
2019

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Peer reviewed
The Elusive Pheo: A Case Report of Pheochromocytoma in the Emergency Department

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Submitted: April 10, 2019; Accepted: August 8, 2019; Electronically Published: October 15, 2019; https://doi.org/10.21980/J8KW63

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ABSTRACT:

Pheochromocytoma is a rare catecholamine-secreting endocrine tumor that can cause symptoms of episodic hypertension, diaphoresis, anxiety, and palpitations. The catecholamine release from a pheochromocytoma can cause life-threatening hypertensive episodes (hypertensive emergency). We illustrate a case of hypertensive urgency caused by a pheochromocytoma.

A 39-year-old female presented to the emergency department (ED) for the second time in one week with a chief complaint of high blood pressure and palpitations. Her past medical history was significant for hypertension (amlodipine 2.5 mg daily). Review of systems was positive for diaphoresis, shortness of breath, chest pain, and palpitations. Her blood pressure was 178/118 mm Hg, heart rate (HR) was 167 beats/minute. She was diaphoretic, distressed, hypertensive, tachycardic, and anxious. Computed tomography (CT) of the abdomen/pelvis showed a 10-cm heterogeneous enhancing left adrenal mass. She was started on a nicardipine infusion, an arterial line was placed, and she was admitted to the medical intensive care unit (MICU).

This case serves as an example of how a very serious condition can present with subtle and common symptoms. Emergency physicians should consider a pheochromocytoma in their differential diagnosis for hypertensive emergency.
**Introduction:** A pheochromocytoma is a catecholamine-secreting endocrine tumor. Albeit rare, a pheochromocytoma can cause a patient to present to the ED with very common symptoms (eg, headache, hypertension, palpitations, diaphoresis, and anxiety), as we see in this case. Without a high-level of clinical suspicion, this diagnosis can easily be missed. An undiagnosed pheochromocytoma can lead to end-organ damage and potentially lethal hypertensive crisis and, therefore, an emergency physician should at least consider it in the list of possibilities of patients presenting with symptomatic high blood pressure.

**Presenting concerns and clinical findings:** A 39-year-old female presented to the ED with a chief complaint of high blood pressure. Her past medical history was significant for hypertension (amlodipine 2.5 mg daily). Review of systems was negative. Her blood pressure was 112/67 mm Hg, HR was 82 and she was in no acute distress. Electrocardiogram (ECG), chest X-ray (CXR), complete blood count (CBC), basic metabolic panel (BMP), urinalysis (UA), B-type natriuretic peptide (BNP) and troponin did not show any significant abnormalities. She was discharged home with primary care follow-up. Three days later, she returned to the ED with a chief complaint of high blood pressure and palpitations. This time review of systems was positive for diaphoresis, shortness of breath, chest pain, and palpitations. Her blood pressure was 178/118 mm Hg; HR was 167 beats/minute. She was diaphoretic, distressed, hypertensive, tachycardic, and anxious. While gathering her history, she states that she had recently seen an endocrinologist for possible “thyroid issues” and that, if her thyroid studies were normal, her endocrinologist may “get a scan to look for other rare causes” of her hypertension. We did not have access to her endocrinologist’s notes, but considering the history and clinical picture in this patient who was re-presenting to the ED with such impressive symptoms, we decided to order a CT abdomen/pelvis with contrast. The CT showed a 10-cm heterogeneous enhancing left adrenal mass.

**Significant findings:** ED work-up: BMP within normal limits (WNL), white blood cell (WBC) 27.4, ECG showed sinus tachycardia, nonspecific ST segment abnormalities, BNP and troponin were within normal limits, HR 146 beats/minute, CT abdomen/pelvis showed a 10-cm-heterogenous enhancing left adrenal mass (anterior-posterior view [shown in red], lateral view [shown in blue]).

**Patient Course:** After discovery of the 10-cm left adrenal mass, it was presumed that the patient had a pheochromocytoma. The patient was then started on a nicardipine infusion for blood pressure management, an arterial line was placed, and she was admitted to the MICU. Endocrinology was consulted for pre-operative blood pressure control. Inpatient labs showed a plasma free normetanephrine level of 113 nmol/L and a plasma free metanephrine level of 4.2 nmol/L. Surgical oncology later excised the mass, which was then sent to pathology. The pathology report described a 10.5 x 10.4 cm, 324-gram pheochromocytoma. The patient was discharged a few days later on amlodipine 2.5 mg daily with endocrinology and surgical oncology follow-up.

**Discussion:** As briefly mentioned in the introduction, a pheochromocytoma is a catecholamine-producing tumor of the
adrenal medulla which can cause hypertensive crisis and even death. A pheochromocytoma, although rare, can present with very common symptoms (eg, hypertension, anxiety, chest pain, diaphoresis, palpitations) which can potentially progress to serious life-threatening complications. During a hypertensive crisis, patients may develop acute pulmonary edema, confusion, headache, intracranial hemorrhages, heart failure, flushing, dysrhythmias, and other complications. Given that the presenting symptoms of a pheochromocytoma can be so common and that potentially life-threatening complications can arise, it is our belief that an emergency physician should at least consider this on their differential diagnoses for patients presenting to the ED (or re-presenting in this case) with a combination of these aforementioned symptoms. Patients with pheochromocytomas are typically diagnosed around forty years old. Classically, ten percent of pheochromocytomas are bilateral, ten percent are malignant, and ten percent are extra-adrenal. This is known as the “rule of tens.” They can be associated with a genetic syndrome (eg, Von Hippel-Lindau disease, multiple endocrine neoplasia, neurofibromatosis type 1), or they can be sporadic. Reportedly, pheochromocytomas occur in two to eight per million people per year. Diagnosis includes measuring fractionated metanephrines and catecholamines in both plasma and 24-hour urine specimens. Advanced imaging (eg, CT abdomen/pelvis with contrast) can help with localization, treatment planning, and essentially make the diagnosis from the ED, as in this case. Treatment options consist of partial or total adrenalectomy. Preoperative blood pressure management is a serious concern for patients undergoing surgery due to the risk of a massive release of catecholamines intraoperatively. Therefore, patients must undergo strict blood pressure control preoperatively with an alpha-1 adrenergic antagonist (eg, phenoxybenzamine or phentolamine). Phentolamine is the preferred medication for an acute hypertensive crisis, but sodium nitroprusside and nicardipine can also be used during an acute hypertensive crisis. In this case, the patient re-presented to the ED with an impressive clinical picture (ie, diaphoretic, tachycardic, anxious, hypertensive) and a suspicious endocrine-related history. However, she did not complain of chest pain or back pain. She did not have a family history of pheochromocytoma and the diagnosis could have easily been missed or attributed to another etiology (ie, panic attack). We were fortunate to have the endocrine-related history component in this case, but we believe an emergency physician with a high clinical suspicion would have considered a pheochromocytoma without this history detail. We hope this case will further encourage emergency physicians to expand their differential diagnosis to consider life-threatening and even rare causes of common chief complaints, including high blood pressure.

In conclusion, it is our hope that this case will encourage emergency physicians to consider a pheochromocytoma on their differential diagnosis for patients presenting with symptomatic high blood pressure or re-presenting to the ED with a combination of the aforementioned symptoms. As in this patient, there was no family history of pheochromocytoma and no complaints of back or flank pain, making misdiagnosis a high possibility, with potentially fatal consequences. Thankfully, the additional history of having an endocrinology workup in progress assisted in making the diagnosis in this case.

References: