Case Report

Cystic Pheochromocytoma with Hypertensive Crisis and Classic Triad Diagnosed Using Point-of-care Ultrasound in the Emergency Department

CME Credits

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Abstract

Pheochromocytoma is a rare catecholamine-secreting neuroendocrine tumor that commonly presents as hypertensive crisis with the classic triad of headache, sweating, and palpitations. However, it is challenging for emergency physicians to diagnose patients who initially present to the emergency department without a medical history. Here, we present the case of a patient diagnosed with cystic pheochromocytoma using point-of-care ultrasound in the emergency department.

Keywords: Cystic pheochromocytoma, Pheochromocytoma, point-of-care ultrasound

Introduction

Pheochromocytoma is a rare and potentially devastating tumor derived from chromaffin cells. Although the tumor usually presents as hypertensive crisis, with the classic triad of headache, sweating, and palpitations, it is frequently discovered incidentally with variable clinical symptoms, as it can sometimes be asymptomatic. Pheochromocytoma is usually a solid tumor of the adrenal medulla. On the other hand, cystic pheochromocytoma is also rare but more likely to be asymptomatic. Herein, we present a case of cystic pheochromocytoma in a patient with hypertension and symptoms of the classic triad, diagnosed using point-of-care ultrasound (POCUS) in the emergency department.

Case Report

A 60-year-old man without a medical history presented to the emergency department with complaints of headache, palpitations, and upper abdominal pain for 1 day. He denied fever, shortness of breath, back pain, nausea and vomiting, or trauma history. His family history was unremarkable.

On physical examination, he was noted to have obvious sweating without neurologic signs, chest tenderness, abdominal tenderness, or knocking pain. He was fully oriented and afebrile with a blood pressure of 254/100 mmHg and heart rate of 73 beats/min. Electrocardiography showed sinus rhythm without ST-segment changes. Laboratory results were normal, except for elevated blood glucose levels (498 mg/dl). POCUS was performed, which revealed a multiloculated cystic mass between the liver and the right kidney [Figures 1 and 2]. The mass was anechoic and polycystic, which was not a vascular structure under color flow [Figure 3]. It was located above the upper pole of the right kidney and was not connected to the liver and the right kidney suspected a right adrenal tumor. Subsequent computed tomography (CT) scan of the abdomen revealed a cystic tumor with the size of 9.6 cm × 8.1 cm × 7.5 cm at the right suprarenal region [Figure 4], a diagnosis of cystic pheochromocytoma was considered. After admission, 24-h measurements of urine showed elevated levels of vanillylmandelic acid (15.48 mg/day; normal range 1–7.5 mg/day) and epinephrine (109 μg/day; upper limit 27 μg/day), which are highly indicative of pheochromocytoma.

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Received: 12-10-2021 **Revised:** 23-11-2021 **Accepted:** 15-12-2021 **Available Online:** 19-05-2022

Access this article online

Quick Response Code:

Website: www.jmuonline.org

DOI: 10.4103/jmu.jmu_190_21

How to cite this article: Lee KY. Cystic pheochromocytoma with hypertensive crisis and classic triad diagnosed using point-of-care ultrasound in the emergency department. J Med Ultrasound 2023;31:60-2.

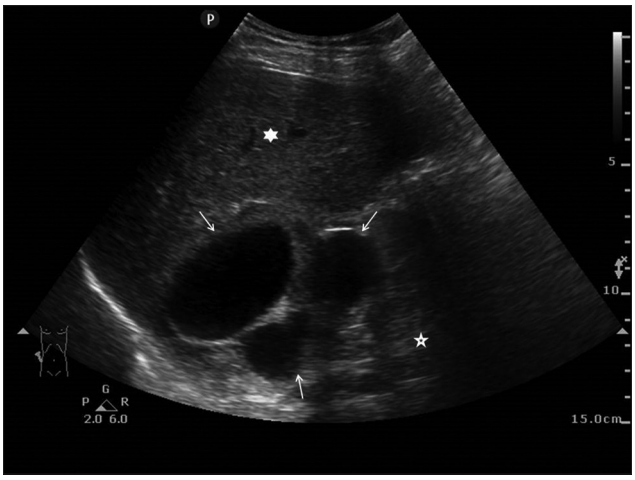

Figure 1: Point-of-care ultrasound image of the longitudinal plane showing multiloculated cystic mass (arrows), between the liver (asterixis) and the right kidney (open star), with thick wall and septations

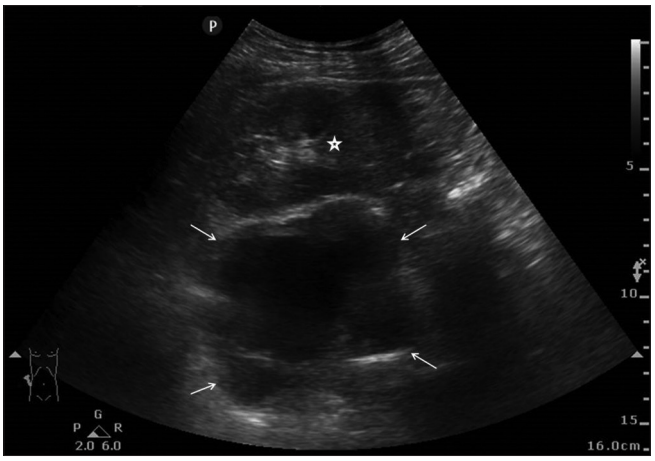

Figure 2: Point-of-care ultrasound image of the transverse plane showing multiloculated cystic mass (arrows) above the upper pole of the right kidney (open star)

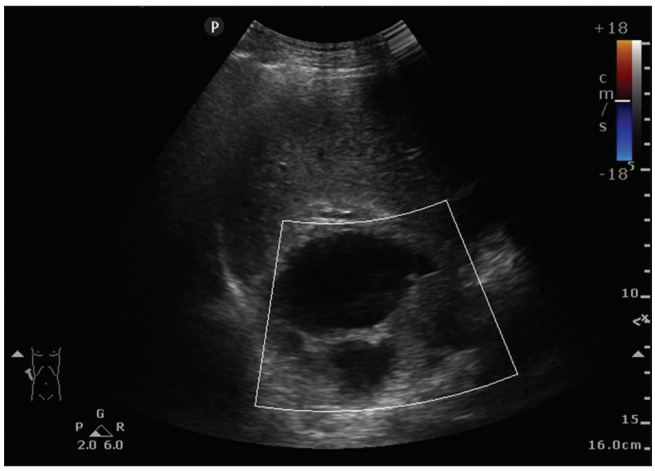

Figure 3: Point-of-care ultrasound image of color flow showing the mass without obvious vascular flow

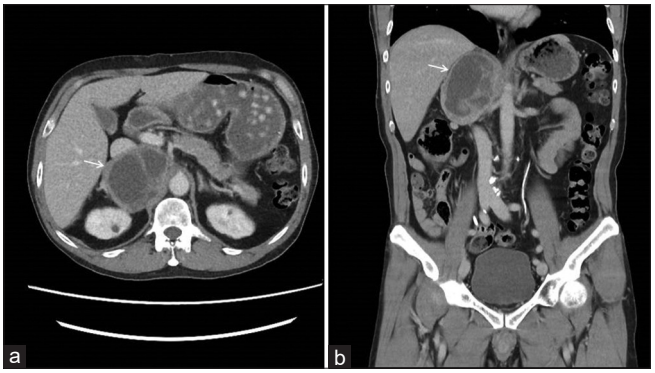

Figure 4: Abdominal computed tomography scan with intravenous contrast, axial view (a) and coronal view (b) showing a well-defined cystic tumor (arrows) with contrast enhancement below the right diaphragm

Discussion

Pheochromocytoma, first described in 1886 by Fränkel,[1] with an incidence of 0.66 cases/100,000 people/year,[2] is a relatively rare catecholamine-secreting tumor derived from the chromaffin cells of the adrenal medulla or extraadrenal paraganglia. The "rule of tens" provides the important characteristics of pheochromocytoma are10% extraadrenal, 10% are bilateral, 10% are malignant, 10% are found in asymptomatic patients, and 10% are inherited.[3]

Patients usually present with hypertensive crisis and a classic triad of headache, sweating, and palpitations. However, these symptoms are not always present and certainly not needed to make a diagnosis. Other signs and symptoms include anxiety, pallor, blurring of vision, tremulousness, nausea, flushing, weight loss, fatigue, orthostatic hypotension, and hyperglycemia. Patients presenting with hypertensive phenomenon are common in the emergency department. The incidence of pheochromocytoma is <0.5% in patients with hypertensive symptoms.[4] However, the diagnosis of pheochromocytoma can be challenging for emergency physicians because of nonspecific signs and symptoms, especially in patients with no medical history.

POCUS is useful in scanning adrenal tumors due to the advantages of including its speed, lack of radiation, real-time evaluation, and low cost. Although it is not accurate and less sensitive for smaller tumors, it still plays an important role for physicians. On ultrasound, variable appearance can be found in pheochromocytoma, ranging from solid, cystic to mixed cystic and solid.[5] Some differential diagnoses of adrenal mass should be considered, including adrenal adenoma, adrenocortical carcinoma, adrenal neuroblastoma, adrenal metastasis, and adrenal lymphoma.[6] CT scan is the most commonly used imaging in the diagnosis of pheochromocytoma. Other imaging studies include magnetic resonance imaging, metaiodobenzylguanidine, and positron emission tomography scan. In the past, CT was usually canceled because of the belief that intravenous contrast may induce hypertensive crisis in a patient who was suspected of pheochromocytoma. In a recent study, it was safe in the administration of intravenous

nonionic iodinated contrast in patients with suspected or known pheochromocytoma. In addition, the prescription of alpha-blockers before intravenous contrast CT scan was not necessary.[7]

Pheochromocytoma is usually a solid neoplasm of the adrenal medulla; the cystic form of pheochromocytoma is rare and more likely to be asymptomatic.[8] Cystic changes may be induced by hemorrhage, necrosis, and liquefaction of the tumor. We present a case of cystic pheochromocytoma in a patient who was hypertensive with complaints of the classic triad.

Surgical resection is the only curative treatment for pheochromocytoma after preoperative blood pressure control with alpha-blockers.[9] After admission, the patient was advised to undergo surgical treatment; however, the patient declined surgery and was instead regularly followed up at the outpatient clinic and prescribed antihypertensive agents.

The diagnosis of pheochromocytoma is challenging for emergency physicians. This case highlights the application of POCUS in the emergency department which helps in the early diagnosis and treatment of this disease.

Declaration of patient consent

The author certifies that he has obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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