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Case Report

Pheochromocytoma presenting with upper abdominal pain and hypertensive crisis: A case report ☆,☆☆

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ABSTRACT

Pheochromocytoma (PHEO) is a rare and complex molecularly driven endocrine disease that can present with a variety of clinical manifestations, including paroxysmal hypertension, episodic anxiety, tremors, devastating acute heart failure and acute pulmonary edema. The variety of PHEO-related symptoms increase the difficulty of identifying and diagnosing PHEO. We reported a case of a 27-year-old Chinese male was diagnosed PHEO by CT scan because of upper abdominal pain. The patient complicated with hypertensive crisis and headache following his admission, and he was underwent adrenal tumor surgical resection by carrying out appropriate clinical, laboratory and radiological imaging. Thus, the patient's PHEO-related symptoms were relieved and the blood pressure returned to normal, and discharged from the hospital with a follow-up plan. Although the PEHO-related symptoms are varied and difficult to diagnose, CT examination can be used for the preliminary detection and diagnosis. This case report emphasizes the importance of considering PHEO by CT scan and atypical symptoms, which help others better understand PHEO for early detection and timely surgical treatment to reduce catecholamine-related complications.

Introduction

Pheochromocytoma (PHEO) is a rare neuroendocrine tumor originating in the adrenal medulla with an annual incidence of 2-8 cases per million, and the average age of diagnosis is 40-50 years old, but can also occur in childhood to late adulthood [1,2]. Paroxysmal hypertension, headache, sweating and palpitations are the most common symptoms of PHEO. However, PHEO-related symptoms range from paroxysmal

Abbreviations: Pheochromocytoma, PHEO; blood pressure, BP; heart rate, HR.

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hypertension, episodic anxiety to devastating acute heart failure and acute pulmonary edema, which all increase the difficulty of identifying and diagnosing PHEO [3,4]. Excessive release of catecholamines and other neuropeptides from PHEO can lead to fatal arrhythmias or hypertensive crisis, all of these may result to sudden death [5], which highlights the importance of timely diagnosis of PHEO and effective therapeutic strategies. Here, we reported a patient who was diagnosed PHEO by abdominal CT scan because of severe upper abdominal pain.

Case report

A 27-year-old male with a negative history of any particular diseases and family presented to the emergency room with upper abdominal pain. Physical examination was unremarkable. To further confirm the diagnosis, Abdominal CT examination was performed and revealed a 60 mm × 51 mm × 60 mm mass in the left adrenal gland (Fig. 1), then the patient was admitted to urology inpatient unit. His body temperature was 36.5°C, blood pressure (BP) was 156/91 mm Hg, respiratory rate was 22 breaths/min, pulse rate was 112 beats/min, and heart rate (HR.) was 112 beats/min on admission. The patient suddenly burst into palpitations and headache with 180/130 mm Hg BP and 128 beats/min HR when communicating with us. Based on this situation, we considered that he had a hypertensive crisis, and his BP dropped to 140/86 mm Hg after about 2 hours of antihypertensive treatment with sodium nitroprusside.

On the second day of admission, the patient felt headache and palpitations again with a 220/137 mm Hg BP and 138 beats/min HR. His BP and HR gradually decreased by maintained on phentolamine 1.6 mg/h and esmolol 0.5 mg/h, and BP fluctuations in 110-130/70-90 mm Hg, heart rate 80-110 beats/min. To further diagnose, he was performed with appropriate laboratory test and CT scan, and the laboratory results were reported in Table 1. While both his plain adrenal gland CT and intravenous contrast-enhanced CT scan showed a left adrenal mass (Fig. 2). All these clinical manifestations and examinations support the diagnosis of PHEO. Considering the patient's young age and typical symptoms of PHEO, he was encouraged but failed to have genetic testing due to high cost.

Considered the patient's age, tumor diameter and typical catecholamine-related symptoms, surgical resection was performed. Before the surgery, terazosin, metoprolol and esmolol were given to control BP and HR. After adequate preoperative fluid infusion and volume expansion, left adrenal tumor resection was performed by open surgery (Fig. 3). While the patient suddenly developed hypotension of 94/41 mm Hg and heart rate of 146 beats/min when central vein was clipped during the operation, but fortunately his BP increased to 136/60 mm Hg and heart rate decreased to 102 beats/min after rapid colloid fluid infusion and norepinephrine 1.2mg/h treatment. The histopathological evaluation report confirmed the diagnosis of PHEO (Fig. 4). Finally, the patient's BP, HR, blood catecholamines remained within the normal range and clinical symptoms disappeared after the surgery, and discharged from hospital on the 10th postoperative day without further medication. Followed up for 6 months, the patient was very satisfied due to the clinically asymptomatic and stable blood pressure.

This case report was approved by the patient and Institutional Review Board.

Discussion

Pheochromocytoma (PHEO) is a rare endocrine tumor that secretes catecholamines. Clinically, it mainly manifests hypertension caused by elevated catecholamines, as well as symptoms such as headache, palpitations, sweating, etc., accompanied with cardio-cerebral and renal complications and metabolic changes [3,6]. But all atypical symptoms such as irritability, abdominal pain, anxiety, etc., are not easily recognized and easily ignored. Most patients have progressive disease associated with manifestations of catecholamine

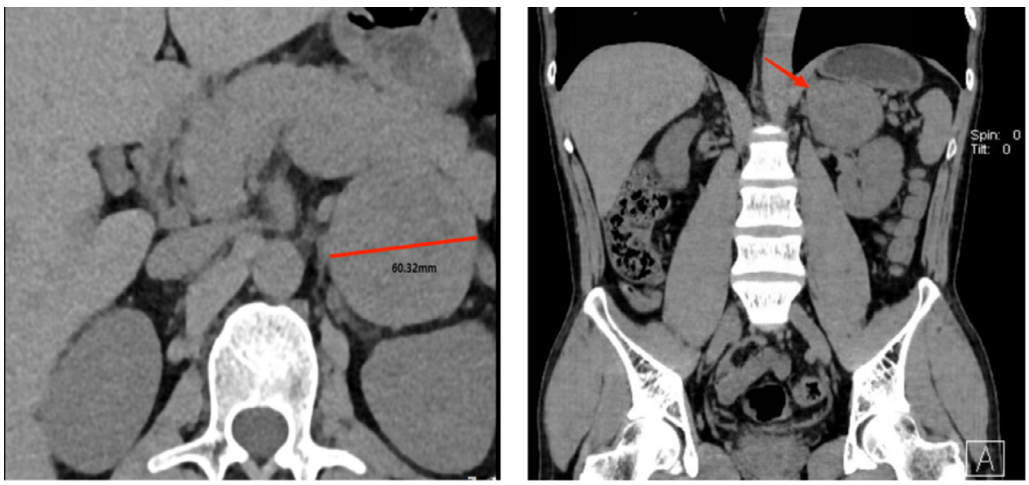

Fig. 1 – Abdominal computed tomographic (CT) scan which depicts a left adrenal gland nodule (over 6 cm diameter).

Table 1 – Laboratory tests results at admission and discharge.

Laboratory tests	Result in admission	Result before discharge	Reference value
WBC ($\times 10^9$/L)	17.93		4-10
RBC ($\times 10^{12}$/L)	5.17		4.09-5.74
PLT ($\times 10^9$/L)	297		85-303
Blood amylase (U/L)	21		25-125
Blood lipase (U/L)	15		1-60
Blood cortisol (nmol/L)			
08:00	1006.08	402.19	185-624
16:00	612.05	74.11	64-340
24:00	747	38.31	≤138
Blood catecholamines (pmol/L)			
Adrenaline	87.6	<65.2	≤605.4
Norepinephrine	17172	346.4	≤414-4435.5
Dopamine	53234	1550.3	≤195.7
Adrenocorticotropic hormone test			
8:00	7.17		AM. 7-64
8:00	2.14		PM. 1-8
12:00	<1		PM. 1-8

WBC, white blood cells count; RBC, Red blood cells count; PLT, platelet count.

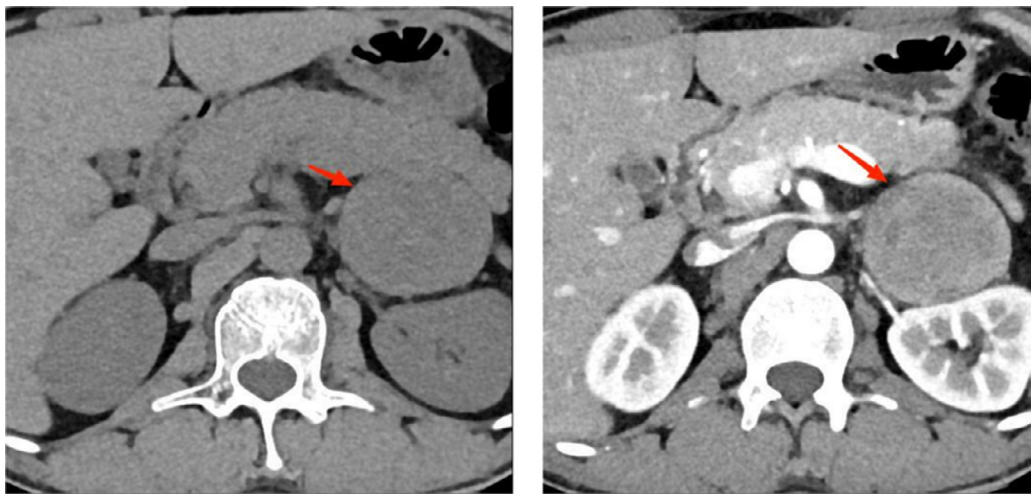

Fig. 2 – The CT scan of the adrenal nodule with intravenous contrast in the patient with a left adrenal mass.

release, and localization imaging such as abdominal CT or MRI and I^{131}-MIBG scintigraphy is required when PHEO is suspected [7,8]. About 85%-90% of catecholamine-releasing tumors occur within the adrenal glands, and about 10%-15% occur outside the adrenal glands called catecholamine-secreting paragangliomas [6]. The patient in our case had a left adrenal mass on CT scan due to acute abdominal pain, and his serum renin and aldosterone were elevated, PHEO was considered in combination with the adrenal mass. After active drug and surgical treatment, his serum renin and aldosterone returned to normal.

Pheochromocytomas larger than 5cm have a high rate of metastasis and thus a short overall survival [9]. Delayed or non-surgical resection of the primary tumor, large primary tumor, synchronous metastasis, large tumor burden, and disease progression are associated with poor prognosis. Tumor resection can greatly reduce the surge of catecholamines and improve symptoms of catecholamine excess [4]. Therefore, whenever possible, surgical resection of the primary tumor and ablation of metastases may improve clinical outcomes and prolong overall survival. Adequate preoperative preparation is required before surgical resection of PHEO with sufficient doses of α, β adrenergic blockers and volume dilation performed to prevent intraoperative cardiovascular events and hypertension risk [10]. The PHEO patient in our case was given adequate α, β adrenergic blockers, hydroxyethyl starch and crystalloid volume expansion before surgery. After surgical resection, the patient's PHEO-related symptoms disappeared and blood pressure returned to normal.

Gruber et al [4] showed that PHEOs are primarily discovered due to incidental finding on cross-sectional imaging rather than PHEO-related symptoms. The authors suggest that this is related to the advent and increasing routine use of CT imaging, as well as to the difficulty in identifying a wide range of

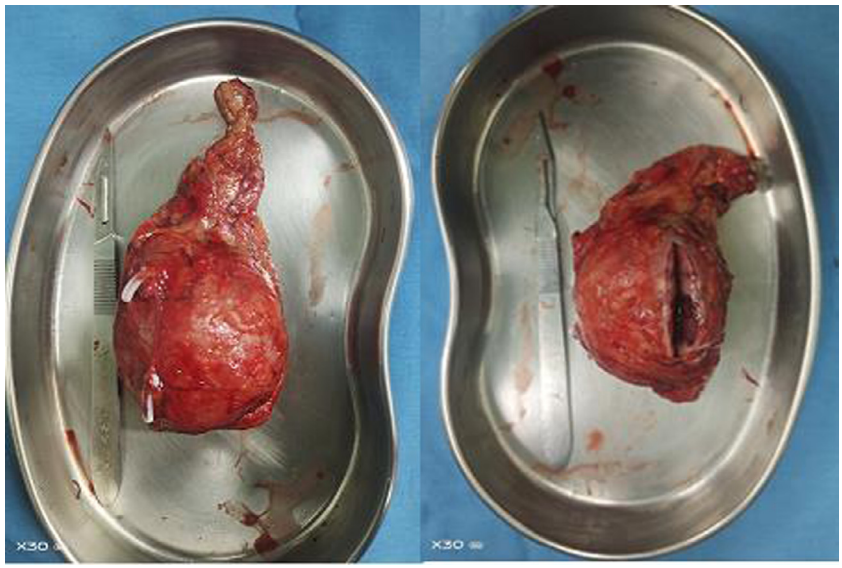

Fig. 3 – Excised left adrenal mass.

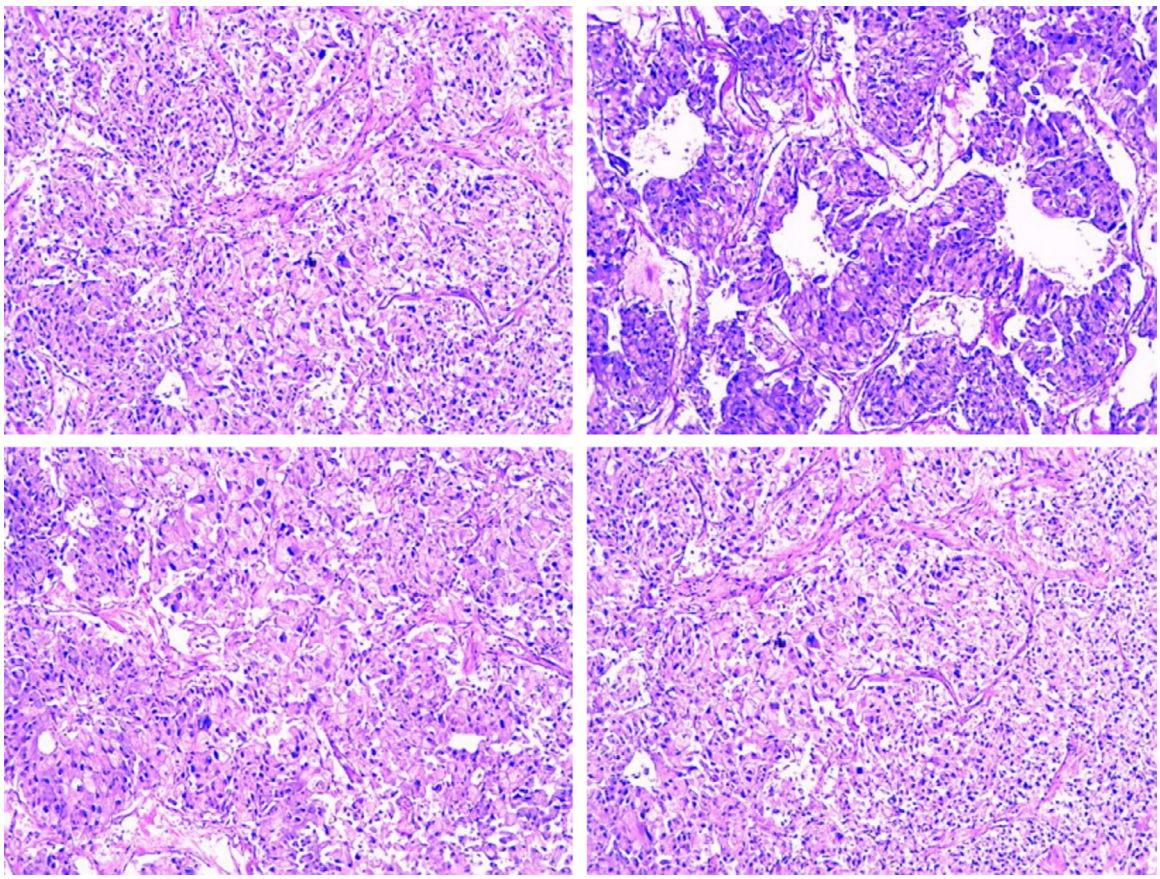

Fig. 4 – Tumor cells are diffusely distributed with focal necrosis. CgA(+), Syn(+), S-100(part +), Ki67(<5% +), MelanA(-), ainhibin(-), PAX-8(-), PAX-2(-), SF-1(-), supporting the diagnosis of pheochromocytoma.

PHEO-related catecholaminergic symptoms. Therefore, many patients may have PHEO for years before being diagnosed based on symptoms, which similar to our case, a left adrenal mass was incidentally discovered on CT scan rather than PHEO-related symptoms.

Conclusion

Pheochromocytoma is a rare and complex molecularly driven endocrine disease which is often studied retrospectively. Our case describes PHEO from patient symptoms, imaging examinations, laboratory tests, and describes the conventional treatment of PHEO through drugs and surgical treatment, which helps others to better understand PHEO for early detection and timely surgical treatment to reduce catecholamine-related complications.

Author contributions

Conceptualization: MHL, LX. Writing—original draft preparation: MHL. Writing—review and editing: MHL, LX. Supervision: CJL. Surgery: YH. The authors revised the manuscript for intellectual content. All authors read and approved the final manuscript.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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