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CASE REPORT

A life-threatening case of pheochromocytoma crisis with hemodynamic instability

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Abstract

Background: Pheochromocytoma crisis (PCC) is a fatal disease characterized by hyper and/or hypotension, hyperthermia, and encephalopathy, and its diagnosis and treatment are challenging.

Case presentation: A 50-year-old woman presented with hypertension, and computed tomography showed an adrenal tumor. Fever, shock, and impaired consciousness were observed, and PCC was diagnosed clinically. Systolic blood pressure fluctuated from 40–220 mmHg within a few minutes, and circulatory agonists were adjusted accordingly. The blood pressure changes gradually stabilized with α-blockade. Surgery was performed on hospital day 26, and the pathological diagnosis was consistent with a pheochromocytoma. She was discharged on hospital day 37.

Conclusion: Computed tomography may facilitate early diagnosis in the acute phase of PCC in case of limited patient medical information and insufficient time to wait for a definitive diagnosis using traditional hormone tests. The shock requires pharmacological therapy to maintain circulation, and paradoxically, the administration of α-blockade can be lifesaving.

KEYWORDS
adrenal gland neoplasms, consciousness, hypertension, pheochromocytoma, shock

INTRODUCTION

Pheochromocytoma crisis (PCC) is a life-threatening condition, with a reported mortality rate of 13.8%–15%.[1,2] Therefore, prompt diagnosis and therapeutic intervention are essential; however, diagnosis and treatment are not always easy during the acute phase of the disease, especially in the emergency department (ED), where limited medical information on patients are available. In this report, we describe a case of PCC in which the patient was saved, although the management of the patient's hemodynamic status was difficult.

CASE PRESENTATION

A 50-year-old woman presented to a local hospital with sudden, severe abdominal pain. At the time of presentation, she had a normal level of consciousness, blood pressure of 204/146 mmHg, heart rate of 132 beats/min, peripheral oxygen saturation (SpO_2) of 92% (oxygen mask, 6 L/min), respiratory rate of 23 breaths/min, and body temperature of 35.6°C. Her abdomen was flat, soft, and tender. Contrast-enhanced computed tomography (CT) showed a 52-mm lesion in the left adrenal gland (Figure 1A). Diltiazem was continuously administered. The hypoxemia due to pulmonary edema worsened, and endotracheal intubation was performed (Figure 1B). Subsequently, she was transferred to our hospital.

Upon arrival, she was unconscious (Glasgow Coma Scale, E2VTM5) and being administered 80 mg/h of propofol, with a blood pressure of 110/63 mmHg, heart rate of 126 beats/min at 15 mg/h of diltiazem, SpO_2 of 98% (inspiratory oxygen fraction, 0.8), and body temperature of 38.0°C. PCC and pulmonary edema were diagnosed based on the high blood pressure at presentation and CT findings, and she was

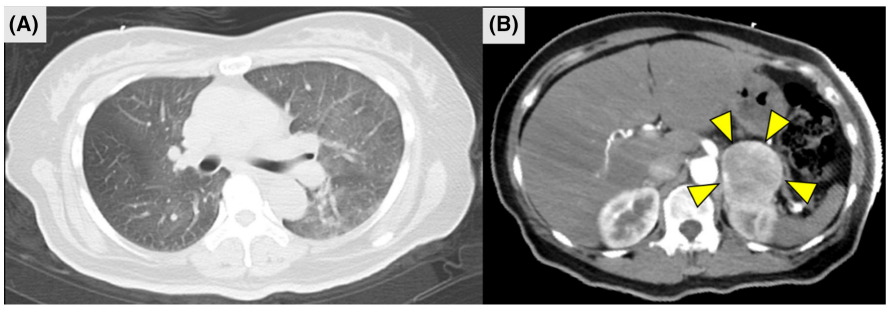

FIGURE 1 Contrast-enhanced computed tomography at the previous hospital. (A) Pulmonary edema. (B) A mass lesion of 52 mm in greatest diameter is observed, continuous with the left adrenal gland and strongly stained in the arterial predominant phase.

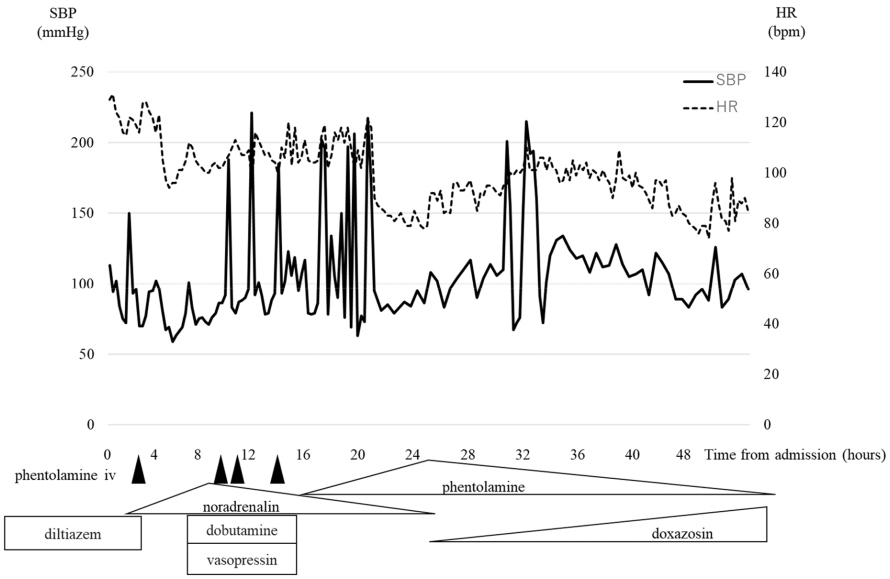

FIGURE 2 Serial changes in the patient's hemodynamic status. The systolic blood pressure fluctuated dramatically in the range of 40–220 mmHg within a few minutes, and the circulatory agonists were adjusted accordingly. Fourteen hours after admission, the combination of phentolamine and doxazosin markedly improved the hemodynamic status. HR, heart rate; iv, intravenous; SBP, systolic blood pressure.

managed with mechanical ventilation and medications for circulatory control.

Figure 2 shows the blood pressure trends and medications administered every 15 min for 48 h after admission. The systolic blood pressure decreased to 80 mmHg within the first hour of admission and further decreased to 50 mmHg after diltiazem administration. Septic shock was suspected, and a continuous noradrenaline line and antibiotics (meropenem, 3 g/day and vancomycin, 1 g/day) were initiated 2 h after admission. The hemodynamic status improved temporarily, and 2 mg of phentolamine was administered intravenously as definitive therapy for PCC. However, her blood pressure markedly decreased 6 h after admission, and cardiac contractility was also reduced. Catecholaminergic cardiomyopathy and septic cardiomyopathy were suspected. Therefore, dobutamine and vasopressin treatment were initiated. Subsequently, her systolic blood pressure fluctuated dramatically in the range of 40–220 mmHg within a few minutes, and the circulatory agonists were adjusted accordingly. She became hypertensive 14 h after admission; therefore, phentolamine was initiated at 0.5 mg/h, and the circulatory agonists were tapered and discontinued. Oral doxazosin was initiated 24 h after admission, and her hemodynamic status stabilized. Intravenous phentolamine was discontinued 80 h after admission.

On hospital day 4, the patient's blood levels of adrenaline and noradrenaline were high, at 1852 pg/mL (normal level, <100 pg/mL) and 28,303 pg/mL (normal range, 100–400 pg/mL), respectively. The sum of urinary metanephrine and normetanephrine levels on hospital day 7 was 1.24 mg/day (normal level, <1.8 mg/day). She was diagnosed clinically with pheochromocytoma, and surgery was performed on hospital day 26. The excised tumor weighed 103 g, had a maximum diameter of 53 mm, and originated from the left adrenal medulla. The pathological diagnosis was consistent with a pheochromocytoma. She was discharged on hospital day 37.

DISCUSSION

A pheochromocytoma is a catecholamine-secreting tumor with a wide range of clinical manifestations, ranging from hypertension and headache to PCC. Three cases of PCC with hyper and/or hypotension, hyperthermia, and encephalopathy were reported in 1988; the condition was defined as severe symptoms of hemodynamic instability caused by catecholamines, leading to organ damage and dysfunction. PCC occurs in 18% of patients with pheochromocytomas.[3] At presentation, 10% of PCC cases have previously been diagnosed as pheochromocytoma and 30% as hypertension.[1] Catecholamines act on α-adrenergic receptors and cause marked arterial vasoconstriction, leading to hypertension and a relatively reduced intravascular volume. This may lead to decreased end-organ perfusion and tissue ischemia, resulting in PCC.[2] Catecholamine cardiomyopathy may occur as a result of myocardial ischemia and coronary vasoconstriction and can reportedly result in shock.[4] The pathogenesis of this shock is not well understood but is thought to be related to vasodilation by β2 receptor stimulation, myocardial dysfunction, and hypovolemia.[5] Whereas pheochromocytoma presents with characteristic symptoms, such as hypertension, sweating, palpitations, and headache, PCC lacks definitive findings. PCC should be considered in cases of unexplained shock, left ventricular failure, multiorgan failure, hypertensive crisis, and unexplained lactic acidosis, all of which are associated with fever, and unstable blood pressure (alternating hypo and hypertension, possibly within seconds to minutes).[6,7] Hypotension, fever, and impaired consciousness in this case necessitated differentiation of PCC from conditions such as septic shock and thyroid crisis. Various triggers of PCC have been reported, including endotracheal intubation, anesthesia, and the administration of contrast medium.[8] In this case, the patient followed a dramatic course after transfer to our hospital, and these factors at the previous hospital might have been the precipitating factors.

Traditional hormone testing is necessary for the diagnosis of pheochromocytomas. However, during the acute phase, the clinician has no time to wait for the results, and the results are masked when catecholamines are administered to maintain the hemodynamic status. Furthermore, in the ED, medical information of patients is insufficient. It has been reported that 8% of incidental adrenal tumors detected on CT are pheochromocytomas.[9] In this case, the adrenal tumor was detected on CT, facilitating the diagnosis of pheochromocytoma. Thus, CT is useful during the acute phase of PCC and should be performed if PCC is suspected.

Although no sufficient consensus has been determined for the treatment of PCC, use of α-blockade is recommended as a first-line treatment and mechanical circulatory supportive treatments, including an intra-aortic balloon pump and use of extra-corporeal membrane oxygenation for hypotension, are suggested.[2] In this case, the patient presented with shock and fever and was managed hemodynamically for septic shock, as one of the differential diagnoses. Noradrenaline was first administered. Dobutamine was subsequently administered to address her decreased heart contractility based on the potential diagnosis of catecholaminergic cardiomyopathy or septic cardiomyopathy. In addition, vasopressin was used as a drug that does not act directly via adrenergic receptors. A similar treatment strategy was used in a previous case.[10] A strong association between the use of α-blockers and survival has been reported, although they are usually contraindicated for use in severe hypotension.[2] In this case, intravenous phentolamine and oral doxazosin were administered as specific therapies, and mechanical circulatory supportive treatments were considered. The combination of phentolamine and doxazosin markedly improved the hemodynamic status.

CONCLUSION

We describe a case of successfully treated PCC that was difficult to manage hemodynamically. PCC is associated with a high mortality rate and requires prompt diagnosis and treatment. However, differentiating PCC from a variety of other diseases is difficult in the ED, given the insufficient patient medical information. CT is useful for rapid diagnosis, and if PCC is suspected, paradoxically, α-blockers should be administered despite the shock.

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CONFLICT OF INTEREST STATEMENT

Authors declare no Conflict of Interests for this article.

DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

ETHICS STATEMENT

Approval of research protocol: N/A.

Informed consent: Informed consent for publication was obtained from the patient.

Registry and the registration no. of the study/trial: N/A.

Animal studies: N/A.

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