Pheochromocytoma Crisis Presenting as Fulminant Cardiopulmonary Failure: A Case Report

CHUN-WEN CHIU¹, CHENG-HSIUNG CHEN²

Fulminant cardiopulmonary failure in a patient with pheochromocytoma is a challenge in the emergency department and intensive care unit. We present a case in a 39-year-old woman with previously undiagnosed pheochromocytoma, who developed acute pulmonary edema after morning exercise. Her condition rapidly deteriorated into cardiac arrest. Cardiopulmonary resuscitation with extracorporeal membrane oxygenation successfully rescued the patient when an intra-aortic balloon pump failed to provide cardiopulmonary support for the pheochromocytoma crisis.

Key words: pheochromocytoma crisis, fulminant cardiopulmonary failure, cardiopulmonary resuscitation (CPR), extracorporeal membrane oxygenation (ECMO), cardiac arrest

Introduction

Pheochromocytoma is a rare neuroendocrine tumor with variable clinical manifestations. The typical presentations are headache, diaphoresis, palpitations, and hypertension. Critical cardiopulmonary complications from pheochromocytoma occur because of excess secretion of catecholamines. Fulminant cardiopulmonary failure with cardiac arrest caused by pheochromocytoma is a rare presentation with a low survival rate. A previous report showed that cardiopulmonary resuscitation (CPR) with extracorporeal membrane oxygenation (ECMO) can provide adequate cardiopulmonary support and improve the survival rate. We present a patient in pheochromocytoma crisis with cardiac arrest who was successfully rescued with ECMO.

A 39-year-old woman without a history of hypertension or heart disease presented to the emergency department with palpitations during morning exercise. On arrival, she was ambulant and had a temperature of 36.8 ℃, heart rate of 112 beats/min, respiratory rate of 22 breaths/min, and blood pressure of 120/80 mmHg and her Glasgow Coma Score was E4M6V5. Her electrocardiogram (EKG) showed sinus tachycardia. Chest radiography showed no infiltration or cardiomegalia. The cardiac enzyme values were as follows: creatine kinase 74 u/L; MB fraction 1.7 ng/mL; and troponin-I 0.08 ng/mL. The patient suddenly developed tachypnea with diaphoresis one hour later. Her heart rate increased to 156 beats/min, her respiratory rate increased to 32 breaths/min, and her blood pressure decreased to 82/42 mmHg. A subsequent
a central venous pressure of 11 mmHg, and a pulmonary artery pressure of 24/17 mmHg. She had a sudden onset of pulseless ventricular tachycardia, followed by cardiac arrest. Immediate defibrillation and standard CPR were performed. The patient received advanced cardiopulmonary life support which involved minimal interruption of chest compressions, ventilation, intravenous administration of epinephrine 1 mg × 7, monophasic shock of 360 J × 3, and intravenous administration of amiodarone 300 mg × 1 and 150 mg × 2. CPR was done for approximately 24 minutes, followed by a return of spontaneous circulation. Her unstable hemodynamic status was refractory to the IABP, as well as inotropic agents and vasopressors (dopamine 20 µg/kg/min and dobutamine 5 µg/kg/min). Echocardiography demonstrated severe, diffuse hypokinesia of the left ventricle with a left ventricular ejection fraction (LVEF) of 10%. The patient required more aggressive mechanical cardiopulmonary life support.

Veno-arterial extracorporeal membrane oxygenation (ECMO) cannulas were inserted in her right femoral vessels. A size 19 Fr cannula was implanted on the venous side, and a size 17 Fr catheter on the arterial side. Heparin was
administered and titrated to maintain an activated clotting time around 200 sec to prevent blood clotting in the ECMO circuit.

The patient’s hemodynamic status became stable 14 hours later, and the hemodynamic calculations showed a pulmonary artery pressure of 21/17 mmHg, a pulmonary wedge pressure of 8 mmHg, and a central venous pressure of 8 mmHg. Serial echocardiography revealed a gradual recovery of the heart contractility (LVEF increased from 10 to 65% in five days), and the patient’s clinical condition recovered. The ECMO and IABP were successfully weaned off on day five, and the patient was successfully extubated on day six.

The 24-hour urine catecholamine levels were elevated. (Table 1) Under high suspicion of pheochromocytoma, the patient was started on oral phenoxybenzamine 20 mg twice daily and subsequent bisoprolol 2.5 mg twice daily. After 40 days of treatment with α- and β-blockers, the patient was stable enough to tolerate surgical intervention. She underwent a laparoscopic left adrenalectomy with tumor excision. The pathology confirmed the diagnosis of pheochromocytoma. She was discharged six days postoperatively and was followed up in the outpatient department.

**Discussion**

The early diagnosis of pheochromocytoma and treatment of its life-threatening cardiopulmonary complications is a challenge in the emergency department and intensive care unit. In our case, we had a high suspicion of pheochromocytoma because of the fluctuations in the blood pressure and palpitations. Pheochromocytoma was confirmed by computed tomography and 24-hour urine catecholamine levels.

This case demonstrates fulminant cardiopulmonary failure secondary to pheochromocytoma with pulseless ventricular tachycardia and cardiac arrest. The pathogenesis of catecholamine-induced cardiomyopathy has been reported.

Excess catecholamines may cause severe, transient, but reversible cardiogenic shock.

The unusual cardiovascular complications of pheochromocytoma include sudden death, arrhythmias, acute myocardial infarction and acute pulmonary edema. Acute pulmonary edema caused by pheochromocytoma is a rare presentation with a high mortality rate. In one report, five of six patients died of pulmonary edema within 24 hours of the onset of symptoms.

These critical patients need mechanical life support. Thiagarajan et al. reported that the use of ECMO-CPR was associated with survival in 27% of adults with cardiac arrest facing imminent mortality.

Initially, our patient was implanted with an intra-aortic balloon pump, and dobutamine and dopamine infusions were administered for fulminant cardiopulmonary failure. However, her condition continued to deteriorate, and sudden onset of pulseless ventricular tachycardia developed with cardiac arrest. Obviously, the IABP, inotropic agents and vasopressors were inadequate. Cardiopulmonary resuscitation with extracorporeal membrane oxygenation and defibrillation with amiodarone was administered, and the hemodynamic status stabilized. We had to decide between emergency resection and pretreatment with an α-adrenergic blockade.

<table>
<thead>
<tr>
<th>Table 1</th>
<th>24-hour urine catecholamine levels</th>
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<tr>
<td></td>
<td>Levels</td>
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<tr>
<td>Noradrenaline (µg/day)</td>
<td>292</td>
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<tr>
<td>Adrenaline (µg/day)</td>
<td>877</td>
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<tr>
<td>Dopamine (µg/day)</td>
<td>49</td>
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<tr>
<td>VMA* (mg/day)</td>
<td>73.8</td>
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VMA: vanillyl mandelic acid
Bos et al. reported that emergency surgery for pheochromocytoma does not have to be structurally avoided and may be considered under life-threatening circumstances\(^4\).

May et al. reported that a patient who had emergency surgery for pheochromocytoma still had severe complications from massive catecholamine excess, including shock, cardiomyopathy, and adult respiratory distress syndrome\(^5\).

In this case, we decided to pretreat with \(\alpha\) blockers. Once the diagnosis of pheochromocytoma was made, \(\alpha\) blockers were administered, and when adequate \(\alpha\) blockade was achieved, \(\beta\) blockers could be initiated for preoperative management.

In conclusion, early diagnosis and treatment were necessary for this patient. Fulminant cardiopulmonary failure without response to conventional treatment may cause a high mortality if there is no aggressive mechanical cardiopulmonary life support system. ECMO successfully provided cardiopulmonary life support until recovery from a critical status secondary to a pheochromocytoma crisis. This was successful CPR with ECMO in a patient with a pheochromocytoma crisis.

References

以猛爆型心肺衰竭呈现的嗜鉻细胞瘤危機：
一病例報告

邱俊文¹ 陳正雄²

嗜鉻细胞瘤病患呈现猛爆型心肺衰竭在急診及加護病房的處置上相當具挑戰性。我們報告一個39歳
女性在之前未診斷嗜鉻细胞瘤，她在運動之後，呈現急性肺水腫然後急速演變成心臟停止。因為傳統的
心肺復甦術以及主動脈內氣球幫浦不足以提供心肺支持需要，故使用葉克膜進行復甦救回病人。

關鍵詞：嗜鉻细胞瘤危機，猛爆型心肺衰竭，心肺復甦術，葉克膜，心臟停止