Pheochromocytoma-Induced Cardiogenic Shock Rescued by Percutaneous Cardiopulmonary Bypass System

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Pheochromocytoma is a rare cause of cardiogenic shock. Clinical management is directed at reducing the heart rate while maintaining blood pressure. However, medical treatment is often unsuccessful because of the high endogenous catecholamine level and low cardiac output. Percutaneous cardiopulmonary bypass system is a circulatory device that allows for safer use of heart-rate-reducing agents, which is advantageous when used early in the course of the cardiogenic shock induced by pheochromocytoma. (Circ J 2009; 73: 1753–1755)

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Pheochromocytoma is a rare cause of cardiogenic shock encountered in emergency departments. The diagnosis is often delayed or not made until post-mortem. Even if a proper diagnosis has been made, the management of cardiogenic shock caused by pheochromocytoma is frequently unsuccessful because reducing endogenous catecholamine levels and maintaining arterial blood pressure are difficult or sometimes impossible. Although there are reports in which intraaortic balloon pumping (IABP) was used successfully for the treatment of medically unresponsive patients, not all are successful.

Percutaneous cardiopulmonary bypass system (PCPS) may help overcome this devastating clinical condition. PCPS is a more powerful circulatory device than IABP and can be used in patients with cardiogenic shock or arrest. The assembly and manipulation of the device is simplified, so rapid setup is possible. Using the Seldinger technique the cannulas for PCPS can be introduced percutaneously through the femoral artery and vein. We present a case in which pheochromocytoma induced cardiogenic shock was successfully managed with PCPS.

Case Report

A 41-year-old man was admitted to the emergency department because of palpitation and abdominal pain. He had a history of intermittent headache and palpitation but had never undergone a medical examination. His initial blood pressure was 140/100 mmHg and heart rate was 171 beats/min. The admission electrocardiogram (ECG) showed narrow-QRS tachycardia with elevated ST-segment in leads V5, V6, II, III and aVF (Figure 1A). Vagal maneuvers failed to terminate the tachycardia and intravenous administration of adenosine (6 mg) and verapamil (5 mg) had no effect. The troponin I and CK-MB levels were 6.27 ng/ml (normal range ≤0.05) and 26 ng/ml (normal range ≤3.6 ng/ml), respectively. Because of the elevated cardiac enzymes and ischemic findings on ECG, the initial impression was acute myocardial infarction and emergency PCI was tried. However, coronary angiography showed no coronary artery abnormality. To determine the cause of the abdominal pain, an abdominal CT scan was done, which revealed a hypervascular left adrenal mass (Figure 2). Meanwhile, his blood pressure decreased to 80/40 mmHg and the patient was transferred to the intensive care unit for mechanical ventilation. Echocardiography showed global hypokinesia with an ejection fraction of 20%.

An IABP was inserted through the right common femoral artery and circulation was assisted with 1:2 counterpulsation. His heart rate was continuously high (168 high/min) and blood pressure dropped further to 53/41 mmHg despite the infusion of inotropics. PaO2 (from the right radial artery) was 51 mmHg with FiO2 100% and peak end-expiratory pressure of 5 cm H2O. Pulmonary edema was present on his chest X-ray (Figure 3). We decided to use a PCPS (EBS, Terumo, Tokyo, Japan). After obtaining informed consent from the patient’s family, the left common femoral artery and vein were cannulated with catheters (17 and 22Fr, DLP, Medtronic Inc, Minneapolis, MN, USA) to provide extracorporeal circulation. The IABP was maintained to facilitate coronary blood flow. After initiation of the PCPS with a flow rate of 3 L/min, the PaO2 in the right radial artery increased to 255 mmHg and the mean arterial blood pressure from 45 mmHg to 63 mmHg. A β-blocker (esmolol 30 μg·kg⁻¹·min⁻¹) was administered to control heart rate and furosemide was used to attenuate the pulmonary edema. A β-blocker propranolol (30 mg/day) and α-blocking agent (doxazosin 1 mg/day) were given by nasogastric tube. The flow of the PCPS was gradually reduced to 0.5 L/min while maintaining the mean arterial blood pressure above 60 mmHg. The PCPS was removed after 26 h of circulatory assist. Several hours later, the IABP was removed because the patient was hemodynamically stable without any inotropic support. The
The patient was extubated on the next day. Follow-up echocardiography showed improved cardiac function with an ejection fraction of 60%, but akinesia of the apex and apical septum was observed. The urinary excretion of vanillylmandelic acid and metanephrine was found to be elevated to 19.2 mg/day (normal range <7 mg/day) and to 13.9 mg/day (normal range <1.3 mg/day), respectively. After withdrawing the mechanical support, a sacral sore was discovered and he developed a fever, which were treated with antibiotics and daily dressing. One month after the mechanical support had ceased the left adrenal mass was removed. The pathologic findings were compatible with those of a pheochromocytoma. Postoperative levels of vanillylmandelic acid and metanephrine were within normal values. Propranolol and doxazosin were stopped postoperatively. The residual sacral sore was managed conservatively. Otherwise, the patient recovered uneventfully. Echocardiography performed when he was discharged revealed improved left ventricular wall motion and the 12-lead ECG showed inverted T-waves on precordial leads (Figure 1B). The patient was discharged on

**Figure 1.** Twelve-lead electrocardiograms (ECGs). Narrow-QRS tachycardia with ST-segment elevation is present on admission (A). The ECG at discharge shows T-wave inversion in the precordial leads (B). No ischemic findings can be seen in the 12-lead ECG taken 7 months after discharge (C).

**Figure 2.** Preoperative abdominal CT scan showing the hypervascular left adrenal mass (white arrow).
the 35th day after admission. Seven months later, echocardiography and ECG showed complete recovery of cardiac function and rhythm (Figure 1C).

Discussion

The mechanism of cardiogenic shock in patients with pheochromocytoma is not fully understood, but some of the suggested mechanisms include coronary vasospasm caused by α1 stimulation, increased myocardial oxygen demand secondary to the tachycardia and high afterload, and direct toxic effect of the increased level of catecholamines.5-6 Regardless of the mechanism, there seems to be a certain degree of myocardial ischemia in most cases. Patients are often suspected to have coronary artery disease and undergo coronary artery angiography, which demonstrates no coronary lesion, as is in the present case.5-7

The mainstay of the clinical management is, therefore, to restore myocardial oxygen supply and reduce oxygen demand. However, once acute heart failure has developed, restoration of cardiac function is difficult. Using α-antagonists to reverse coronary vasoconstriction or β-blockers for tachycardia is not an optimal choice when the blood pressure is too low, nor is the administration of cardiotonic drugs helpful in these patients, who already have surplus endogenous catecholamines. IABP has been used in the treatment of medically unresponsive patients,3 but even with IABP support, some fail to recover.4

One alternative solution is PCPS. To our knowledge, there have been only 3 reports of using PCPS to manage the cardiogenic shock induced by pheochromocytoma.7-9 Although it has not been used frequently, the PCPS has potential advantages in the management of catecholamine-induced cardiogenic shock. First, it can restore arterial blood flow regardless of the heart rate or arrhythmia. Second, a β- or α-blocker can be used under more stable hemodynamic conditions. Third, arterial oxygenation can help end-organ oxygenation, even if there is severe pulmonary edema because of the cardiogenic shock. Lastly, intravascular catecholamines might be diluted by the priming solution of the device. PCPS has the disadvantages of increasing afterload because of its continuous blood flow.10 Furthermore, end-organ perfusion in continuous flow is decreased compared with pulsatile flow because it has less hemodynamic energy.11 These problems can be overcome by concomitant use of IABP, adding pulsatility to the blood flow.12

There are 2 forms of cardiogenic shock caused by pheochromocytoma.13,14 One is catecholamine-induced diluted cardiomyopathy and patients with this type of cardiac dysfunction require a long period of recovery after surviving the acute cardiogenic shock and surgical resection of the tumor. The other form is myocardial stunning caused by myocardial ischemia. Elimination of the ischemia restores myocardial function in a relatively short period of time. In the present case, using PCPS early in the course of the heart failure, and IABP in combination with medical therapy, ensured that myocardial function recovered rapidly.

Because the myocardial dysfunction in pheochromocytoma-induced cardiogenic shock is reversible, the diagnosis must be prompt and treatment initiated before extensive irreversible myocardial damage takes place. Considering the high mortality rate and poor outcomes with traditional management, the early use of PCPS should be considered in the management of cardiogenic shock induced by pheochromocytoma.

References