



Life-Threatening Pheochromocytoma Crisis Operated On Emergency Under Extracorporeal Life Support: About 4 Case Reports

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Abstract

Pheochromocytoma crisis may induce severe circulatory collapse, and mechanical support requirement as a bridge to recovery before pheochromocytoma surgical withdrawal. Here we report 4 cases of patients who suffered from an fulminant pheochromocytoma crisis with a severe cardiogenic shock. The pheochromocytoma surgical removal was successfully performed on emergency under Veino-Arterial Extracorporeal Membrane Oxygenation (VA ECMO). The definitive treatment for pheochromocytoma is surgical excision of the tumor but often delayed to allow preoperative pharmacologic preparation. In case of life threatening condition, VA ECMO support may allow surgery to be performed without delay. Though bypassing catecholamine blockade, this approach may reduce the risk of patient exposure to prolonged cardiotoxic effect of catecholamines and may favor shorter and better cardiac recovery.

Introduction

Presentations of pheochromocytoma as circulatory collapse are increasingly reported [1-3]. The circulatory collapse may require a mechanical support which was used as a bridge to recovery before pheochromocytoma surgical withdrawal [1,3,4]. Few cases have reported surgical tumorectomy under veino-arterial Extra Corporeal Membrane Oxygenation (ECMO), where ECMO was used as a protective condition [4,5]. Here we report 4 cases of patients who underwent pheochromocytoma surgical removal under ECMO because of an extreme critical condition, where tumor removal appeared as the best option to save the patient, although bypassing medical preparation.

Case Studies

Case 1

A 49-year-old man suddenly complained of palpitations, precordialgia, and emesis. He was referred to the cardiac intensive care unit with the hypothesis of myocardial ischemia as ECG showed sinus tachycardia and ST depression in posterior leads.

The clinical presentation was acute pulmonary oedema and marked swings in arterial Blood Pressure (BP). Tropinin IC level was elevated to 6.6 µg/L (TnIC, normal <0.04) and transthoracic echo showed severe Left Ventricle (LV) global hypokinesia (ejection fraction, EF <15%). Abdominal palpation found a painful mass in the right hypochondrium. A thoraco-abdominal tom densitometry revealed a retroperitoneal tumor, with inferior vena cava compression, which was highly suspected of being a pheochromocytoma of the right adrenal gland.

Hypotension and low cardiac output was associated with lactic acidosis (plasma lactate 4.5 mmol/L) and was treated with dobutamine infusion but ventricle arrhythmias occurred and worsened into a ventricular fibrillation 24 hrs after admission. Sinus rhythm was restored with a cardio version but the patient needed sedation, intubation and respiratory support.

Due to the persistent BP instability and worsening in LV EF (10%) despite epinephrine infusion, ECMO was inserted through right femoral vein and artery. ECMO provided adequate flow (4 to 5 l/min) and allowed epinephrine weaning within few hours.

Tumor removal was performed on the same day through a median laparotomy. Several episodes of ventricle arrhythmias occurred with hypertensive crises during tumor manipulation which were treated with nicardipine and esmolol infusion, and 3 electrical cardio versions. Tumor ablation was followed by neither hypotension that was controlled by nor epinephrine (1.25 mg/h). Amiodarone

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Received Date: 19 Dec 2018

Accepted Date: 07 Jan 2019

Published Date: 10 Jan 2019

Citation:

Colson P, Gaudard P, Coves R, Zeroual N, Eliet J. Life-Threatening Pheochromocytoma Crisis Operated On Emergency Under Extracorporeal Life Support: About 4 Case Reports. Clin Surg. 2019; 4: 2293.

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was administered because of recurring ventricular tachycardia. Anatomopathology was in favor of a pheochromocytoma (10 cm × 9 cm × 5 cm, 163 g) not at the expense of the right adrenal gland.

The day after surgery, the patient woke up but was re-sedated because of recurring arrhythmias and ECMO needs of a high level PEEP controlled ventilation (10 cm H₂O). Transthoracic echo showed global akinesia (EF <5%), without LV dilatation or intracavity sludge. Diuresis was spontaneous and abundant.

Five days after surgery, a trans-oesophageal echo showed LV EF at 20% which improved to 30% while reducing ECMO flow by half. ECMO was withdrawn the day after; small doses of epinephrine and dobutamine were required for few hours. LV function improved further and EF reached 40% to 50% 2 days after ECMO explantation. Plasma TnIc concentration peaked at 84.62 µg/L the day after surgery decreased slowly without complete normalization (0.23 µg/L on day 14). As a high level of brain natriuretic peptide was also observed (2000 ng/ml, normal <100), a coronary angiography was performed that excluded obstructive coronary arteries. Follow-up was then marked by pulmonary infection but full recovery allowed hospital discharge on day 39.

Case 2

A 49-year-old woman was scheduled for an arthrolysis of the right foot performed under general anesthesia. During and immediately following the surgery, she presented hypertension and pulmonary edema. She was breathless so that she was sedated and assisted ventilation was resumed just after reintubation. On EKG, a new left bundle block had appeared. A quick echocardiography showed a severe left ventricle failure, with global hypokinesia. The cardiogenic shock was associated with a metabolic acidosis (plasma lactate 9.5 mmol/L), liver cytolysis, and bad perfusion of her extremities.

She was referred to the coronary unit of the tertiary hospital with the hypothesis of myocardial ischemia. The coronary angiography showed no abnormality. The patient was transferred to the intensive care unit where she experienced a cardiac arrest just after her ICU admission. A sinus rhythm was resumed with resuscitation maneuvers, but the cardiac function was severely impaired (ejection fraction, EF <10%). An ECMO was implanted to restore perfusion; plasma lactate peaked at 20.8 mmol/L, and persisted despite ECMO and dialysis (6.9 mmol/L).

A thoraco-abdominal tom densitometry was performed the day after admission and revealed a retroperitoneal surrenal tumor, which was highly suspected of being a pheochromocytoma of the right adrenal gland. Tumor removal was performed on the same day through a median laparotomy.

Anatomopathology confirmed a pheochromocytoma (8 cm; 124 g) at the expense of the right adrenal gland. Two days after surgery, a trans-oesophageal echo showed a good recovery of LV EF at 40% allowing ECMO explantation on day 4 after implantation. Unfortunately, she experienced a septic shock in relation to a mesenteric ischemia, which required surgery (left hemicolectomy) on day 5. Follow-up showed slow improvement due to cerebral ischemic lesions, but full recovery of cardiac function. Hospital discharge was effective on day 185.

Case 3

A 40-year-old woman was addressed at the emergency department for headache, dyspnea, and hyperthermia (38°C). Her

condition worsened quickly, with hemodynamic shock within one day, with low LV EF (5% to 10%) and tachycardia. She was sedated, intubated, while receiving inotrope (dobutamine) and vasopressor (noradrenaline) to restore blood pressure. She was transferred in the cardiac assistance unit where cardiogenic shock was confirmed (plasma lactate 5.1 mmol/l) despite the inotropic support.

A VAECCMO was quickly inserted. Circulatory assistance allowed removal of inotropic and vasopressive drugs. Hypertensive crises occurred incidentally. Initial symptoms and hypertensive crises under ECMO makes the diagnosis of pheochromocytoma crisis likely, that was confirmed by thoraco-abdominal tom densitometry, showing a tumor at the expense of the right surrenal gland.

It was decided after consensus discussion between surgeon, intensivist and anesthetist to proceed to the surgical tumor resection under ECMO. The surgery was performed on day 3 after admission. Besides tachycardia controlled by beta-blocker (esmolol) the surgery was uneventful (tumor 48 g; 6.5 cm × 6 cm × 4 cm). ECMO was removed on day 4 without hemodynamic consequences. Her follow-up was complicated by pulmonary infection acquired under assisted ventilation which evolved favorably in few days. Hospital discharge was possible on day 30.

Case 4

A 46-year-old man suddenly complained of palpitations just following a physical effort. At emergency admission, he was hypertensive (200/130 mmHg), with tachycardia (150) and chest pain. A thoraco-abdominal tom densitometry excluded aortic dissection but revealed a heterogeneous tumor at the expense of the right surrenal gland. The hemodynamic check-up found out a low LF EF (20%) with a typical aspect of inverted Takotsubo. His condition worsened quickly, despite hemodynamic support with dobutamine, nor-adrenaline, sedation and assisted ventilation after intubation. The cardiogenic shock was severe (plasma lactate 11.7 mmol/l). A cardiac arrest occurred after a sustained ventricular tachycardia, but was resumed with cardiac resuscitation. An ECMO was then inserted by the mobile ECMO Unit, before the patient was transferred to the cardiac intensive care unit. Hemodynamic remained unstable under ECMO support, with alternance of hypertension and hypotension treated respectively by nor-adrenaline and fluid infusion.

It was decided to remove the tumor surgically without delay. The surgery was performed on day 3 under ECMO support, and was uneventful (tumor 46g; 7.5 cm × 5 cm × 4 cm). ECMO was weaned with complication on day 5. Follow-up was marked by a complete recovery of LV function hypertension and tachycardia which were treated by oral nicardipine and carvedilol. Hospital discharge was possible on day 21.

Discussion

Presentation of pheochromocytoma as circulatory collapse is not usual but not uncommon [1-7]. In some cases, the clinical presentation is misleading into erroneous diagnosis of myocarditis or myocardial infarction [1]. In the present cases, besides the first one where the abdominal mass gave a direct lead to the diagnosis, other cases showed various clinical presentations. Fortunately, CT scan known has a good sensitivity for detecting adrenal pheochromocytoma [7].

Pheochromocytoma is a curable cause of induced cardiomyopathy. In previous published cases, the surgical tumor removal was often postponed after ECMO explantation, mainly

because pheochromocytoma was diagnosed after cardiac recovery and ECMO explantation [1,3]. The sustained cardiogenic shock does not allow pharmacologic blockade of catecholamine synthesis or effects before surgery as usually recommended in preparation of elective pheochromocytoma removal surgery [7]. In only one case report, surgery had been performed under ECMO, not in emergency but because of ongoing hemodynamic instability despite circulatory assistance and calcium-channel blocker, α - β -blocker, and vasodilator therapy [2]. Besides, surgery performed after prolonged exposure to catecholamines may reveal the need of subsequent prolonged left ventricle assistance [2].

Conversely, the present cases show that it is possible to remove pheochromocytoma tumor under ECMO without delay. A strategy that may reduce the risk of patient exposure to prolonged cardiotoxic effect of catecholamines, which is known to lead to focal myocardial necrosis with possibly dramatic ending [5]. The consistent myocardial damage as assessed by high levels of troponin release is indeed the potential result of myocardial perfusion catecholamine-induced jeopardizing [7]. The definitive treatment for pheochromocytoma is surgical excision of the tumor, and the surgery performed on emergency under ECMO may have favored cardiac recovery and a happy ending.

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