Case report - Assisted circulation
Cardiogenic shock due to pheochromocytoma rescued by extracorporeal membrane oxygenation

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Abstract

This is the case of a 49-year-old female presenting in sustained cardiogenic shock due to an adrenal pheochromocytoma. She was rescued by venaarterial extracorporeal membrane oxygenation. The presence of a catecholamine-secreting tumor was confirmed by highly elevated plasma metanephrines and catecholamines. Successful open adrenalectomy was performed under protective extracorporeal life support and full anticoagulation early after cardiogenic shock. The patient could be weaned off mechanical support rapidly and made a full cardiopulmonary recovery.

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1. Case report

A previously healthy 49-year-old female with chest pain after exercise was admitted to the emergency department of a local hospital. She reported to have had short-lasting palpitations once a month and headache with occasional nausea. At presentation, her vital signs were normal. Electrocardiography showed self-limiting ventricular tachycardia and bigeminus, but no other abnormalities. Creatine kinase was 351 U/l (normal <195), troponin-I level 3.7 ng/ml [1.0 μg/l] (<0.1). B-type natriuretic peptide was 10 pg/ml [1.0 ng/l] (<100). Assuming that the patient suffered from acute non-ST-elevation myocardial infarction, aspirin, heparin, clopidogrel and metoprolol were administered. Later, the patient developed supraventricular tachycardia (150 beats/min), diastolic hypertension (135/110 mmHg) and severe hyperglycemia (33 mmol/l). Four hours after admission, she had to be intubated and ventilated due to acute pulmonary edema and lactic acidosis (pH 6.97, lactate 100). Blood pressure stabilized gradually under ECMO support and the clinical condition markedly improved until day 4. Norepinephrine, epinephrine and dobutamine were all stopped by then. To lower blood pressure, escalating doses of phentolamine were needed despite large doses of vaspressors and inotropic drugs (norepinephrine 1.7 μg/kg/min, epinephrine 1.0 μg/kg/min, and dobutamine 13 μg/kg/min). After end-to-side grafting of the right subclavian artery (Fem-Flex II 18 Fr, Edwards Lifesciences, Irvine, CA, USA) and percutaneous cannulation of the right femoral vein, ECMO was started with a blood flow of 5 l/min (Rotaflow pump, membrane oxygenator Quadrox, Maquet Cardiopulmonary AG, Hrllingen, Germany). In addition to mechanical ventilation and circulatory support, continuous venovenous hemofiltration was started because of anuric renal failure. At that time, fulminant myocarditis was assumed. However, in the face of potentially severe complications, endomyocardial biopsy was not performed. During sonography for quantification of pleural effusion, a solid mass of 5.5 cm in the right adrenal gland was incidentally detected. Plasma epinephrine and norepinephrine levels were measured by high-performance liquid chromatography and found to be strongly elevated (Table 1). Blood pressure stabilized gradually under ECMO support and the clinical condition markedly improved until day 4. Norepinephrine, epinephrine and dobutamine were all stopped by then. To lower blood pressure, escalating doses of phenolamine were needed and subsequently combined with esmolol. To be sure that plasma metanephrine elevation most probably derived from a real secreting tumor rather than from previous catecholamine administration, catecholamines were measured again on day 7. All fractionated plasma hormones remained excessively high (Table 1). Abdominal computed tomography confirmed the presence of a heterogeneous, contrast-
Enhanced adrenal mass measuring 5.3 × 4.0 × 5.8 cm (Fig. 1).

An open right adrenalectomy was performed with protective ECMO in place under full anticoagulation with unfractionated heparin on day 8. After ligation of adrenal vessels, phenolamine and esmolol could be both tapered rapidly. The ECMO was weaned off immediately. The patient’s cardiac function remained stable postoperatively with improved left ventricular ejection fraction of 50%. No antihypertensive therapy was needed thereafter. Complete resection of a 6-cm adrenal pheochromocytoma was histologically demonstrated. The patient made a complete cardiopulmonary recovery and was successfully extubated on day 17. After discharge, she still needed intermittent outpatient hemodialysis for chronic renal failure.

2. Discussion

Catecholamine-secreting tumors may rarely cause an acute adrenergic cardiomyopathy at initial presentation [1], although classic symptoms consist of episodic headache, sweating, tachycardia, and paroxysmal hypertension. In patients with suspected pheochromocytoma fractionated metanephrines and catecholamines in a 24-hour urinary specimen or occasionally fractionated plasma free metanephrines should be measured [2, 3]. Critical illness causes increased levels of endogenous catecholamines and their metabolites [4]. Administration of norepinephrine and epinephrine also elevates plasma levels substantially leading to false-positive test results. To establish the presence of a secreting tumor measurement of plasma free metanephrines was repeated three days after stopping all catecholamines. At that time, analytical interference with administered exogenous catecholamines, which all have extremely short plasma half-lives, was therefore highly unlikely.

Mechanical life support with ECMO is a valuable option for the treatment of refractory cardiogenic shock [5], especially when the heart failure is likely to be reversible. Several cases of patients with hemodynamic collapse secondary to pheochromocytoma rescued by ECMO have been reported [6–8]. In these patients, ECMO support was stopped before tumor resection. a-Adrenergic blockade to control blood pressure should precede surgery, because pheochromocytoma crisis may be precipitated during induction of anesthesia or surgery [9]. In the present case, early tumor resection was performed under full anticoagulation by the open transabdominal approach leaving ECMO in place in case of circulatory re-collapse after recent catecholamine cardiomyopathy. ECMO was discontinued immediately after uneventful adrenalectomy.

In conclusion, patients with profound cardiogenic shock secondary to adrenal pheochromocytoma can be successfully rescued by mechanical support with emergency ECMO. Surgical resection of a catecholamine-secreting tumor can safely be performed under protective extracorporeal circulation and oxygenation early after cardiogenic shock.

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References