Venoarterial extracorporeal membrane oxygenation for refractory cardiogenic shock induced by adrenal lesions: a case report and review of the literature

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Abstract

Background

Most adrenal disorders have an insidious onset and are not severe. Only a minority of adrenal lesions can lead to life-threatening cardiovascular crisis. Venoarterial extracorporeal membrane oxygenation (VA-ECMO) is an extracorporeal life support technology that can partially replace human cardiopulmonary function. To date, many case reports have indicated that this technology can temporarily help patients survive critical condition and to achieve a good prognosis.

Case presentation: A 27-year-old male patient presented to the emergency department with sudden dyspnoea and haemoptysis. Pulmonary oedema, cardiogenic shock and cardiac arrest occurred immediately after admission, prompting VA-ECMO treatment. After the patient’s haemodynamics were stabilized, multiple endocrine neoplasia type 2 (MEN2) was detected via laboratory tests and imaging examination. After the withdrawal of VA-ECMO support, surgical resection of the lesions was performed, and a good prognosis was achieved.

Conclusion

VA-ECMO can be used as an emergency treatment for refractory cardiogenic shock caused by adrenal lesions. However, a large number of prospective studies are still needed to confirm its safety and efficacy and to clarify the timing of initiation and assessment of patient prognosis.

Background

The adrenal gland is an important endocrine organ in the human body that secretes a variety of hormones that regulate human physiological functions. Diseases caused by adrenal lesions can have an insidious onset, a chronic process, or a sudden onset presenting as a critical illness, which is easily missed or clinically misdiagnosed.

Recently, several young patients with unknown sudden and refractory cardiogenic shock or cardiac arrest were successfully treated in our emergency department by the application of venoarterial extracorporeal membrane oxygenation (VA-ECMO); these life-threatening conditions were finally determined to have been caused by adrenal lesions. These cases highlight the bridging role of VA-ECMO and merit further review. [1]

Case Presentation

A 27-year-old male patient who had experienced diarrhoea for the prior two days was admitted to the emergency room due to palpitations, shortness of breath, cough and haemoptysis for 6 hours. His family disclosed that he had a long history of refractory hypertension and paroxysmal supraventricular tachycardia that could not be medically managed as well as intermittent headaches and palpitations in the past two years. The patient did not have a history of diabetes, but had smoked approximately 10-20 cigarettes a day for the past 10 years. No relevant family history was reported. Physical examination showed tachypnoea (26 breaths/min), tachycardia (131 beats/min), a blood pressure of 117/80 mmHg, no fever, and blood oxygen saturation in room air of 84%. The cardiac auscultation was normal, but pulmonary auscultation revealed bilaterally dispersed crackles. The patient was pale, with cold and clammy extremities and did not exhibit oedema in either lower extremity. Emergency laboratory findings showed blood gas pH = 7.37, PaCO₂ = 29 mmHg, PaO₂ = 55 mmHg, base excess (BE) = -9 mmol/L, HCO₃⁻ =16.7 mmol/L, white blood cell count (WBC) = 20.8x10⁹/L, platelet count (PLT) = 387x10⁹/L, hemoglobin (Hb) = 164 g/L, creatine kinase (CK) = 334.1 U/L, CK-MB = 33.2 U/L, Myoglobin = 163.8 µg/L, amylase (AMS) = 249.4 U/L, lactic acid (Lac) = 4.8 mmol/L, high-sensitivity cardiac troponin I (hs-cTnI) = 2.54 ng/mL, N-terminal pro b-type natriuretic peptide (NT-proBNP) < 100 pg/mL, procalcitonin (PCT): negative, D-dimer = 1.7 mg/L, and liver and kidney function: normal. A chest CT scan showed diffuse multiple ground-glass opacities in both lungs and suspected alveolar haemorrhage (Figs 1-2). The clinical impression was unexplained pulmonary oedema, respiratory failure type II.

The patient was given high-flow oxygen in the rescue room but still experienced shortness of breath, dyspnoea, irritability, and low pulse oxygen saturation, thus, he was transferred to the emergency intensive care unit (EICU). The patient was successively provided with noninvasive and invasive ventilators to assist breathing in the EICU, and pink foamy sputum was seen in the airway during orotracheal intubation. He then underwent sudden cardiac arrest. Spontaneous circulation was restored immediately after cardiopulmonary resuscitation for approximately 1 minute, but a large dose of vasoactive drugs (norepinephrine: 1 µg/kg/min) had to be administered and he still exhibited low blood pressure and anuria. Bedside echocardiography showed normal atrioventricular cavity size and decreased left ventricular function (left ventricular ejection fraction [LVEF] of approximately 9.5%) (Fig 3). The refractory cardiogenic shock was treated by initiation of VA-ECMO (flow = 1.9 L/min). However, approximately 3 hours later, the ultrasound evaluation revealed LV dilatation and restricted aortic valve patency, thus, an intra-aortic balloon pump (IABP) was implanted immediately. Cardiotonic, diuretic, sedative, analgesic, and high-dose αβ-adrenergic antagonists were administered simultaneously. The patient was weaned off VA-ECMO after 5 days, the IABP was withdrawn after 6 days, and tracheal intubation was removed after 7 days, replaced with high-flow oxygen. After 17 days, the patient was transferred out of the EICU.

During EICU treatment, the test results showed the following blood hormone levels: dopamine = 0.202 nmol/L (0.0-0.13), epinephrine = 10.948 nmol/L (0.055-1.09), norepinephrine = 2.895 nmol/L (0.47-3.07), calcitonin > 2,000 pg/ml (0-9.52), parathyroid hormone = 246.8 pg/ml (15-65), tumour
marker carcinoembryonic antigen (CEA) = 7.32 ng/ml (<5.0), and RET variant in the disease-causing gene. After VA-ECMO and artificial ventilation were discontinued, imaging examination of the patient revealed masses in the bilateral adrenal glands. The largest slice on the right side was approximately 44 mm × 39 mm, and the largest slice on the left side was approximately 42 mm × 31 mm. The shape was irregular with a clear boundary, and the mass density was uneven (Figs 4-5). A neck ultrasound showed a 12 × 6 mm hypoechoic nodule in the middle of the left lobe of the thyroid and a 6.9 × 4.3 mm hypoechoic nodule in the middle of the right lobe of the thyroid. Multiple hypoechoic nodules were detected in the bilateral VI area of the neck, and the larger nodules were 11 × 5 mm (right) and 16 × 7 mm (left). Therefore, this patient was suspected of multiple endocrine neoplasia type 2 (MEN2), catecholamine cardiomyopathy, and cardiogenic shock.

After leaving the EICU, he was transferred to the endocrinology department for preoperative preparation and was given α/β adrenergic receptor blockers and fluid replacement therapy. A laparoscopic bilateral adrenal tumour resection was performed in the urology department, the pathological result was pheochromocytoma (Fig 6). The patient was given hydrocortisone replacement therapy after surgery. A bilateral thyroidectomy, bilateral central neck lymph node dissection and bilateral parathyroidectomy were performed in the thyroid department approximately 1 month later. The pathological results were medullary thyroid carcinoma and parathyroid neoplastic hyperplasia. The patient was treated with levothyroxine after surgery. Therefore, the final diagnosis was MEN2, pheochromocytoma, medullary thyroid carcinoma, and parathyroid neoplastic hyperplasia.

**Treatment timeline:**

<table>
<thead>
<tr>
<th>Date and time</th>
<th>Event</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oct 17th at 22:43</td>
<td>Admitted to the emergency department</td>
</tr>
<tr>
<td>Oct 18th at 6:30</td>
<td>Administered high-flow oxygen, but still exhibited shortness of breath and palpitations, heart rate (HR) = 135 bpm, respiratory rate (RR) = 32 breaths/min</td>
</tr>
<tr>
<td>Oct 18th at 10:35</td>
<td>Transferred to the EICU</td>
</tr>
<tr>
<td>Oct 18th at 13:30</td>
<td>Provided with a noninvasive ventilator assist, but restless and unable to achieve mask fit, still exhibiting shortness of breath and palpitations</td>
</tr>
<tr>
<td>Oct 18th at 17:00</td>
<td>Orotracheal intubation and invasive ventilator assist (pressure-controlled synchronized intermittent mandatory ventilation [P-SIMV], positive end-expiratory pressure [PEEP] = 8 cmH₂O, PEEP⁺ = 10 cmH₂O, FiO₂ = 80%)</td>
</tr>
<tr>
<td>Oct 18th at 17:30</td>
<td>Heart rate and blood pressure dropped, norepinephrine and vasopressin administered</td>
</tr>
<tr>
<td>Oct 18th at 18:05</td>
<td>Cardiac arrest, return of spontaneous circulation after approximately 1 min of cardiopulmonary resuscitation</td>
</tr>
<tr>
<td>Oct 18th at 18:30</td>
<td>Patient experienced anuria. Central venous catheter placement and left lower extremity artery catheterization for haemodynamic monitoring, bedside ultrasound assessment</td>
</tr>
<tr>
<td>Oct 18th at 20:15</td>
<td>Right femoral artery and vein catheterization, VA-ECMO initiated</td>
</tr>
<tr>
<td>Oct 18th at 23:30</td>
<td>Catheterization of the left femoral artery to initiate the IABP</td>
</tr>
<tr>
<td>Oct 23rd at 16:30</td>
<td>Weaning from VA-ECMO</td>
</tr>
<tr>
<td>Oct 24th at 10:30</td>
<td>Withdrawal of the IABP</td>
</tr>
<tr>
<td>Oct 25th at 14:45</td>
<td>Removal of the endotracheal tube, high-flow oxygen provided</td>
</tr>
<tr>
<td>Nov 2nd at 9:30</td>
<td>Nasal cannula for oxygen</td>
</tr>
<tr>
<td>Nov 4th</td>
<td>Out of the EICU</td>
</tr>
<tr>
<td>Dec 2nd</td>
<td>Laparoscopic bilateral adrenal tumour resection</td>
</tr>
<tr>
<td>Jan 19th</td>
<td>Bilateral thyroidectomy, bilateral central cervical lymph node dissection, and bilateral parathyroidectomy</td>
</tr>
<tr>
<td>Jan 22nd</td>
<td>Discharged from the hospital</td>
</tr>
</tbody>
</table>
A review of the literature

Some adrenal lesions can lead to refractory cardiogenic shock and even cardiac arrest, requiring the support of VA-ECMO. We conducted a literature review of the MEDLINE database through the PubMed search engine between 2016 and 2022 using the keywords "adrenal and extracorporeal membrane oxygenation" and "pheochromocytoma and extracorporeal membrane oxygenation" in the "AB/TI" field. Observational studies and case series reports on adult patients who required VA-ECMO for adrenal lesion-induced cardiogenic shock were eligible for inclusion. Studies on children or newborns, that utilized VV-ECMO, and those not in English were excluded.

A total of 36 papers were located. Three papers involving VV-ECMO, 5 papers on children, 2 papers not in English and 2 unrelated papers were excluded, thus, 24 papers remained. See Table 1 for a summary. Of these papers, two were single-centre retrospective studies: one study documented 13 VA-ECMO cases with pheochromocytoma but did not record the lesion site, and the other study documented 9 VA-ECMO cases, including 1 nonadrenal case with paraganglioma. The other 22 papers were all case reports that covered 23 VA-ECMO cases, including 3 nonadrenal cases.

A total of 28 cases with adrenal lesions requiring VA-ECMO support were finally summarized and analysed. Twenty-one (75%) patients were women, including 3 pregnant women. The median patient age was 38 years old (range: 19-59 years). Cardiac arrest occurred in 11 (39%) patients before and during VA-ECMO initiation. The duration of VA-ECMO support was recorded in 25 case reports, the average duration was 5.32 days (range: 1-14 days). Four patients received a combination of IABPs, and 3 patients received a combination of Impella. The ICU duration was documented in 9 cases, with an average of 20 days (range: 4-40 days).

Three patients died without surgery in 8 cases documented in a retrospective study. All 20 patients in the case reports were discharged. A total of 20 case reports documented adrenal surgery, of which 17 documented the duration from onset to surgery, which had an average of 32.6 days (range: 7-70 days). Four patients with adrenal insufficiency did not require surgery.

The aetiology of adrenal lesions requiring VA-ECMO was mainly pheochromocytoma (23 cases, 82.1%), of these, 22 cases were unilateral (including 1 case of MEN1) and 1 case was bilateral (MEN2). The second cause of these adrenal lesions was adrenal insufficiency, which was primary in 3 cases (1 case of adrenal tuberculosis, and 2 cases of autoimmune adrenal disease) and secondary to pituitary insufficiency in 1 case. Another cause was right adrenal Ewing's sarcoma (1 case).

The hormones that play a major role in adrenal lesions that induce cardiogenic shock and even cardiac arrest are catecholamines secreted by the adrenal medulla, the clinical manifestations of which are catecholamine cardiomyopathy, stress cardiomyopathy, Takotsubo syndrome or broken heart syndrome. Additionally, adrenal insufficiency leads to a lack of corticosteroids, which clinically manifests as adrenal crisis, electrolyte imbalance, and circulatory failure.

### Table 1

| #: retrospective observational studies | *: Cases of nonadrenal lesions |

**Discussion**

The adrenal gland is an important endocrine organ in the human body and includes the medulla and the cortex. The hormones secreted by the adrenal cortex are divided into three categories. Zona glomerulosa cells secrete mineralocorticoids (mainly aldosterone), zona fasciculata cells secrete glucocorticoids (mainly cortisol), and zona reticularis cells mainly secrete sex hormones. The adrenal medulla secretes epinephrine and norepinephrine, which are catecholamines. The hormones with the greatest impact on the cardiovascular system and haemodynamics are aldosterone, cortisol, and the catecholamines.

Aldosterone is an important hormone that regulates the body's water and salt metabolism. Excessive aldosterone secretion results in the retention of sodium and water, causing hyponatremia, hypertension and a decrease in serum potassium. In contrast, if aldosterone is deficient, too much sodium and water are excreted, leading to a decrease in serum sodium levels, blood pressure, and urinary potassium excretion as well as an increase in serum potassium levels.

Glucocorticoids affect carbohydrate, protein, and fat metabolism; water and salt balance; blood cells; the circulatory system; and the stress response. They can enhance the sensitivity of vascular smooth muscle to catecholamines, which helps to increase the tension of blood vessels and maintain blood pressure. In addition, these hormones can reduce the permeability of capillary walls, reduce plasma leakage, and help to maintain blood volume.

Medullary hormones are secreted in large quantities during the stress response. They act on the central nervous system, increasing its excitability, and placing the body in a state of alertness and sensitivity: breathing deepens and accelerates, and pulmonary ventilation increases; the contractility of muscles increases; cardiac output and blood pressure increase, thus accelerating blood circulation; and visceral vasoconstriction, skeletal muscle vasodilation and blood flow increase, redistributing blood flow throughout the body to facilitate supply to vital organs during the stress response.
<table>
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<tr>
<th>First author</th>
<th>Publication Year</th>
<th>Patients (n)</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Adrenal lesions</th>
<th>CA before ECMO initiation</th>
<th>MCS, duration (days)</th>
<th>ICU length of stay (days)</th>
<th>Admission-to-surgery interval (days)</th>
<th>Hospital survival</th>
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<td>Daniel[11]</td>
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<td>Gender</td>
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<td>Treatment 2</td>
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<td>20</td>
<td>F</td>
<td>Right adrenal</td>
<td>Ewing’s sarcoma</td>
<td>Yes</td>
<td>ECMO, 11</td>
<td>19</td>
<td>NR</td>
<td>Yes</td>
</tr>
</tbody>
</table>

This article reported a case of a complex rare disease in which the cause of pulmonary oedema, cardiogenic shock, and cardiac arrest was not immediately identified on admission to the emergency department. Laboratory tests, imaging, and pathological findings later confirmed MEN2, bilateral pheochromocytoma with medullary thyroid carcinoma and parathyroid neoplastic hyperplasia. The patient's hormone levels were disturbed, causing haemodynamic instability and cardiac arrest. VA-ECMO combined with an IABP helped the patient to survive critical condition. After surgery, the patient was given oral hormone replacement therapy, and recovered cardiac function without experiencing any after-effects, such as lower extremity ischaemia.

Through a review of the literature, we found that adrenal lesions lead to changes in hormone levels in the body, resulting in various crises that require ECMO treatment, especially among patients under the age of 60; these crises may clinically manifest as water and electrolyte acid-base imbalance, sudden cardiac arrest, haemodynamic instability, stress cardiomyopathy, Takotsubo syndrome, etc. Pheochromocytoma is the most common cause of adrenal lesions, followed by adrenal insufficiency. These patients may be admitted to the emergency department upon sudden deterioration, but their condition often changes rapidly, and they also recover rapidly. VA-ECMO can be used as a bridge to recovery, to a more durable bridge, to definitive treatment, or to decision for unexplained refractory cardiogenic shock and cardiac arrest in contemporary emergency departments. The support from VA-ECMO can provide an opportunity to clarify the aetiology of the patient, restore cardiac function, and help the patient survive crisis. Thus, the use of VA-ECMO should be promoted and applied (strictly complying with the indications of the machine), and a good prognosis can be achieved with later surgical resection of the lesions or hormone replacement therapy.
Conclusions

Adrenal disease should be considered in patients presenting with acute cardiovascular crisis, especially in young patients admitted to the emergency department. If emergency treatment for cardiogenic shock is ineffective, VA-ECMO should be implemented to temporarily replace cardiopulmonary function and create an opportunity to treat the primary disease. However, a large number of prospective studies are still needed to confirm the safety and efficacy of VA-ECMO and to clarify the timing of its initiation and assessment of prognosis in these patients.

Abbreviations

VA-ECMO
veno-arterial extracorporeal membrane oxygenation
VV-ECMO
veno-venous extracorporeal membrane oxygenation
MEN2
multiple endocrine neoplasia type 2
CT
computerized tomography
LV
left ventricular
CA
cardiac arrest
MCS
mechanical circulatory support
PHEO
pheochromocytoma
PARA
paraganglioma
ELVV
endovascular left ventricular venting
APS
autoimmune polyendocrinopathy syndrome
PSAI
panhypopituitarism with secondary adrenal insufficiency
TB
tuberculosis
VAD
ventricular assist device
AAD
autoimmune Addison's disease
NR
not reported
NA
not applicable

Declarations

Ethical approval and consent to participate

Ethics approval and consent were waived because this study is a review of literature with a retrospective case report on one patient that gave consent to participate for publication.

Consent for publication

The patient gave consent for publication.

Availability of data and materials

All data relevant to the study are included in the article.
Competing interests

All authors declare that they have no competing interests.

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Author's contributions

ZLP, MXY, WP, ZZ, LB, YN were responsible for treatment and management of this patient. HGQ, SCS, HSS were responsible for data collection. YN was responsible for paper-writing. All authors have read and approved the manuscript.

Acknowledgements

All ECMO team members who actively participated in providing healthcare for that patient should be fully acknowledged.

References


**Figures**

**Figure 1**

CT scan at admission showing diffuse multiple ground-glass opacities in both lungs and suspected alveolar haemorrhage.
Figure 2

CT scan at admission showing diffuse multiple ground-glass opacities in both lungs and suspected alveolar haemorrhage.

Figure 3

Bedside transthoracic echocardiography at admission showed decreased left ventricular function.
Figure 4

Transverse plane of CT scan prior to surgery showing masses in the bilateral adrenal glands. (Red arrow)

Figure 5

Coronal plane of CT scan prior to surgery showing masses in the bilateral adrenal glands. (Red arrow)
Figure 6

Macroscopic view of the surgical specimen. (Bilateral adrenal masses)