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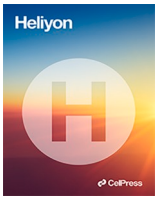

Case report

Temporary mechanical circulatory support as a bridge to durable left ventricular assist device as destination therapy in fulminant giant cell myocarditis : A case report

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ABSTRACT

Fulminant giant cell myocarditis is a fatal form of acute myocarditis leading to a rapid-onset clinical presentation with lethal arrhythmias, acute heart failure, or cardiogenic shock requiring mechanical circulatory support. We report the case of a 52-year-old female diagnosed with fulminant myocarditis requiring veno-arterial extracorporeal membrane oxygenation (V-A ECMO) and intra-aortic balloon pump(IABP) support. Due to hemodynamic instability, she was transferred to our hospital by helicopter on day 4. On arrival at our hospital, she underwent percutaneous balloon atrial septostomy to decompress the left ventricle. Although the left ventricular distension and pulmonary edema improved after atrial septostomy, no signs of biventricular function recovery were identified on day 14. On day 23, V-A ECMO and IABP were switched to a durable left ventricular assist device(LVAD) system and a right ventricular assist device(RVAD) with ECMO (RVAD-ECMO) under median sternotomy. On day 37, RVAD-ECMO was eventually removed and rehabilitation was started with the remaining LVAD support as destination therapy. On day 78, the patient was finally discharged with LVAD support to follow-up as an outpatient. This case underscores the importance of a multidisciplinary approach and rigorous monitoring to optimize outcomes in the treatment of fulminant giant cell myocarditis.

1. Introduction

Fulminant giant cell myocarditis, defined by the presence of either cardiogenic shock, lethal arrhythmias, or hemodynamic instability, is a fatal form of acute myocarditis and is associated with an increased risk of death [1]. An inflammatory infiltrate with giant cells and myocardial necrosis is the characteristic of fulminant giant cell myocarditis [2]. Fulminant giant cell myocarditis requires active treatment, including aggressive circulatory support when deemed necessary. We report a case of fulminant giant cell myocarditis which was supported with veno-arterial extracorporeal membrane oxygenation for inter-hospital air transport and as a bridge to implantation of a left ventricular assist device(LVAD).

The patient provided a written informed consent form, agreeing to the treatment plan, and willing to disclose the details of the medical record.

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2. Case presentation

A 52-year-old woman with a history of Sjogren's syndrome, an autoimmune disease affecting the body's moisture levels, initially presented to a regional hospital complaining of sudden chest tightness. A cardiac catheterization performed revealed normal coronary arteries. However, on day 3, the patient developed severe respiratory distress and experienced cardiac arrest characterized by severe ventricular arrhythmias refractory to conventional cardiopulmonary resuscitation efforts. Following tracheal intubation, she underwent peripheral venous-arterial extracorporeal membrane oxygenation (V-A ECMO) as a rescue therapy, accompanied by intra-aortic balloon pump (IABP) implantation. The presumptive diagnosis was fulminant myocarditis, prompting the initiation of immunosuppressive therapy with intravenous methylprednisolone administered at a dose of 500mg/day for 3 days. On day 4, due to the worsening cardiovascular status and persistent electrical instability despite the utilization of V-A ECMO and IABP, the patient was transferred to our hospital by helicopter for further management of her cardiac condition.

Upon arrival at our hospital, the patient's blood pressure was recorded as 83/64 mmHg while receiving mechanical circulatory support with ECMO (operating at a flow rate of 3.5 L/min) and IABP. Blood tests revealed several abnormalities: a platelet count of 14 $\times$ 10^9/L; serum troponin T levels at 14210 ng/L (significantly above the reference range 0–14 ng/L); an N-terminal pro B-type natriuretic peptide(NT-proBNP) level of 8296 pg/mL (exceeding the normal range of 0–125 pg/mL); a serum alanine aminotransferase (ALT) level of 116 IU/L(reference range 0–33 IU/L); and a serum creatinine level of 134 μmol/L(reference range 45-84umol/L). Arterial blood gases showed metabolic acidosis of a pH of 7.25 was present, and a lactate level of 4.87 mmol/L (reference range, 0.5–2.2mmol/L). The electrocardiogram (ECG) revealed ventricular tachycardia (Fig. 1). Bedside transthoracic echocardiography showed that the global wall motion was severely depressed, with an estimated left ventricular ejection fraction(LVEF) of approximately 10 %. Chest X-ray showed pulmonary edema(Fig. 2A). The patient's low platelet count was attributed to a combination of factors, including inflammation, heparin use during ECMO, and mechanical damage resulting from ECMO and IABP transitions. These factors could have acted individually or in concert to produce this effect. To address this issue, treatments such as platelet transfusions were administered. Due to severe LV distension and intractable pulmonary congestion, a decision was made to proceed with percutaneous balloon atrial septostomy to decompress the LV. She was transferred to the operating room and underwent atrial septostomy (Fig. 2B). The timeline and clinical course chart are shown in Table 1.

After atrial septostomy, on day 7, the left ventricular distension and pulmonary edema were improved(Fig. 2C), and her incessant slow ventricular tachycardia resolved to sinus rhythm. However, no cardiac recovery was observed within the first two weeks. The patient's hepatic and renal functions continued to deteriorate. The patient underwent a tracheostomy. Although she was started on continuous venous-venous hemodialysis, received transfusions of blood products, inotropic, and had vasopressor therapy adjustment, no signs of biventricular function recovery were identified on day 14. Therefore, the cardiology, cardiothoracic surgery, and critical care teams reassessed the patient's heart function, and it was determined that she might require a biventricular assist device. On day 23, V-A ECMO and IABP were switched to a durable LVAD system and a right ventricular assist device. A left VAD system(CH-VAD,

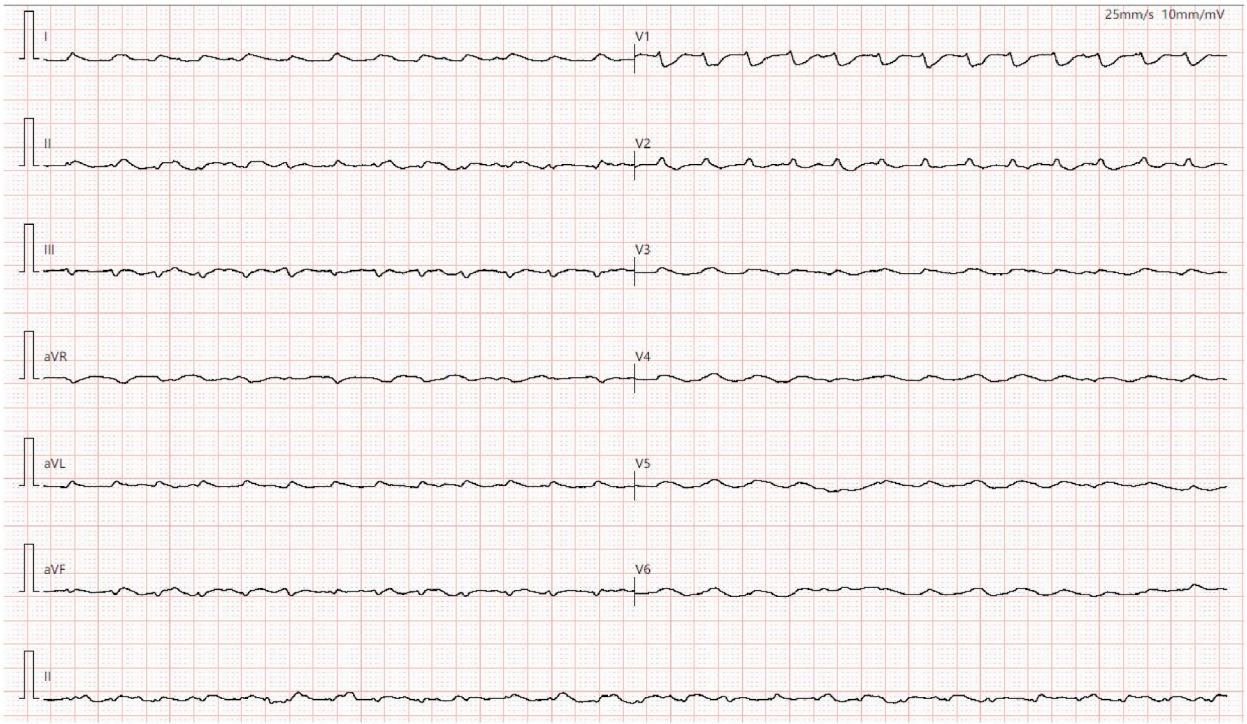

Fig. 1. The patient's electrocardiogram on day 4 after onset showed ventricular tachycardia.

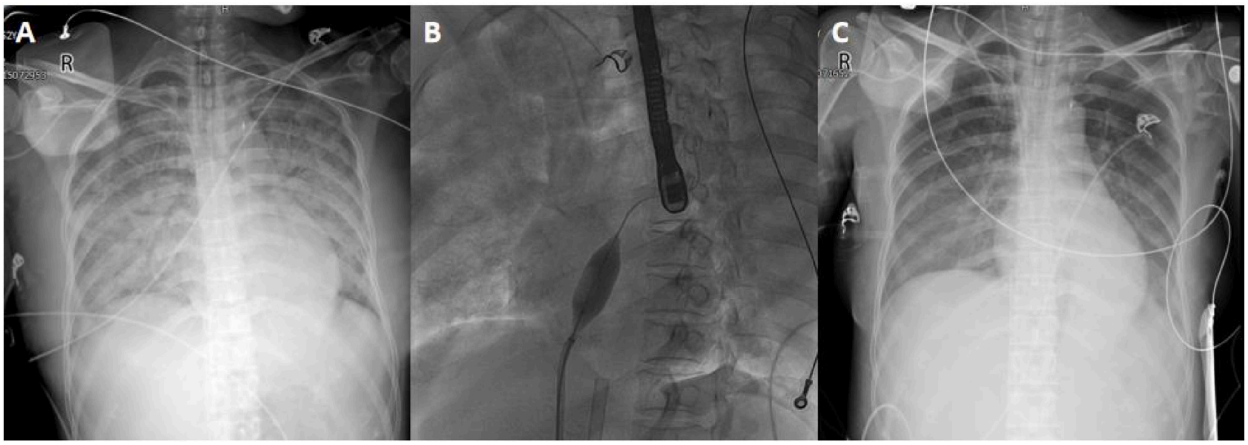

Fig. 2. On hospital admission, the chest X-ray showed pulmonary edema (A); The patient underwent percutaneous balloon atrial septal septostomy (B); The left ventricular distension and pulmonary edema improved after atrial septostomy (C).

Table 1
Timeline and clinical course chart.

Timeline	Assist device	MAP (mmHg)	CVP (mmHg)	PAP (mmHg)	Dobutamine (μg/kg/min)	Dopamine (μg/kg/min)	Noradrenaline (μg/kg/min)
On day 1–3(Before admission)	V-A ECMO + IABP	64	NO	NO	NO	10	0.5
On day 4(admission)	V-A ECMO + IABP Atrial septostomy	70	21	NO	5	8	0.2
On day 7(ICU)	V-A ECMO + IABP	74	15	NO	5	5	0.2
On day 14(ICU)	V-A ECMO + IABP	65	18	NO	5	8	0.15
On day 23(ICU)	LVAD + V–P ECMO	77	16	22	5	5	0.05
On day 25(ICU)	LVAD + V–P ECMO	81	10	24	3	NO	NO
On day 37(ICU)	LVAD	86	8	26	0	NO	NO

MAP, mean arterial pressure; CVP, central venous pressure; PAP,pulmonary arterial pressure; V-A ECMO, veno-arterial extracorporeal membrane oxygenation; V–P ECMO, veno-pulmonary extracorporeal membrane oxygenation.

ChinaHeart Biomedical,Inc., Suzhou, China) was established from the left ventricular apex to the ascending aorta, and a right ventricular assist device with ECMO(RVAD-ECMO) was established from the right atrium, using a 14mm artificial graft, to the pulmonary artery, using a 10mm artifical graft, under median sternotomy.

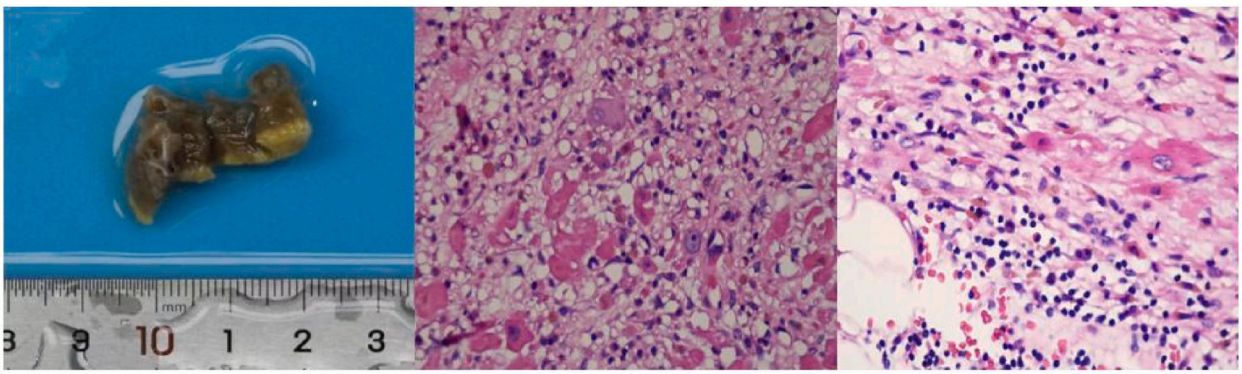

Fig. 3. Histological findings from the left ventricular apex, showed myocardial necrosis, interstitial edema, and fibrosis with infiltration by diffuse monocytes and scattered multinucleated giant cells.

With the assistance of both LVAD and RVAD-ECMO mechanical support, the patient's hepatic and renal functions gradually improved. On the 25th day of treatment, the patient was successfully weaned off ventilation. Histological analysis of a resected specimen from the left ventricular apex, revealed myocardial necrosis, interstitial edema, and fibrosis, accompanied by infiltration of diffuse monocytes and scattered multinucleated giant cells(Fig. 3). Based on these findings, a pathologic diagnosis of giant cell myocarditis was established. As the patient's RV function continued to improve, the RVAD-ECMO was eventually removed on Day 37 through a median re-sternotomy. Following this, rehabilitation commenced with the patient relying on durable LVAD support as a long-term therapy. On Day 78, the patient's LV function had progressively improved, achieving an ejection fraction (EF) of approximately 40 %(Fig. 4). She was ultimately discharged from the hospital with LVAD support and scheduled for outpatient follow-up care.

3. Discussion and conclusions

The present case report describes a severe cardiogenic shock associated with fulminant giant cell myocarditis necessitating bridge with ECMO to durable LVAD implantation as destination therapy. It has been demonstrated that short-term mechanical circulatory support should be switched to either durable mechanical circulatory support or as a bridge to urgent heart transplantation when cardiac function recovery cannot be expected [3].

Giant cell myocarditis, when associated with other autoimmune disorders like systemic lupus erythematosus, vasculitis, Hashimoto's disease, myasthenia gravis, and inflammatory bowel disease, is considered an autoimmune disorder [4]. One study indicated the presence of autoimmune disorders in approximately 20 % of the affected population [5]. Sjögren's syndrome is also an autoimmune condition, and as seen in our current case, giant cell myocarditis can potentially be associated with it. Giant cell myocarditis is a fatal manifestation, known as T-cell-mediated inflammatory myocarditis, frequently resulting in a rapid-onset clinical presentation characterized by refractory ventricular arrhythmias, atrioventricular block, cardiogenic shock, or sudden cardiac death [4,6,7].

Despite the early symptoms of fulminant giant cell myocarditis being similar to those of other froms of acute myocarditis, this condition can progress rapidly towards fatality, often within days to months [8]. During the acute phase, hemodynamic instability necessitates cardiovascular support. Therefore, mechanical circulatory support devices might be required as a bridge to recovery, as a form of destination therapy or for cardiac transplantation [7]. Currently, several types of mechanical circulatory support devices are in use. The intra-aortic balloon pump (IABP) is the most widely available mechanical circulatory support device and is adequate for most patients with fulminant myocarditis. However, in cases where patients experience progressive hemodynamic deterioration, the IABP alone may not restore essential circulation. In such situations, additional mechanical support, such as ECMO should be promptly employed [9].

As a short-term mechanical assist device, ECMO, which guarantees rapid and full cardio-respiratory assistance, is considered a well-known bridging therapy in the setting of fulminant giant cell myocarditis. However, while V-A ECMO can effectively provide systemic circulatory support, it does not completely unload the LV in cases of severe LV failure and the prognosis of patients with LV failure is reported not to be as good as expected [10,11]. It may be likely that V-A ECMO alone might increase LV afterload, leading to a further rise in end-diastolic LV pressure and a subsequent reduction of subendocardial coronary flow [12]. The rapid increase in LV diastolic pressure can lead to pulmonary congestion; thus, additional LV venting strategies to prevent LV distension and pulmonary edema may be required during V-A ECMO support [13]. Low-dose inotropes and vasodilators and/or IABP implantation are the most common strategies for reducing LV afterload [14]. Other strategies for directly unloading the left heart, such as the microaxial pump Impella system, TandemHeart, and percutaneous balloon atrial septostomy, have been increasingly used in recent years [15–17]. In our case, V-A ECMO was used in combination with IABP. However, IABP had a limited effect in reducing LV pressure and pulmonary congestion in this case. Impella can drain left ventricular blood to the aorta, achieving a good left ventricular decompression effect. When used in combination with ECMO, it can increase perfusion flow. As our institution does not have Impella, we performed percutaneous balloon atrial septostomy to decompress the LV as soon as she was transported to our hospital when her hemodynamic status was not significantly improved. After atrial septostomy, the LV distension and pulmonary edema were significantly improved. The incessant ventricular arrhythmia also automatically resolved into sinus rhythm due to the decrease of LV tension.

Giant cell myocarditis can affect both ventricles, necessitating biventricular support in certain cases. Tominaga et al. [18] reported a case in which a combination of V–P ECMO and an Impella® 5.0 left ventricular aortic pump was successfully used as a temporary biventricular assist device for managing cardiogenic shock due to fulminant myocarditis. Ma et al. [19]described a similar case of a patient with fulminant giant cell myocarditis who also required biventricular support. In a recent systematic review conducted by Patel et al. [20], the outcomes of patients with giant cell myocarditis requiring mechanical circulatory support were examined. The study found that 76.7 % of patients needed biventricular support, and 58.5 % underwent heart transplantation. In the present case, although the LV dilation and pulmonary edema improved after LV unloading, there was no recovery of biventricular function even after approximately three weeks of support with ECMO and IABP. Therefore, the decision was made to replace the ECMO and IABP assist devices with a long-time LVAD and a temporary right ventricular assist device with ECMO support. Fortunately, with this enhanced biventricular support, the RV function recovered quickly, allowing for the successful removal of the RVAD-ECMO. Ultimately, the patient was able to retain the durable LVAD as a long-term therapeutic option and was discharged after completing rehabilitation.

In conclusion, we have reported a challenging case of fulminant giant cell myocarditis complicated by cardiogenic shock and life-threatening arrhythmia, necessitating the utilization of mechanical circulatory support. The patient's initial stabilization with short-term devices, such as V-A ECMO, IABP, and RVAD-ECMO, followed by a successful transition to long-term LVAD therapy, underscores the effectiveness of a multifaceted approach combining temporary and permanent mechanical support in managing this critical condition. This case highlights the importance of a tailored strategy that considers the patient's hemodynamic status and potential complications, emphasizing the need for a comprehensive and sequential utilization of mechanical circulatory support devices in the

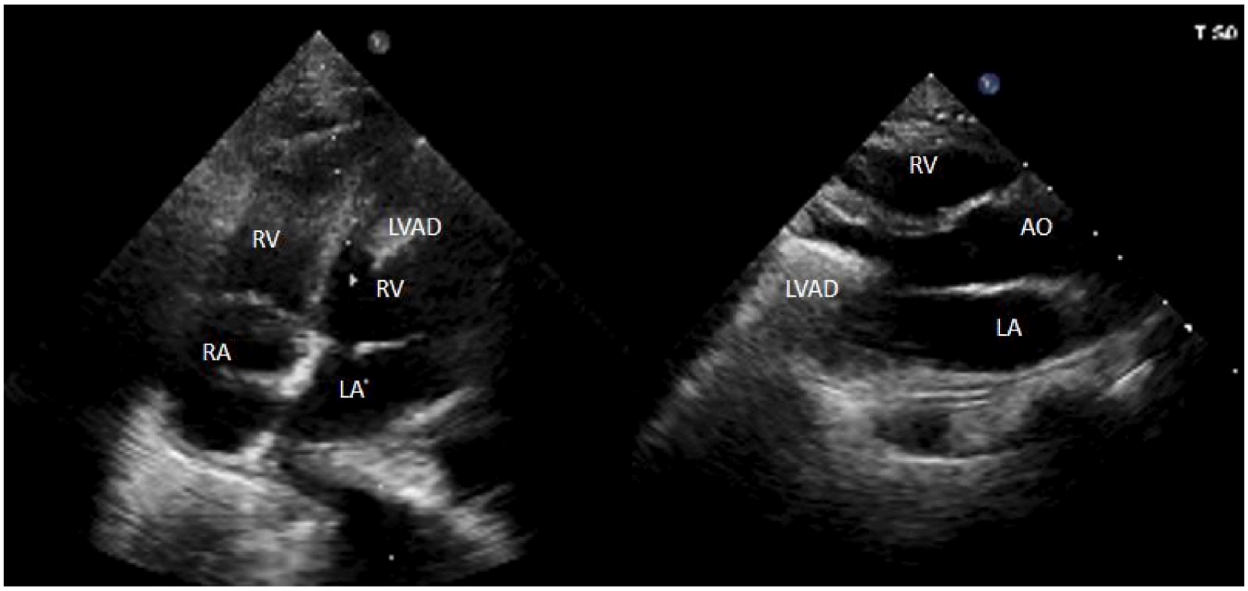

Fig. 4. Transthoracic echocardiography before discharge showed progressive improvement in left ventricular function with an EF of approximately 40 % under LVAD support.

management of fulminant giant cell myocarditis.

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Additional information

No additional information is available for this paper.

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Data included in article/supp. Material/referenced in article.

CRediT authorship contribution statement

Zhi-Ping Zhang: Writing – review & editing, Writing – original draft, Supervision, Resources, Methodology, Investigation, Formal analysis, Data curation, Conceptualization. **Pen-Yu Guo:** Writing – review & editing, Supervision, Data curation. **Qi-Shen Ye:** Data curation. **Yong Zhang:** Supervision, Investigation.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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