Journal of Cardiology Cases 29 (2024) 7–10

Contents lists available at ScienceDirect

Journal of Cardiology Cases

journal homepage: www.elsevier.com/locate/jccase

Case Report

A case of progressive right ventricular failure with ventricular arrhythmia and aortic insufficiency after implantable left ventricular assist device implantation

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ARTICLE INFO

Article history:
Received 9 March 2023
Received in revised form 17 August 2023
Accepted 23 August 2023

Keywords:
Right ventricular failure
Ventricular assist device
Ventricular arrhythmia
Cardiac implantation

ABSTRACT

Right ventricular failure (RVF) is a serious complication after left ventricular assist device (LVAD) implantation. In this report, a case of RVF that developed over two years after LVAD implantation is presented. The patient was a 12-year-old male with dilated phase of hypertrophic cardiomyopathy. He had no risk factors for early or late-onset RVF. However, his right ventricular function worsened after he developed ventricular arrhythmia (VA), and right ventricular dysfunction became exacerbated with an increasing frequency of VAs. He also developed moderate aortic insufficiency (AI), which became severe. Two years after implantation, he was admitted for treatment of recurrent ventricular tachycardia and became inotropic-dependent during hospitalization. Finally, he underwent successful heart transplantation 2 years and 9 months after LVAD implantation. This case suggests that vicious cycle of RV dysfunction, recurrent VAs and severe AI could lead to RVF in patients without known risk factors for RVF, even long after LVAD implantation.

Learning objective: This report shows a progressive right ventricular failure (RVF) two years after left ventricular assist device (LVAD) implantation. Although the patient had no known risk factor, vicious circle of RV dysfunction, ventricular arrhythmias (VAs) and aortic insufficiency (AI) lead to RVF. Patients with LVAD as destination therapy will increase and require long-term LVAD management. We should recognize that these patients could develop RVF even years after LVAD implantation in association with VAs and AI.

Introduction

Implantable left ventricular assist devices (LVADs) are increasingly used for the treatment of patients with severe heart failure. However, 3.9–53 % of patients with LVAD has been reported to develop right ventricular failure (RVF), a major cause of morbidity and mortality after LVAD implantation [1]. RVF after LVAD implantation is classified according to its onset, early or late [2]. Several preoperative risk factors for early RVF have been identified, such as low right ventricular stroke work index (RVSWI), high central venous pressure, and pulmonary vascular resistance [3]. Late RVF usually appears weeks after LVAD implantation [4,5]. Past studies have reported several risk factors, such as smaller LV diastolic diameter and renal dysfunction, albeit the clinical characteristics and pathophysiology of late RVF remain elusive [4,5].

In this report, a case of dilated phase of hypertrophic cardiomyopathy (D-HCM), who underwent implantation of EVAHEART (Sun Medical Technology Research Corp., Nagano, Japan), is presented. The patient developed progressive RVF accompanied by ventricular arrhythmia (VA) and aortic insufficiency (AI) more than two years after LVAD implantation.

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https://doi.org/10.1016/j.jccase.2023.08.017

Case report

A 12-year-old male patient presented to the clinic with a systolic heart murmur. He was diagnosed as having HCM, and optimal medical therapy was initiated. Two years later, he was admitted to a hospital for syncope due to complete atrioventricular block. Since left ventricular systolic function was decreased moderately, he received cardiac resynchronization therapy with a defibrillator (CRT-D) device.

Two years later, his symptoms of heart failure worsened to New York Heart Association IV, and an echocardiogram showed LV dilatation and severe systolic dysfunction, consistent with D-HCM (Table 1). Right-heart catheterization revealed atrial pressure of 6 mmHg, RVSWI of 7.6 g·m/m^2, and pulmonary vascular resistance of 0.83 Woods units. An echocardiogram showed that RV function was preserved (Video 1). Because his heart failure was severe (INTERMACS profile 2) and inotrope-dependent, he was considered a candidate for heart transplantation and received an EVAHEART LVAD as a bridge to transplant. The patient had one episode of ventricular tachycardia (VT) requiring defibrillation a month after implantation. Echocardiogram showed slightly decreased RV function (Table 1), but right heart catheterization demonstrated no elevation of central venous pressure (CVP, 10 mmHg). The patient had no other sign of early RVF and was discharged 1.5 months after LVAD implantation.

Four months after implantation, he presented with VA several times. All VAs were successfully terminated by defibrillation, and amiodarone was started. Blood B-type natriuretic peptide (BNP) levels were decreasing (Fig. 1), and an echocardiogram showed mild AI, with recovered RV function (Table 1, Video 2).

One year after implantation, he was admitted for recurrent VAs, which caused low output syndrome (LOS). LOS subsided soon after termination of VT storm by defibrillation. The bilirubin level was also elevated to 3.4 mg/dl and decreased to normal range over time. An echocardiogram showed RV dysfunction (Video 3) and relatively unchanged LV function and dilatation (Table 1) with moderate AI (Video 4). We initiated eplerenone and sildenafil, but he could not tolerate sildenafil because of digestive symptoms. There was no apparent suction of the LV cavity or contact event between the inflow cannula and ventricular wall. Since premature ventricular contractions (PVCs) led to the initiation of VAs, treatment with carvedilol and mexiletine was started, and the setting of the minimum rate of CRT-D was changed to 100 bpm to prevent PVCs. These treatments in combination enabled successful prevention of VAs.

However, 1.5 years after LVAD implantation, he again developed several episodes of VAs, and the frequency of VAs increased each week. He was admitted for treatment of VAs two years after LVAD implantation, but the VAs were refractory to procainamide, lidocaine, carvedilol, amiodarone, and nifekalant hydrochloride. Catheter ablation was not performed considering that the VTs were multifocal (Online Fig. 1) and the perioperative risks. Eventually, the patient needed to be put under sedation with propofol and dexmedetomidine for control of VAs. RV function deteriorated with development of VT storms (Table 1, Video 5), and AI worsened (Video 6). Right ventricular dilation also led to severe tricuspid regurgitation (Table 1). Even after the VAs were controlled, the patient frequently presented with RVF. The bilirubin level was elevated to 2.1 mg/dl with accumulation of pleural effusion. Blood BNP levels were further elevated (Fig. 1) with the presentation of LOS. Dobutamine and milrinone were initiated. In addition, furosemide was administered by intravenous infusion, and dosage was managed using CVP, lactate level, and urine volume as indicators. The patient developed end-stage and inotrope-dependent heart failure even under LVAD support.

Under the administration of inotropes, the hemodynamics were maintained, and alteration of pump speed or invasive strategy including aortic valve replacement (AVR) was not performed in view of the time to transplant. Two years and nine months after LVAD implantation, the patient finally underwent successful heart transplantation and was discharged without any complications.

Table 1
Changes in right and left ventricular function in echocardiogram.

Months after LVAD implantation	Pre-operation	1 month	5 months	12 months	28 months
LVDd (mm)	68	68	60	56	69
LVDs (mm)	63	65	56	52	68
LV ejection fraction (%)	16 %	11 %	17 %	16 %	2 %
RVEDA (cm^2)	48.0	26.2	28.5	31	33.7
RVESA (cm^2)	26.4	18.1	18.6	24.2	29.5
RVFAC (%)	45	30	35	22	10
Tricuspid regurgitation	Mild	Mild	Mild	Mild	Severe

LVDd, left ventricular end-diastolic diameter; LVDs, left ventricular end systolic diameter; LVAD, left ventricular assist device; RVEDA, right ventricular end-diastolic area; RVESA, right ventricular end-systolic area; RVFAC, right ventricular fractional area change.

Discussion

RVF remains a crucial complication after LVAD implantation. In this case, the pre-operative echocardiogram and catheterization showed no risk factors for early or late RVF. Left ventricular function seemed to improve with a decrease in BNP level after LVAD support. However, BNP levels increased again, and the right ventricular functional area change decreased with the VA events (Fig. 1). The clinical course of this patient indicates that RVF became worse, accompanied with the aggravation of VAs.

The incidence of VAs after LVAD implantation is high, ranging from 20 % to 50 % [6]. In general, VAs can be well tolerated in patients with LVAD support, but RV dysfunction potentially leads to inadequate perfusion and dysfunction of end-organs, as observed in this patient. In this case, recurrent VAs seemed to cause biventricular dilation and contractile dysfunction. Further, antiarrhythmic medications for VAs could have negative inotropic effect on RV function. Biventricular remodeling and worsening hemodynamics due to RVF reciprocally increased susceptibility to VAs. Progressive RVF and VAs could exacerbate each other, leading to a vicious cycle.

In addition, severe AI might contribute to progressive RVF. The development of moderate or severe AI during CF-LVAD support was estimated to be about 30 % by 3 years of support, and patients on longer LVAD support tend to demonstrate worse AI [7]. In the present patient, recurrent VAs could also decrease the frequency of aortic valve opening, leading to valve degeneration and subsequent development of AI. Further, worsening AI could also interfere with the reduction of LV end diastolic pressure, provoking frequent VAs [7]. Patients with pre-existing RV dysfunction were reported to poorly tolerate significant AI. Increasing RV afterload worsened RV function-provoked decompensation and finally caused severe RVF in the present patient.

In patients developing severe RVF after LVAD implantation, RV assist device (RVAD) implantation is indicated, but the mortality is still high (50 % to 60 % at 6 months) [8]. RVAD implantation has been reported to increase the risk of complications and end-organ dysfunction at implantation may worsen the prognosis [8]. In this case, RVAD implantation was not considered, because we managed to control his RVF with sedation and inotropic/antiarrhythmic medications.

This report demonstrates that RVF, VAs, and AI could have an adverse effect on each other in the context of long-term management of LVAD, which finally lead to decompensation of heart failure (Fig. 2) [9]. A thorough assessment of hemodynamics and optimization of inotropic/antiarrhythmic medications, pump speed, and fluid management should be attempted to break out of this vicious cycle. In addition, invasive strategy including AVR or catheterization ablation could be considered for patients refractory to conservative management [6,7]. Patients with LVAD as a destination therapy will increase, and longer periods of LVAD management will be required. We should recognize that these

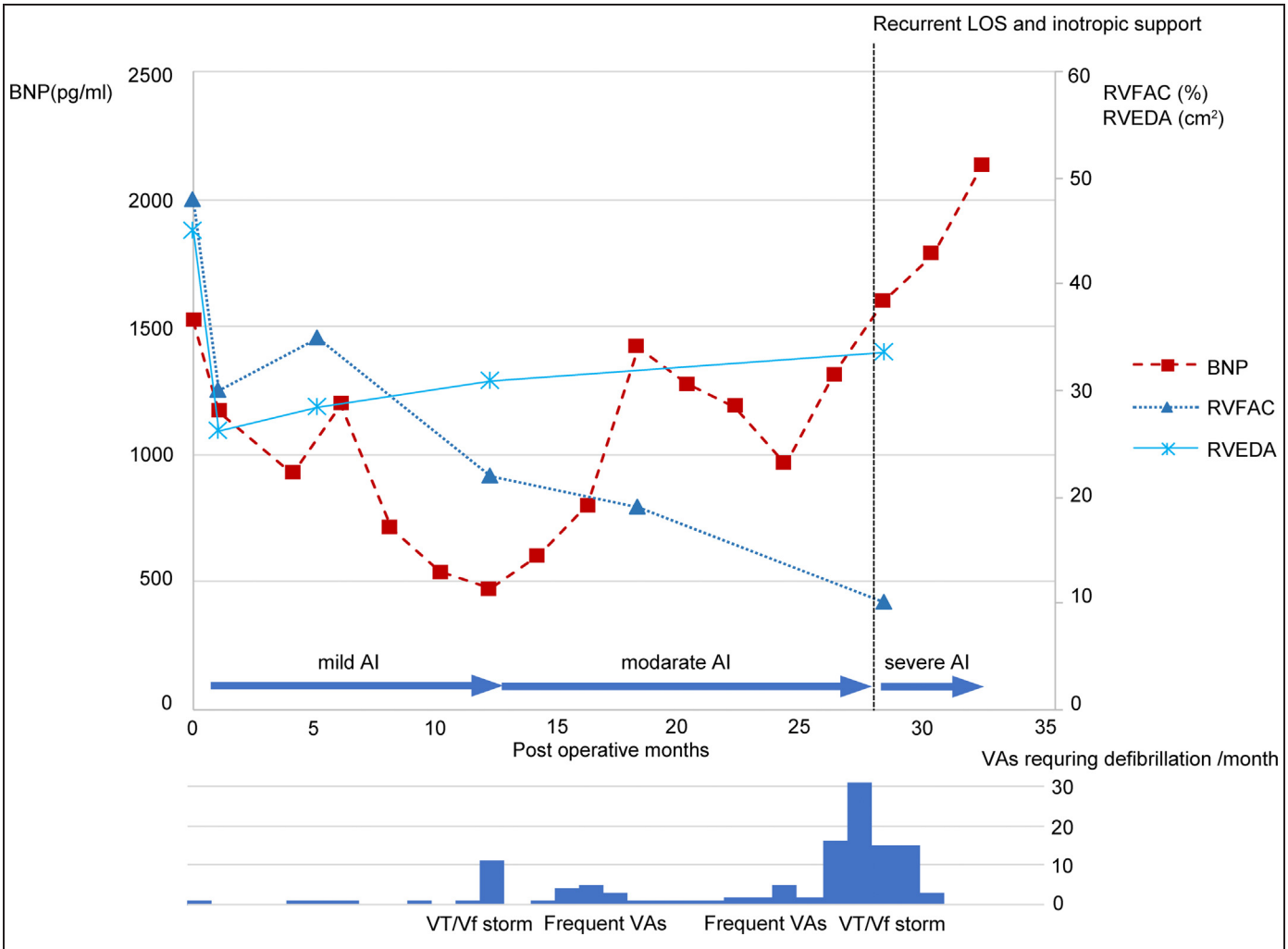

Fig. 1. Clinical course.
AI, aortic insufficiency; BNP, brain natriuretic peptide; LOS, low output syndrome; VA, ventricular arrhythmia; Vf, ventricular fibrillation; VT, ventricular tachycardia; RVFAC, right ventricular fractional area change; RVEDA, right ventricular end-diastolic area.

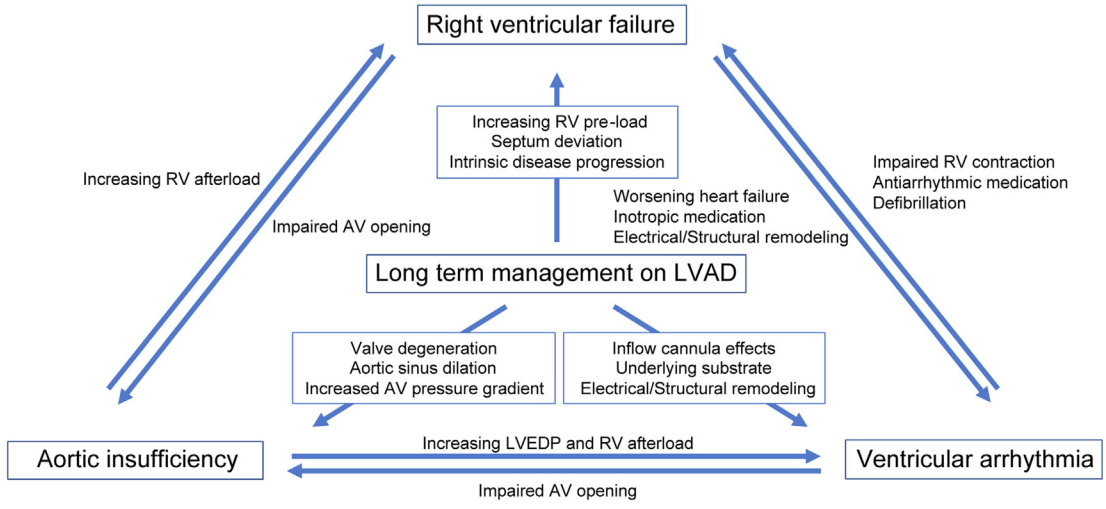

Fig. 2. The possible relationship between right ventricular failure (RVF), aortic insufficiency (AI), and ventricular arrhythmia (VA). RVF, VAs, and AI could have an adverse effect on each other in the context of the long-term management of LVAD. This vicious circle could finally lead to the progression of RVF. Reproduced, with permission, from Hatano et al. [9].
AV, aortic valve; RV, right ventricular; LVAD, left ventricular assist device; LVEDP, left ventricular end-diastolic pressure.

patients have the potential to develop RVF progression even years after implantation in association with recurrent VAs and AI.

Supplementary data to this article can be found online at https://doi.org/10.1016/j.jccase.2023.08.017.

Consent statement

Written informed consent was obtained from the patient.

Declaration of competing interest

The authors declare that there is no conflict of interest.

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