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Letter

Extreme Right Ventricular Pseudohypertrophy Due to Myocardial Edema

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INTRODUCTION

While some degree of right ventricular hypertrophy (RVH) is common in patients with long-standing pulmonary hypertension, extreme right ventricular (RV) wall thickening (RV free wall >10 mm) is a very unusual phenotype that is found only in a few disorders such as cardiac amyloidosis, hypertrophic cardiomyopathy (HCM), and certain congenital heart diseases.[1,2] Therefore, the presence of extreme RVH generally warrants further investigations and may signify a poor prognosis.

We present a patient who was admitted to the cardiac critical care unit after a prolonged cardiopulmonary resuscitation (CPR). Initial echocardiogram revealed biventricular hypertrophy with an RV free wall thickness of 13 mm. The combination of significant RV wall thickening on echocardiogram and low voltage on electrocardiogram (ECG) raised the suspicion for undiagnosed infiltrative cardiomyopathy. However, repeat echocardiogram several days later showed normalization of RV wall thickness to <5 mm. Therefore, we concluded that RV wall thickening was not due to true hypertrophy but rather significant myocardial edema from myocardial ischemia with associated reperfusion injury and mechanical trauma in the setting of prolonged CPR.

CASE

A 58-year-old male with a history of tobacco smoking and untreated hypertension presented with acute chest pain and shortness of breath of one day duration. He was found to have an ST-elevation myocardial infarction (STEMI) complicated by Killip class IV acute heart failure. He emergently underwent aspiration thrombectomy and intravascular ultrasound-guided percutaneous coronary intervention (PCI) to a 100% occluded right coronary artery (RCA) with 2 stents placed (**Figure 1**). He had residual non-obstructive disease involving distal left main, obtuse marginal, and diagonal branches. The procedure was complicated by cardiac arrest with incessant ventricular tachycardia (VT) storm, requiring multiple attempts of defibrillation and prolonged CPR lasting for over an hour, ultimately resulting in peripheral venoarterial extracorporeal membrane oxygenation (ECMO) insertion and Impella CP for left ventricular (LV) venting. He also required support with multiple vasopressors and inotropes including epinephrine, norepinephrine, vasopressin, and dobutamine.

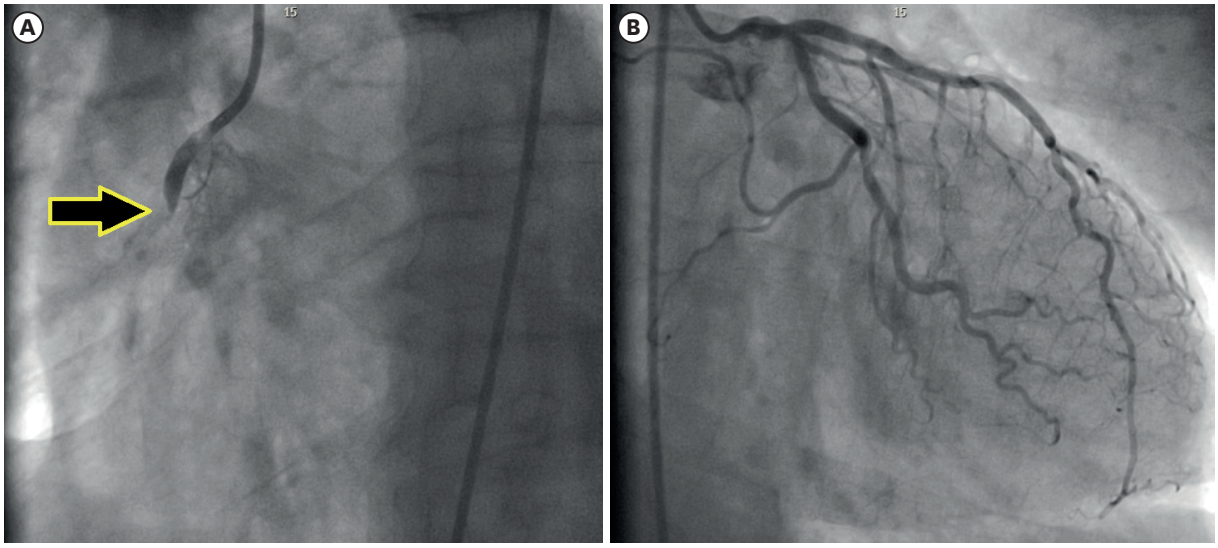

Figure 1. Coronary angiogram. (A) Acute thrombotic occlusion of right coronary artery (arrow). (B) Patent left coronary system.

Investigations

Laboratory workup post-PCI was remarkable for creatinine of 2 mg/dL, alanine aminotransferase 137 U/L, aspartate aminotransferase 758 U/L, lactic acid 14.2 mmol/L, and significantly elevated high sensitivity troponin-I over the detection limit of 229,730 ng/L up from 34 ng/L pre-PCI. Post-revascularization echocardiogram demonstrated an left ventricular ejection fraction of 15–20%, moderately reduced RV systolic function (tricuspid annular plane systolic excursion 1.1 cm), biventricular hypertrophy with extremely thickened RV free wall measuring 13 mm (**Supplementary Video 1**). RV global longitudinal strain (GLS) was −4%. ECG did not show signs of RVH, however, despite significant RV wall thickening on echocardiogram (**Figure 2**). Invasive hemodynamics prior to ECMO insertion showed a pulmonary artery pressure of 23/21 mmHg (mean 21 mmHg), and a pulmonary capillary wedge pressure of 14 mmHg. Post ECMO insertion, the pulmonary artery pressure and pulsatility improved to 30/16 mmHg with a central venous pressure of 6 mmHg, and RV pressure of 35/5 mmHg. Computed tomography angiogram of the chest showed a normal pulmonary artery diameter of 25 mm indicating lack of pre-existing pulmonary hypertension.

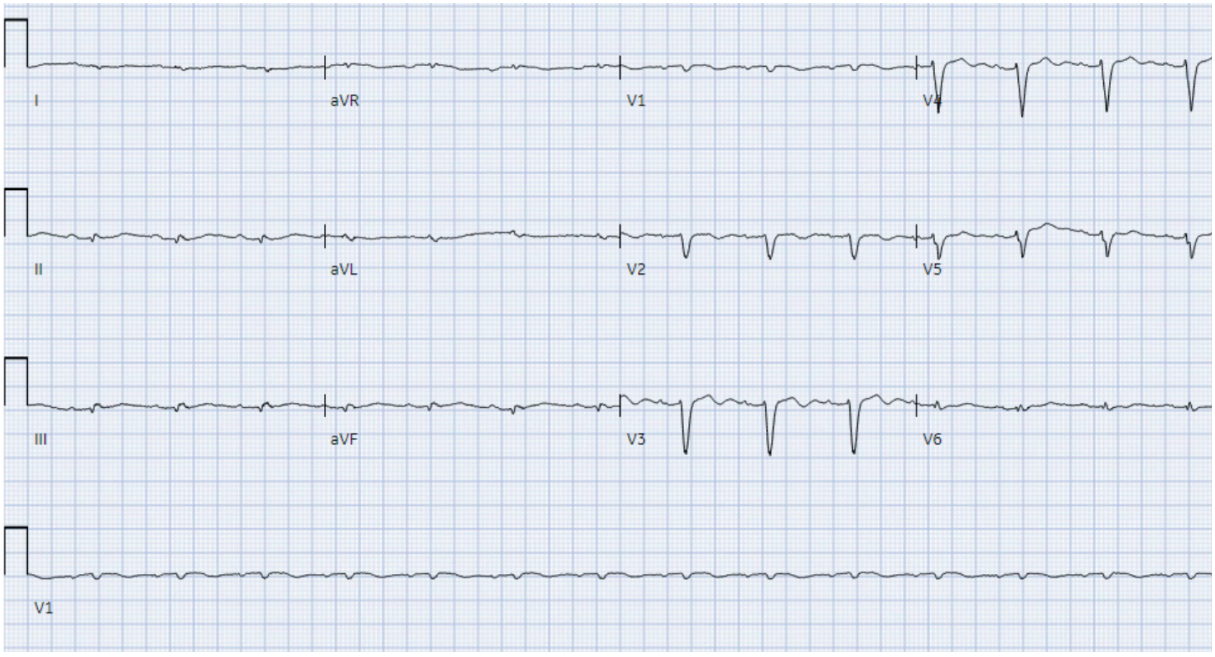

Figure 2. Post-revascularization electrocardiogram showing sinus rhythm, low voltage, with no signs of right ventricular hypertrophy.

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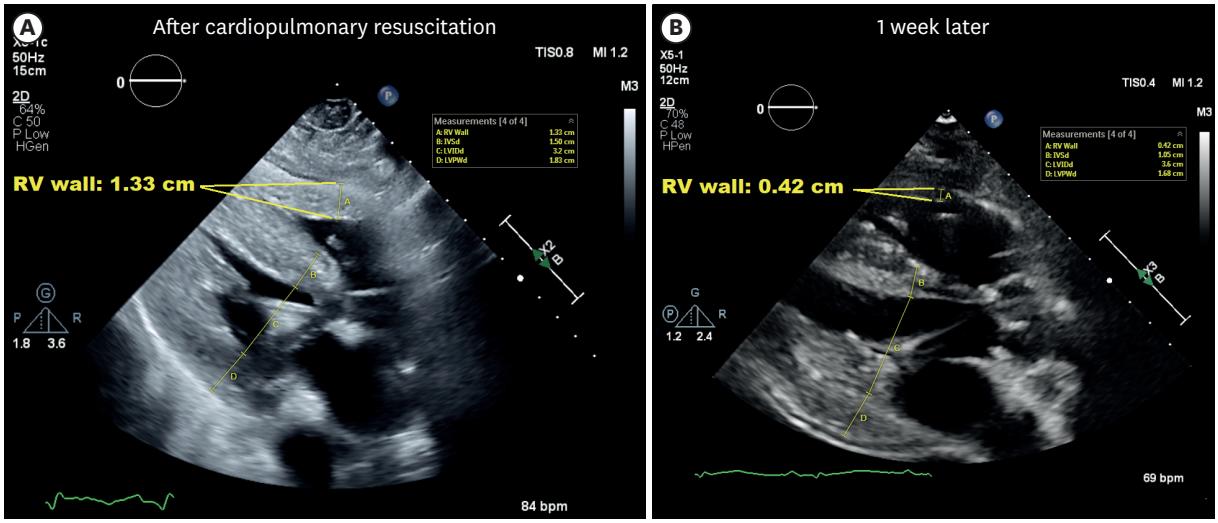

Figure 3. Transthoracic echocardiogram in parasternal long axis view showing right ventricular wall thickness. (A) After cardiopulmonary resuscitation and (B) 1 week later.

Outcome and follow-up

Repeat echocardiogram one week later showed significant improvement of RV wall thickness to <5 mm (**Figure 3** and **Supplementary Video 2**) with some improvement of biventricular function. RV GLS improved to –18%. While LV wall thickness improved, the patient had residual LV hypertrophy due to hypertensive heart disease from long-standing untreated systemic hypertension (**Supplementary Video 2**). Additionally, RV sustained more insults than LV in this case due to severe ischemia from occluded RCA as well as its anatomic location anteriorly thus receiving large mechanical impact from CPR. Resolution of RV wall thickening suggested a reversible process rather than a chronic systemic condition, and as such, we concluded that the patient had a better chance of recovery. Ultimately, he underwent successful ECMO decannulation on post-operative day 8, with a sustained recovery of RV function, and no recurrence of VT.

DISCUSSION

RVH, a pathologic increase in RV muscle mass, can result from chronically elevated RV afterload from elevated pulmonary pressure or anatomic RV outflow tract obstruction. The latter is mostly a congenital anomaly such as in tetralogy of Fallot or double outlet RV, certain genetic disorders (e.g., Noonan syndrome), or maternal rubella syndrome.[3] In the adult population, RVH is most commonly a manifestation of long-standing pulmonary hypertension. In these cases, RV free wall thickness is usually limited to 5–10 mm. However, extreme RV wall thickening (>10 mm) is found only in a few disorders including infiltrative diseases such as cardiac amyloidosis (especially TTR amyloidosis), HCM, and certain congenital heart diseases. Extreme RVH is found in only 1.3% of patients with HCM and is associated with increased mortality.[1,2,4] Therefore, understanding the underlying mechanism responsible for RVH is clinically important because various etiologies have different principles of management and imply dissimilar prognosis.

Infiltrative diseases such as cardiac amyloidosis, Danon disease, and Fabry disease are characterized by accumulation of extra or intra-cellular deposits resulting in biventricular wall thickening and loss of elasticity.[5,6] Therefore, when extreme RV wall thickening is identified (especially with unknown baseline), infiltrative cardiomyopathy (most commonly cardiac amyloidosis) is amongst the top differentials, and may indicate presence of an extensive systemic disease with a poorer chance of myocardial recovery. Such patients may not tolerate weaning off mechanical circulatory support (MCS) in the setting of cardiogenic shock, and may better be considered for cardiac transplantation. Moreover, patients with RV failure should be closely monitored with complete hemodynamic profiling to guide therapy. This is because physical examination alone can be highly inaccurate, and often underestimating the degree of RV failure leading to hypoperfusion and congestion.[7]

Although it is common to refer ventricular wall thickening as "hypertrophy," it is important to make the distinction between actual myocyte hypertrophy (true hypertrophy) and wall thickening due to the expansion of extracellular space (pseudohypertrophy). An example of true hypertrophy is the athletic heart which typically has normal diastolic function and global longitudinal strain (GLS). Conversely, pseudohypertrophy syndromes such as cardiac amyloidosis and longstanding renal disease typically result in diastolic dysfunction with restrictive filling pattern and

impaired GLS. While patients with hypertensive heart disease and severe aortic stenosis are theoretically expected to have true LV hypertrophy from increased LV afterload, these patients end up developing progressive interstitial fibrosis over time resulting in late Gadolinium enhancement (LGE) with expansion of extracellular volume (ECV) and elevated T1 values on cardiac MRI. Further studies investigating RV diastology, GLS, LGE and ECV may be of value to further understand the impact of different disease processes that lead to RV wall thickening.[8]

CPR is known to carry a risk of injuries such as sternal or rib fractures, pneumothorax, hemopericardium, or other visceral injuries due to mechanical forces exerted during compressions. However, RV edema has not previously been documented as potential sequelae of CPR. This is likely because CPR is often aborted in a much shorter time frame than in the present case. In our case, we concluded that RV wall thickening was due to myocardial edema due to prolonged CPR with repetitive trauma to the RV wall, multiple attempts of defibrillation for VT storm, with associated myocardial ischemia and reperfusion injury. We believe that mechanical injury to myocardium can result in reversible myocardial edema, myocyte injury, and interstitial hemorrhage in a process similar to blunt cardiac contusion.[9,10] Myocardial edema can hinder myocardial oxygen delivery further exacerbating ischemic injuries. These traumatic and ischemic insults resulted in transient myocardial inflammation and edema which improved over several days. In the current era of progressive utilization of MCS and extracorporeal CPR, identification of massive RV wall edema can have a significant impact on decision making while managing critically ill patients with cardiogenic shock.

In conclusion, prolonged CPR can result in significant RV wall swelling which may be mistaken for infiltrative diseases. Identifying such a reversible process can significantly impact decision making while managing critically ill patients. As such, these should be monitored with serial echocardiograms to evaluate for reversible etiologies and to further guide management.

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Conflict of Interest
The authors have no financial conflicts of interest.

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SUPPLEMENTARY MATERIALS

Supplementary Video 1
Transthoracic echocardiogram in parasternal long-axis view done 2 hours post-revascularization showing severe biventricular wall thickening and significant biventricular dysfunction. Note the Impella device in the visualized portion of the aortic root.

Supplementary Video 2
Transthoracic echocardiogram in parasternal long-axis view done 1 week after CPR showing normalization of RV wall thickness to <5 mm with some improvement of biventricular function. Persistent left ventricular hypertrophy is likely due to long-standing untreated systemic hypertension.

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