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Case Report

Mid-ventricular Obstruction in Hypertrophic Cardiomyopathy: A Case Report

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Abstract

Mid-ventricular hypertrophic obstructive cardiomyopathy (HOCM) is an uncommon type of hypertrophic cardiomyopathy (HCH). In this case study, we reported the case of a 48-year-male patient with HCH, he was admitted because of severe rheumatic MS in pulmonary edema. After initial treatment, decision was made to put the patients on a peripheral VA extracorporeal membrane oxygenation (ECMO). He recovered well by minimal invasive therapy under the support of ECMO. Thereafter, he was shifted from intensive care unit with a decreasing creatinine values, good urine output, and good left ventricular and right ventricular functions and released from hospital without any further complications.

Key words: Mid Ventricular Obstruction, Hypertrophic Cardiomyopathy, Systolic Anterior Motion Of Anterior Mitral Leaflet

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is characterized by the occurrence of abnormal left ventricular (LV) wall thickening that is typically asymmetrical and not exclusively explained by conditions such as hypertension, aortic valvular stenosis, or valvulopathies.[1,2] It is also linked with myocardial fiber disarray.[3,4] The LV hypertrophy is mainly affecting the interventricular septum. In adults, majority of HCM is caused by mutations of the cardiac sarcomere genes. It is inherited by autosomal dominant trait.[5] Genetic variation in at least eleven genes and mutations involving myosin heavy chain (MYH7) and myosin-binding protein C3 (MYBPC3) are most common in HCM.[6]

In majority of HCM patients, dynamic LV outflow tract (LVOT) obstruction occurs which is a classic pathophysiological characteristic of the disease.[7] Outflow obstruction is usually produced by mitral valve systolic anterior motion (SAM) and septal contact due to flow drag, also resulting in mitral regurgitation. Under resting conditions or during exercise (physiological provocation), the LV outflow obstruction (gradients ≥30–50 mmHg) is found in approximately 70% of HCM patients.[8] LVOT obstruction occurs mainly at the subaortic level, mostly due to SAM of the mitral valve.[1,2] SAM is formed by a pull effect in the occurrence of high-velocity LV ejection, and it is mainly accountable for concomitant mitral regurgitation due to partial leaflet apposition.[6,7] Moreover, in variant HCM cases, the obstruction to flow arises at the mid-ventricular level, unlike to SAM. Mid-ventricular is mainly caused by contact of septal hypertrophy with a hypercontractile anterolateral LV wall (interposition of anterolateral papillary muscle and hypertrophic longitudinal muscle bands on the posterolateral wall of the LV).

Mid-ventricular hypertrophic obstructive cardiomyopathy (HOCM) is a rare variety of HCM, mostly overlooked. HOCM is characterized by asymmetric LV hypertrophy and by a pressure gradient between basal and apical sites in the left ventricle. Mid-ventricular HOCM in association with an apical aneurysm has rarely been reported. This can remain occult and manifest only when a trigger worsens it. HOCM is a big challenge for clinician, once it is linked with severe symptoms and unstandardized management and treatments. This case study is an attempt to address this matter; therefore, we conducted a case report of mid-ventricular obstruction in HCM patients. Our case conference describes such an example where occult HOCM became evident with high

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gradients in the post-operative period triggered by worsening of the right ventricular (RV) dysfunction.

CASE DESCRIPTION

A middle-aged person of 48 years old, known case of rheumatic heart disease (RHD), had undergone AVR in 1998 for severe AS and AR. Nine years down the line, he had stuck aortic valve prosthesis, got lysed successfully. Now, he complained of mild dyspnea and frequent palpitations. He presented with severe rheumatic MS, in pulmonary edema. Transthoracic echocardiogram (TTE) showed severe MS, dilated LA, good LV function, no PAH, RV basal TDI 6.8, and mild TR. He was taken up for MVR. Induction with etomidate, fentanyl, and vecuronium was uneventful. Re-do sternotomy was uneventful. The echocardiogram (ECG) showed asymmetric hypertrophy of the LV, MVR with a 23 mm OnX valve, clamp time 90 min, and pump time 120 min. He had one e/o VT during rewarming, was defibrillated with 50 J came off CPB on dobutamine 5 mcg/kg/min. Furthermore, TEE post-CPB showed good LV systolic function, dysfunctional RV with TAPSE 15, FAC was 18%, trivial TR, prosthetic mitral valve functioning well, mean gradient 3, DVI 1.1, and aortic valve prosthesis functioning well.

Moreover, the patient was wheeled into ICU with a cardiac index of >2, lactates of 5, and P/F ratio 400, on mechanical ventilation sedated. After 24 h, the patient experienced rapid breathing (tachypnea) with rise in airway pressures, also fall in urine output and rise in potassium levels. However, the cardiac index (CI) was found to be relatively stable around 1.8–2.00, the sudden increase in CVP from 7 to 23, with mild PAH. About 22% of MVO_2 was observed at this time. Furthermore, the systemic pressures dropped, along with fall in SVR. Transthoracic echocardiogram (TTE) further showed good LV function but dysfunctional RV and an LV mid-ventricular gradient of 120 mmHg (Figure 1) and dysfunctional Right ventricle (Figure 1). At that moment, to maintain systemic pressures, noradrenaline and vasopressin infusions were administered. In view of the MVO, dobutamine was discontinued for its inotropic action that would worsen the obstruction. Then, iNO was initiated to recover and improve RV forward output. With iNO, the airway pressures stabilized and CVP came down to ~10–11. Thereafter, the patient was carefully filled using crystalloid solutions. MVO_2 saturation improved further to 40%. Then, inhaled nitric oxide (iNO) was weaned due to rise in metHb and was bridged with Sildenafil (tab). Following day, the patient went into oliguria, and RV dysfunction further worsened dramatically, with a fall in P/F ratios up to <100. Although small dose of adrenalin was initiated for the RV inotropy, there was not much positive response

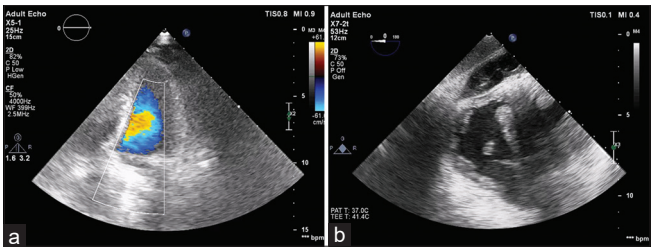

Figure 1: (a) Mid-ventricular turbulence. (b) Empty left ventricular with dysfunctional right ventricular

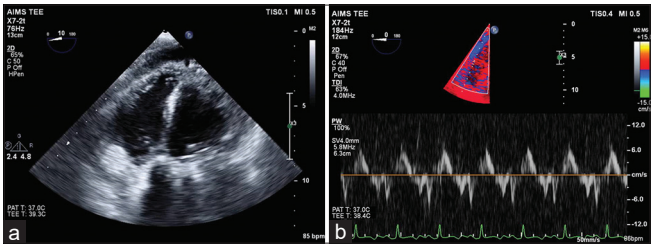

Figure 2: After 4 days of extracorporeal membrane oxygenation (ECMO) support. (a) Post-ECMO weaning, (b) post-ECMO right ventricular (RV) function better: Peak velocity TDI- RV lateral wall ≈8

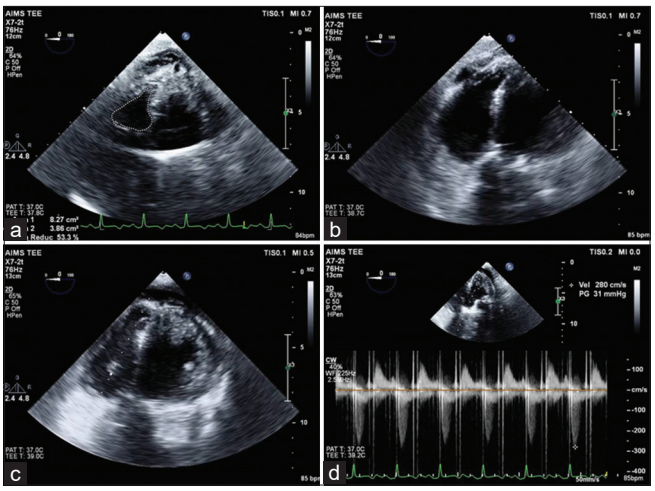

Figure 3: (a) FAC improved, (b) right ventricular (RV) contractility longitudinal excursion, (c) RV contraction short axis, and (d) RV function better, mid left ventricular gradient disappeared

and in that circumstances, decision was made to put the patients on a peripheral VA extracorporeal membrane oxygenation (ECMO). Moreover, femoral vein and artery were cannulated under TEE guidance with a limb perfusion cannula and ECMO was initiated. Perfusion pressures were maintained at `60 mean, and vasopressin was tapered off. Furthermore, UF was done using the ECMO circuit. ECMO was discontinued on the 4th day. During weaning off ECMO, a minimal dose of adrenalin and noradrenalin was maintained. TEE revealed fairly good RV function and LV function, with minimal turbulence but no mid ventricular gradient (Figures 2 and 3).

There was a transient rise in CVP after ECMO decannulation but it settled with a hike up of adrenalin dose and fluid removal with CRRT. Patient was tracheostomized and subsequently over the 2 days was weaned off inotropes and mechanical ventilation. By 2 weeks, parent renal parameters started improving and HD could be made less frequent. He had grown *Klebsiella* and *Enterococcus* from blood, was treated with sensitive antibiotics. The patient was then shifted to the ward on POD 25 with a decreasing creatinine values, good urine output, and good LV and RV functions.

DISCUSSION

Mid-ventricular HOCM is an uncommon type of HCH that is frequently accompany by the apical aneurysm.[9] Prior study suggested that patients with a history of HCM and mid-ventricular obstruction, had great chance of sudden death.[10] The exact pathogenesis of HOCM remains unclear, however, it had been demonstrated that apical aneurysm might be resulting to the enlargement after increase of apical pressure from the mid-ventricular obstruction seen in the degenerative process of hypertrophic HCH.[11] Other causes of apical aneurysm development are small-vessel disease with reduced coronary flow reserve, strained of coronary artery because of the increased wall pressure in the hypertrophic myocardial part, reduced coronary perfusion pressure due to mid-ventricular obstruction, and decreased capillary myocardial fiber ratio and coronary artery spasm.[11,12] Since the patient exhibited dysfunctional RV in the severe stage of his illness, we assume that the apical aneurysm has its beginning in a severe coronary artery episode, for instance, coronary artery compression or coronary microcirculation due to elevated pressure overload and systolic myocardial wall pressure linked with an intensely occurred mid-ventricular obstruction.

Appropriate management and treatment of HOCM is ambiguous, however, failure to intrude may result in lethal ventricular arrhythmias and death. In HOCM condition, β-blockers are the initial choice of treatment,[13] however, the management for mid-ventricular HOCM has still unknown. The presented case was considered as drug-resistant mid-ventricular HOCM since even after initiation of two medicines, the LVOT pressure gradient was found abnormal, and thereafter, ECMO support was administered. Thus, the patient was suitable candidate for minimal invasive therapy, in view of his age and responses.

CONCLUSION

Mid ventricular obstruction is a rare variant Hypertrophic cardiomyopathy and awareness of this mid ventricular obstruction and management can avoid catastrophe.

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