

Hypertrophic Obstructive Cardiomyopathy and Cardiogenic Shock: A Challenging Hemodynamic Situation

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Abstract: *Hypertrophic cardiomyopathy is a genetic disease due to a mutation in cardiac muscle protein resulting in left ventricular wall and septal hypertrophy. HOCM is characterized by an excessively thick interventricular septum, which obstructs left ventricular outflow during ventricular systole. It is quite for a patient with hypertrophic cardiomyopathy to present in a state of cardiogenic shock, except in the context of major factors of decompensation leading to disruption of balance between preload and after load. When it is the case, the whole difficulty is to know the hemodynamic specifics of this pathology. Here, we report a case of 50-years-old female admitted for hypertrophic obstructive cardiomyopathy complicated by critical cardiogenic shock. This case illustrates the challenges of managing such complex hemodynamic situation.*

Keywords: hypertrophic cardiomyopathy, left ventricle outflow tract obstruction, cardiogenic shock

1. Introduction

Hypertrophic obstructive cardiomyopathy (HOCM) can lead to left ventricular outflow tract obstruction and cardiogenic shock, only few prior cases have been reported. In this article, we present a case of cardiogenic shock in patient with HOCM illustrating the importance of a well understanding of pathophysiologic process in order to better manage this difficulty of this situation.

2. Case Report

This is a case of a 55 year old female known to have hypertension, suffering from progressive shortness of breath since 2 month, she became severely dyspnoeic two days before, she was transferred from a peripheral hospital to our unit because of acute pulmonary edema, she received intravenous furosemide and was on Dobutamine infusion from the referring hospital. At her admission, she was in a real cardiogenic shock that required endotracheal intubation. She was directly hospitalized in reanimation, ECG showed sinus tachycardia and left ventricular hypertrophy (LVH) with strain pattern. Physical exam revealed elevated jugular venous pressure, bilateral leg edema, bilateral lung crackles throughout the lung field and a loud systolic murmur at the left sternal edge. Laboratory values revealed a severe metabolic acidosis (lactate >15, pH 7.03), and over the next 12 hours, end-organ function worsened with a creatinine 24 g/ml, AST 3130 UI/l, ALT 2770 UI/l. Her WBC count and CRP were elevated, She had massive pulmonary vascular congestion on Chest X-ray and her Transthoracic Echocardiography demonstrated severe concentric LVH with asymmetrical septal hypertrophy (basal septal thickness = 22 mm), severe left ventricular outflow tract (LVOT) obstruction with peak gradient of 150 mmHg (Fig.1), small LV cavity and systolic anterior motion (SAM) of mitral valve with moderate MR. LV systolic function was normal with impaired LV relaxation.

During this time, Phenylephrine was started but the patient remained resistant to treatment. In order to relieve the dynamic LVOT obstruction, an intravenous bolus of esmolol was administered and within 20 min there was rapid improvement in the patient's symptoms. Real-time echocardiography demonstrated a dramatic reduction in the outflow tract gradient to 70 mmHg (Fig. 2). An esmolol infusion was continued for a further 6 h and then replaced with oral beta-blocker therapy after extubation. Subsequently, Patient had a protracted course of illness due to cardiogenic shock leading to multi-organ involvement. She was proposed for percutaneous septal alcohol ablation. Her Coronary angiogram was normal, she was discharged with propranolol 160 mg twice a day.

3. Discussion

Hypertrophic cardiomyopathy (HCM), the most common monogenic cardiovascular disorder, is diverse in presentation and natural history, frequently misunderstood, and often under recognized in clinical practice [1-3]. In patients with HOCM, systolic septal bulging into the LVOT, malposition of the anterior papillary muscle, drag forces, and hyperdynamic LV contraction (causing the Venturi effect) may contribute to creation of the LVOT gradient[4]. LVOT obstruction in HOCM is dynamic and the degree of obstruction depends more on cardiac contractility and loading conditions. Decrease in LV preload and augmentation of myocardial contractility increase the LVOT obstruction. Usual inotropes and vaso pressors are contraindicated in such situation with the exception of Phenylephrine pure α -agonists that reduced obstruction by after load augmentation [5].

The current case illustrate the critical management dilemma existing in patient with HOCM presenting with cardiogenic shock. Conventional therapy directed at hypotension and pulmonary edema used in peripheral hospital (positive inotropic agents and diuretics) had precipitated hemodynamic deterioration of the patient by increasing

contractility and reducing preload which contribute to worsening of left ventricular obstruction.

On the contrary, the use of esmolol, a short-acting beta-blocker, associated with phenylephrine reversed the gradient leading to resolution of the patient's symptoms. Esmolol has been reported to be of beneficial use in cardiac emergencies related to hypertrophic cardiomyopathy [6, 7].

Concerning invasive treatment, myectomy is considered first-line therapy for medically refractory HCM with LVOT obstruction. This is because of its very low perioperative mortality, higher success rates, lower risk for PPM implantation, and availability of long-term efficacy data. Myectomy should be performed in younger patients (especially in those ≤ 40 years old), patients with massive hypertrophy, and those with concomitant valvular or other cardiac disease. Alcohol septal ablation is ideally for patients who are not surgical candidates (elderly individuals). Pacing should only be considered if a patient has an existing device or has refractory symptoms with LVOT obstruction and is not a candidate for septal reduction therapy [8]. In case of hypertrophic obstructive cardiomyopathy (gradient, ≥ 50 mm Hg), the patient should be directed at time to invasive treatment before hemodynamic deterioration.

4. Conclusion

The use of vasodilators and positive inotropes in cardiogenic shock with hypertrophic obstructive cardiomyopathy can be more life-threatening. The mainstay of therapy consist of IV β -blockers and phenylephrine.

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Figure legend:

Figure 1: Apical five-chamber view demonstrating
A: The asymmetrical hypertrophy of interventricular septal (maximum thickness is 22mm).
B: RVOT obstruction determined by colour flow mapping.

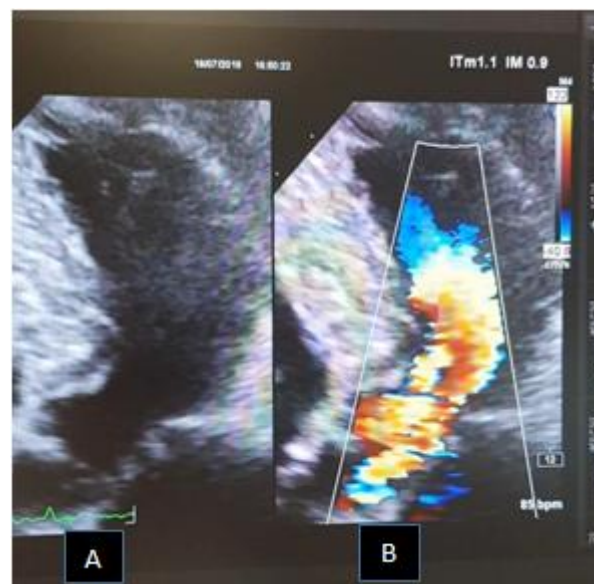


Figure 1



Figure 2