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Refractory cardiogenic shock in hypertrophic cardiomyopathy complicated by apical ballooning: A case report

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Abstract

Hypertrophic cardiomyopathy can be accompanied by dynamic obstruction in the left ventricular outflow tract and acute apical ballooning, which are among the very rare causes of cardiogenic shock. This condition requires a specific treatment approach that in many ways differs from the treatment of other causes of cardiogenic shock. We present a case and our treatment strategy (including extracorporeal life support) for refractory cardiogenic shock in a patient with previously undiagnosed hypertrophic cardiomyopathy.

Keywords

Cardiogenic shock, hypertrophic cardiomyopathy, Takotsubo syndrome, left ventricular outflow tract obstruction, extracorporeal membrane oxygenation, mechanical circulatory support, acute heart failure, case report

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Introduction

Hypertrophic cardiomyopathy (HCM) is a genetically determined heart muscle disease characterized by left ventricular hypertrophy with a wide range of clinical manifestations. A small subgroup of patients with HCM presents acutely with severe obstruction in the left ventricular outflow tract

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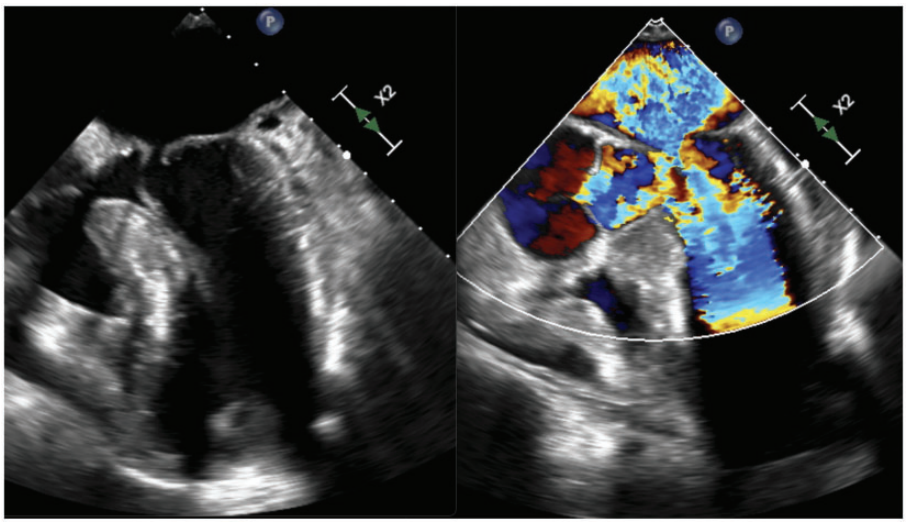

Figure 1. Transesophageal echocardiography: systolic anterior motion of the elongated anterior mitral valve leaflet, resulting in obstruction of the left ventricular outflow tract (left panel) and significant mitral regurgitation (right panel).

obstruction (LVOTO) and apical ballooning (AB), which can eventually progress to severe heart failure or cardiogenic shock (CS).[1,2] This subset of patients shares several epidemiologic, clinical and echocardiographic features with Takotsubo syndrome (TTS).[2–4] The treatment approach in patients with CS caused by LVOTO differs from that in patients with CS caused by other conditions. We present a case of a refractory CS in a patient with previously undiagnosed HCM, treated with extracorporeal life support.

Case report

A woman in her late 60s with no significant medical history was urgently admitted to our department due to severe hemodynamic instability at the end of the elective neurosurgical procedure (thoracic and lumbar spine stabilization after a fall). The procedure was conducted in a prone position and was complicated by significant blood loss. Despite appropriate perioperative management, including transfusions and fluid resuscitation, the patient's hemodynamic status progressively deteriorated, with necessitating escalating doses of vasopressor (norepinephrine dose, 0.5–1 μg/kg/min). Electrocardiography (ECG) revealed diffuse ST-segment elevation. Rapid bedside transthoracic echocardiography (TTE) and transoesophageal echocardiography demonstrated preserved ejection fraction (EF) in an asymmetrically hypertrophic left ventricle (LV) without regional wall motion abnormalities but with severe mitral regurgitation (MR) due to systolic anterior motion (SAM) of the anterior leaflet and dynamic LVOTO (Figure 1). Initial management of the shock included volume resuscitation and vasopressors (argipressin), guided by pulmonary artery catheter monitoring. However, despite these interventions, the patient's conditions continued to deteriorate, with worsening left ventricular function. Repeat TTE revealed AB of the LV with a reduced EF of approximately 15%. Cardiac output (CO) was measured at 2 L/min, cardiac index (CI) at 1 L/min/m^2 and heart rate at 90/min, and lactate levels

were elevated at 5.7 mmol/L. The mean arterial pressure was approximately 50 mm Hg. Given the refractory nature of the shock, the decision was made to escalate treatment with venoarterial extracorporeal membrane oxygenation (V-A ECMO). Extracorporeal membrane oxygenation (ECMO) cannulation was performed percutaneously via the femoral vessels (21-Fr venous and 19-Fr arterial cannula with a 7-Fr distal perfusion catheter), with the pump flow set to 4 L/min. Owing to the recent extensive neurosurgical procedure, in addition to an initial bolus, ECMO was run without continuous heparin infusion during the first 24 h. Coronary angiography performed during ECMO implantation ruled out obstructive coronary artery disease (Figure 2).

The patient's hemodynamic status improved rapidly with ECMO support, allowing for the gradual weaning of vasopressors and inotropes. Subsequent TTE showed regression of SAM and MR, with progressive improvement in left ventricular EF. The dynamic LVOTO was effectively managed with a continuous infusion of the β-blocker metoprolol (0.2 μg/kg/min). Both CO and CI normalized, and ECMO support was successfully discontinued on the third day without complications. The patient was discharged from the intensive care unit on the 14th day. A follow-up TTE showed normalized left ventricular function without regional wall motion abnormalities but with persistent severe dynamic LVOTO (peak gradient approximately 200 mmHg) provoked by a small dose of nitroglycerin (Figure 3). Septal reduction therapy (SRT) was discussed and offered to the patient, but she declined. The patient was discharged home on β-blockers and has been followed up annually at our center. She subsequently underwent orthopedic surgery without any complications and remains asymptomatic, with no evidence of obstruction (at rest or provoked) on follow-up TTE, and has preserved left ventricular function.

Discussion

LVOTO is a characteristic feature of HCM, present in approximately 70% of cases. LVOTO has a complex pathophysiology. It consists of SAM of the anterior mitral leaflet caused by Venturi effects or flow drag forces in a narrowed left ventricular

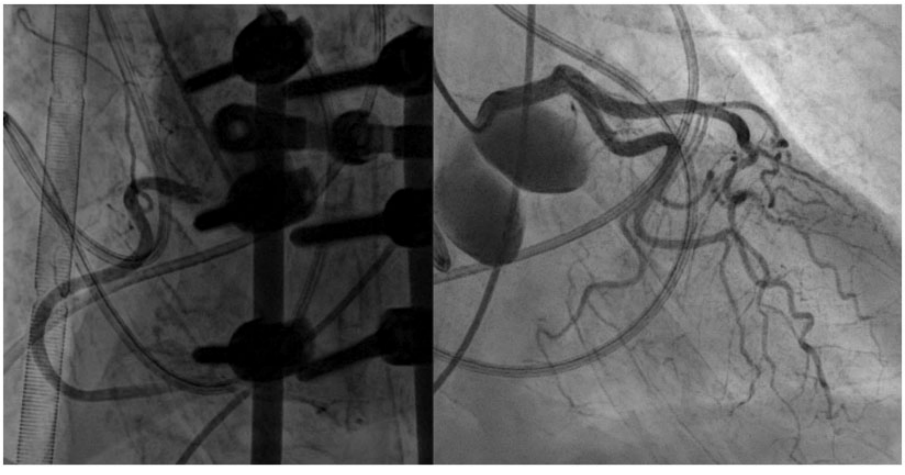

Figure 2. Selective coronary angiography: demonstration of nonobstructive coronary artery disease.

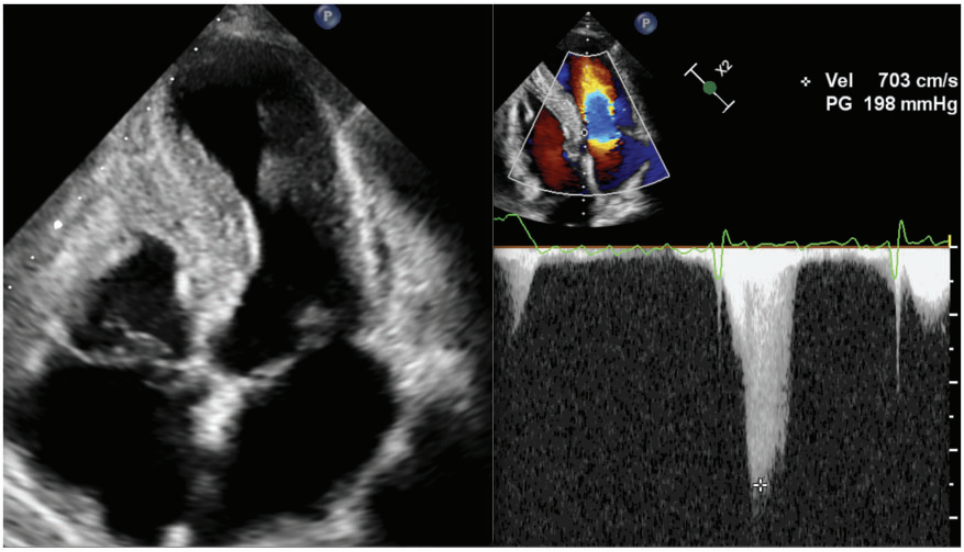

Figure 3. Transthoracic echocardiography: asymmetrical hypertrophy of the left ventricle (16 mm at the basal septum), with severe provoked obstruction in the left ventricular outflow tract following administration of a low dose of nitroglycerin.

outflow tract. LVOTO is notably dynamic, being dependent on preload and afterload conditions.[1] TTS was initially considered among the differential diagnoses in our case. Although LVOTO can occur in TTS, it is less common, being present in approximately 20% of cases.[4] Conversely, AB, the most common variant of TTS, is occasionally observed in patients with HCM (1%) with acute LVOTO, often triggered by a rapid decrease in preload or afterload, in line with the pathophysiologic concept of supply-demand ischemia (elevated left ventricular pressures, increased left ventricular work and reduced diastolic coronary pressure), afterload mismatch and microvascular dysfunction.[2,5] The pathogenesis of AB in TTS is believed to involve catecholamine excess and microcirculatory dysfunction, typically in the context of physical or emotional stressors.[4] An overlap between HCM and TTS is possible in one small, unselected group of patients; approximately 30% of those with TTS may exhibit phenotypic features of obstructive HCM.[6]

Patients with HCM and AB share several characteristics with TTS, including epidemiologic (predominantly postmenopausal women in their 60s), clinical (presentation with chest pain, dyspnea and syncope), ECG (ST-segment changes), echocardiographic (apical hypokinesis with hypercontractile basal left ventricular segments) and coronary angiographic (nonobstructive coronary artery disease) features.[2] A detailed echocardiographic assessment is essential for distinguishing HCM with AB from TTS. This includes measurement of left ventricular wall thickness, assessment of LVOTO and evaluation of the mitral valve (including leaflet elongation), as these abnormalities often persist beyond the acute phase. The clinical course can also aid in diagnosis; in cases of HCM with AB, relief of LVOTO often leads to rapid improvement in shock and resolution of left ventricular ballooning.[2,5]

Management of CS due to LVOTO with or without AB is primarily guided by observational data and expert consensus. Initial

management should focus on optimizing contributing factors such as preload (e.g. volume optimization, blood transfusion for anemia) and rhythm control (e.g. cardioversion for tachyarrhythmias). Vasopressors, including norepinephrine and vasopressin, are often required to maintain perfusion pressure, ideally at the lowest effective doses, and may positively impact LVOTO by increasing afterload. In contrast, inotropes, vasodilators and diuretics should generally be avoided as they can exacerbate dynamic obstruction through increased basal segment hypercontractility and decreased preload or afterload, potentially worsening shock.

Paradoxically, β-blockers (e.g. esmolol, landiolol, metoprolol) are central to the management of LVOTO, provided the patient is not in fully developed shock or experiencing deteriorating hemodynamics, as they can effectively reduce the LVOTO gradient. Mechanical circulatory support (MCS) should be considered if CS cannot be controlled through conservative measures. Intra-aortic balloon pumps should be avoided due to their potential to reduce left ventricular afterload and exacerbate LVOTO. Percutaneous ventricular assist devices can provide hemodynamic stabilization, but by reducing left ventricular preload, they may also worsen LVOTO. V-A ECMO is often the preferred MCS for this type of CS, as LVOTO can mitigate the negative impact of V-A ECMO on left ventricular performance.[7] SRT, such as alcohol septal ablation or myectomy, represent definitive treatments for LVOTO and should be considered in cases of refractory shock at specialized centers.[2,5] The reporting of this study conforms to CARE guidelines,[8] and all patient information was deidentified.

Conclusion

This case of CS secondary to LVOTO and AB in a patient with previously undiagnosed HCM underscores the critical importance of early diagnosis in this rare but potentially catastrophic condition. HCM with AB caused by severe LVOTO necessitates a tailored therapeutic approach distinct from other causes of CS. In our patient with refractory CS, the implantation of V-A ECMO was crucial in enabling effective relief of LVOTO with β-blockers, which constitute the cornerstone of treatment.

Author contributions

MS: wrote and edited the original draft. PT and JK: visualized and conceptualized the study. RP, PT and JK: supervised the study. All authors discussed the results and contributed to the final manuscript.

Data availability

No data sets were generated or analyzed during this study.

Declaration of conflicting interest

The authors declare that there is no conflict of interest.

Ethics statement

This case report did not require approval by the ethics committee at our institution. Verbal informed consent for publication was obtained from the patient and all information was deidentified. Our case report followed CARE guidelines.

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