Summary

Hypertrophic obstructive cardiomyopathy (HOCM) is a rare genetic disorder characterized by left ventricular outflow tract (LVOT) obstruction. Clinical presentation ranges from absence of symptoms to sudden death. Our 60 year old patient scheduled for left modified radical mastectomy had HOCM since seventeen years with severe LVOT obstruction and mitral regurgitation. An implantable cardioverter defibrillator (ICD) and permanent pacemaker (PPM) was inserted 15 months earlier for ventricular tachycardia. Anesthetic management of these patients presents considerable challenges and requires maintenance of desired hemodynamic parameters and management of specific complications. Factors like tachycardia, hypovolemia, vasodilation and increased cardiac contractility leads to exacerbation of the obstruction. In our patient there was the additional consideration of the ICD which required to be turned off during surgery with full provision for external defibrillation. We managed to successfully maintain the desired hemodynamics throughout the surgery and the patient was discharged home on the seventh postoperative day.

Introduction

Hypertrophic obstructive cardiomyopathy (HOCM) is a rare disorder characterized by massive asymmetric hypertrophy of the myocardium resulting in left ventricular outflow tract (LVOT) obstruction\textsuperscript{1}. Clinical presentation ranges from absence of symptoms to sudden unexpected death\textsuperscript{2}. Decrease in venous return and systemic vascular resistance or increase in myocardial contractility worsens the LVOT obstruction\textsuperscript{3}. These patients are highly prone to arrhythmias like atrial fibrillation and ventricular tachycardia (VT)\textsuperscript{2}. Management of anesthesia in these patients poses considerable challenges for the anesthesiologist. We report successful conduct of anesthesia in a patient with HOCM undergoing modified radical mastectomy (MRM).
Case Report

A 60 year old female was scheduled for MRM for carcinoma of the left breast. She was a known case of HOCM for 17 years. Past medical history revealed hypertension and diabetes since six years. An implantable cardioverter defibrillator (ICD) and permanent pacemaker (PPM) was inserted 15 months earlier when she had presented with VT at our tertiary care university hospital. She was taking atenolol, amlodipine, aspirin and soluble insulin. She complained of breathlessness on climbing a flight of stairs and had an ejection systolic murmur in the mitral area. The routine laboratory investigations were within normal limits. Her echocardiogram showed gross concentric left ventricular hypertrophy with a Grade II diastolic dysfunction. There was also systolic anterior motion of anterior mitral leaflet resulting in severe LVOT obstruction with a peak gradient of 130 mmHg and severe mitral regurgitation.

The patient was premedicated with tablet midazolam 7.5 mg. The pacemaker was altered to a fixed mode at 70 b/minute and the ICD was turned off before taking the patient to OR. External defibrillator with electrocardiogram (ECG) was accompanied with the patient. After initiating monitoring with ECG lead II, non-invasive blood pressure, peripheral oxygen saturation and insertion of peripheral venous line, an arterial catheter was placed in the right radial artery under local anesthesia. General anesthesia was induced with fentanyl, etomidate and vecuronium and a size 7.0 endotracheal tube was inserted smoothly. Intermittent positive pressure ventilation with 50% oxygen in air and isoflurane adjusted to 1.2 MAC with fentanyl and vecuronium boluses were used for maintenance of anesthesia. After induction a 7.5 French Swan sheath was inserted in right internal jugular vein for monitoring of central venous pressure (CVP) and a provision for floating a pulmonary artery catheter if there was a need. Sterile external pads for defibrillation and pacing were placed after ensuring that they were kept away from the surgical field and the ICD.

The procedure lasted three hours during which there was one episode of hypotension ten minutes after induction, which responded to a 200 ml bolus of normal saline and 100 micrograms phenylephrine. Thereafter she remained hemodynamically stable and her CVP was kept at 12-13 cmH₂O. She was extubated at the end of the procedure after reversal of the neuromuscular block and was transferred to the coronary care unit (CCU) where the ICD was switched on and PPM mode was reverted to the preoperative status. Patient remained stable hemodynamically except for an episode of supraventricular tachycardia (SVT) on the first postoperative day, which was reverted by the ICD without any additional measures. She was discharged home on the seventh postoperative day.

Discussion

HOCM is a rare genetic disorder characterized by massive asymmetric myocardial hypertrophy. Systolic anterior motion of the mitral valve leads to LVOT obstruction and often precipitates mitral regurgitation. Diastolic dysfunction occurs due to impaired ventricular compliance. All these features were present in our patient. Factors such as tachycardia, hypovolaemia, vasodilation and increased cardiac contractility exacerbate the obstruction. Anesthetic management entails maintenance of desired hemodynamic parameters, and management of specific complications like hypotension, dysrhythmias and congestive heart failure.

Sinus rhythm is crucial in these patients because of the dependence of preload on atrial contraction. The synchronous contractile pattern induced by pacing may be therapeutic as pacing has been shown to be effective in reducing the pressure gradient between the left ventricle and the aorta. In our patient the presence of dual chamber pacemaker was probably helpful in maintaining the hemodynamic state. The ICD needed to be inactivated during surgery to prevent its damage by cautery. It is important to arrange its prompt activation in the CCU. External pads were placed for delivery of shock if required. Our patient had a severe LVOT obstruction with peak gradient of 130 mmHg. Negative inotropic drugs like beta blockers and calcium channel blockers are used to decrease the degree of outflow tract obstruction. Our patient was on both these agents which were continued perioperatively, while intraoperatively the dose dependent myocardial depression caused by inhalation anesthetics might have helped in this regards.

These patients should be given adequate
premedication to reduce stress. Avoidance of vasodilators and agents that increase contractility is essential during the anesthetic management\textsuperscript{2,9}. Episodes of hypotension can be treated with volume replacement and/or vasoconstrictors like norepinephrine or phenylephrine. Our patient responded well to fluids and phenylephrine. We used IV fluids judiciously and kept the CVP at 12-13 cm of H\textsubscript{2}O as adequate preload is necessary to maintain optimal cardiac output and avoid undue increase in contractility because of hypovolemia\textsuperscript{9}. Although CVP may be an inaccurate guide to filling pressures due to abnormalities in left ventricular compliance, we decided to monitor CVP with a provision of floating a pulmonary artery catheter if required as we were not expecting major fluid shifts during an MRM procedure.

**Conclusion**

HOCM is a rare disorder imposing challenging conditions for the anesthesiologist. Thorough understanding of the pathophysiology of the condition is mandatory. It is important to maintain adequate preload and afterload, prevent arrhythmias and minimize outflow obstruction. In our patient there was the additional consideration of the ICD which required to be turned off during the surgery.
References


