CASE IN POINT

Importance of Recognizing Hypertrophic Cardiomyopathy in the Preoperative Clinic

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Background: Hypertrophic cardiomyopathy is a relatively common inherited disorder that may be asymptomatic. It is a common cause of sudden cardiac death and can lead to catastrophic cardiovascular collapse in the operating room if previously undiagnosed. This case report discusses the implications of unsuspected hypertrophic cardiomyopathy for anesthesia management.
Case Presentation: A veteran aged 55 years presented to the preadmission testing clinic prior to undergoing outpatient surveillance colonoscopy under anesthesia. His initial medical history was unremarkable. On physical examination, a murmur with benign characteristics was detected. However, further evaluation with echocardiography, revealed severe hypertrophic obstructive cardiomyopathy. The patient was ultimately referred for septal myectomy consideration.
Conclusions: Hypertrophic cardiomyopathy is a relatively common disorder that often remains undiagnosed. This condition has critical implications for preoperative evaluation, as patients with this disorder may develop sudden, unexpected, and refractory hypotension upon induction of anesthesia.

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Hypertrophic cardiomyopathy (HCM) is a relatively common inherited condition characterized by abnormal asymmetric left ventricular (LV) thickening. This can lead to LV outflow tract (LVOT) obstruction, which has important implications for anesthesia management. This article describes a case of previously undiagnosed HCM discovered during a preoperative physical examination prior to a routine surveillance colonoscopy.

CASE PRESENTATION

A 55-year-old Army veteran with a history of a sessile serrated colon adenoma presented to the preadmission testing clinic prior to planned surveillance colonoscopy under monitored anesthesia care. His medical history included untreated severe obstructive sleep apnea (53 apnea-hypopnea index score), diet-controlled hypertension, prediabetes (6.3% hemoglobin A_{1c}), hypogonadism, and obesity (41 body mass index). Medications included semaglutide 1.7 mg injected subcutaneously weekly and testosterone 200 mg injected intramuscularly every 2 weeks, as well as lisinopril-hydrochlorothiazide 10 to 12.5 mg daily, which had recently been discontinued because his blood pressure had improved with a low-sodium diet.

A review of systems was unremarkable except for progressive weight gain. The patient had no family history of sudden cardiac death. On physical examination, the patient's blood pressure was 119/81 mm Hg, pulse was 86 beats/min, and respiratory rate was 18 breaths/min. The patient was clinically euvolemic, with no jugular venous distention or peripheral edema, and his lungs were clear to auscultation. There was, however, a soft, nonradiating grade 2/6 systolic murmur that had not been previously documented. The murmur decreased substantially with the Valsalva maneuver, with no change in hand grip.

Laboratory studies revealed hemoglobin and renal function were within the reference range. A routine 12-lead electrocardiogram (ECG) was unremarkable. A transthoracic echocardiogram revealed moderate pulmonary hypertension (59 mm Hg right ventricular systolic pressure), asymmetric LV hypertrophy (2.1 cm septal thickness), and severe LVOT obstruction (131.8 mm Hg gradient). Severe systolic anterior motion of the mitral valve was also present. The LV ejection fraction was 60% to 65%, with normal cavity size and systolic function. These findings were consistent with severe hypertrophic obstructive cardiomyopathy (HOCM). Upon more detailed questioning, the patient reported that over the previous 5 years he had experienced gradually decreasing exercise tolerance and mild dyspnea on exertion, particularly in hot weather, which he attributed to weight gain. He also reported a presyncopal episode the previous month while working in his garage in hot weather for a prolonged period of time.

The patient's elective colonoscopy was canceled, and he was referred to cardiology. While awaiting cardiac consultation, he was instructed to maintain good hydration and avoid any heavy physical activity beyond walking. He was told not to resume his use of lisinopril-hydrochlorothiazide. A screening 7-day Holter monitor showed no ventricular or supraventricular ectopy. After cardiology consultation, the patient was referred to a HCM specialty clinic, where a cardiac magnetic resonance imaging confirmed severe asymmetric hypertrophy with resting obstruction (Figures 1-4). Treatment options were discussed with the patient, and he underwent a trial with the β-blocker metoprolol 50 mg daily, which he could not tolerate. Verapamil extended-release 180 mg orally once daily was then initiated; however, his dyspnea persisted. He was amenable to surgical therapy and underwent septal myectomy, with 12 g of septal myocardium removed. He did well postoperatively, with a follow-up echocardiogram showing normal LV systolic function and no LVOT gradient detectable at rest or with Valsalva maneuver. His fatigue and exertional dyspnea significantly improved. Once the patient underwent septal myectomy and was determined to have no detectable LVOT gradient, he was approved for colonoscopy which has been scheduled but not completed.

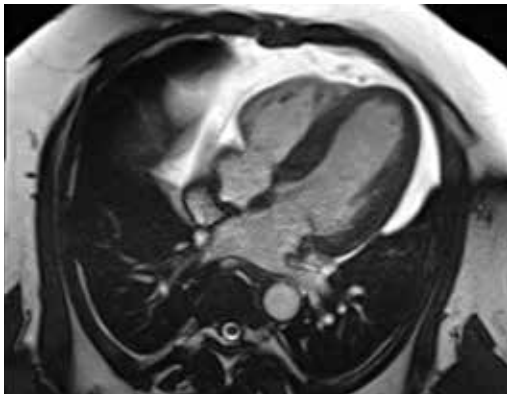

FIGURE 1. Static steady-state free precession magnetic resonance image in the 4-chamber view demonstrating asymmetrical septal hypertrophy.

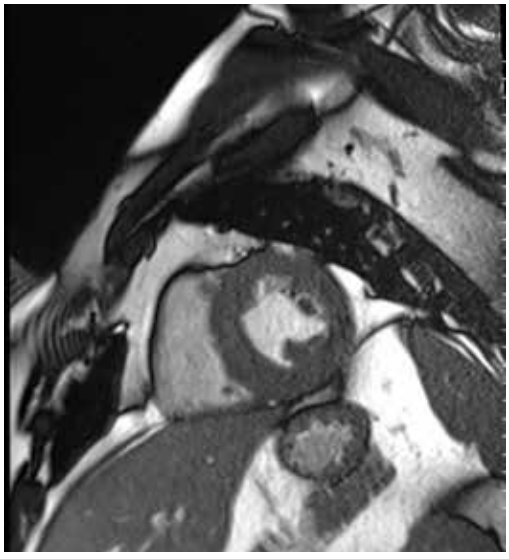

FIGURE 2. Static steady-state free precession magnetic resonance image in the short axis view demonstrating predominately septal hypertrophy.

DISCUSSION

Once thought rare, HCM is now considered to be a relatively common inherited disorder, occurring in about 1 in 500 persons, with some suggesting that the actual prevalence is closer to 1 in 200 persons.[1,2] Most often caused by mutations in ≥ 1 of 11 genes responsible for encoding cardiac sarcomere proteins, HCM is characterized by abnormal LV thickening without chamber enlargement in the absence of any identifiable cause, such as aortic valve stenosis or uncontrolled hypertension. The hypertrophy is often asymmetric, and in cases of asymmetric septal hypertrophy, dynamic LVOT obstruction can occur (known as HOCM). The condition is inherited in an autosomal dominant pattern with variable expression and is associated with myocardial fiber disarray, which can occur years before symptom onset.[3] This myocardial disarray can lead to remodeling and an increased wall-to-lumen ratio of the coronary arteries, resulting in impaired coronary reserve.

Depending on the degree of LVOT obstruction, patients with HCM may be classified as nonobstructive, labile, or obstructive at rest. Patients without obstruction have an outflow gradient ≤ 30 mm Hg that is not provoked with Valsalva maneuver, administration of amyl nitrite, or exercise treadmill testing.[3] Patients classified as labile do not have LVOT obstruction at rest, but obstruction may be induced by provocative

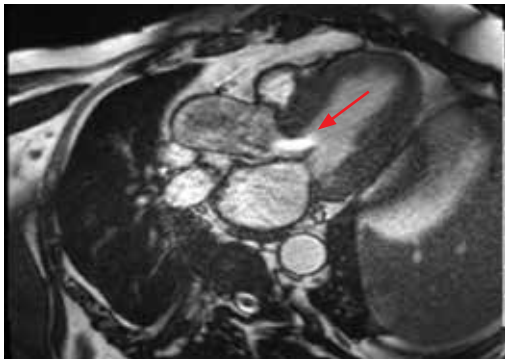

FIGURE 3. Static steady-state free precession magnetic resonance image in 3-chamber view; arrow indicates predominately septal hypertrophy with flow acceleration at level of left ventricular outflow tract secondary to systolic anterior motion of mitral valve.

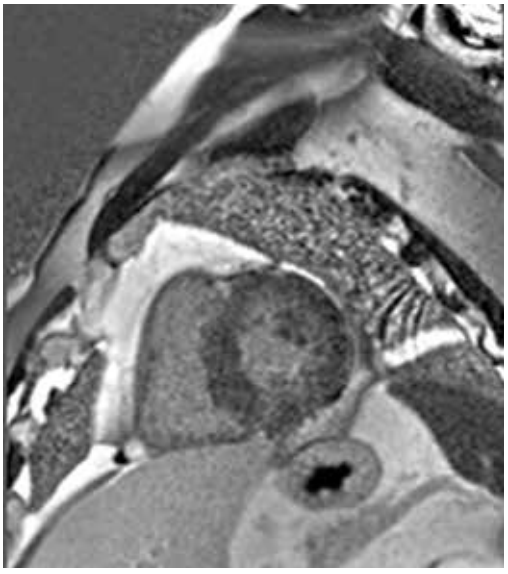

FIGURE 4. Short axis view of the mid ventricular level on delayed gadolinium magnetic resonance image showing no significant myocardial fibrosis.

measures. Finally, about one-third of patients with HCM will have LVOT gradients of > 30 mm Hg at rest. These patients are at increased risk for progression to symptomatic heart failure and may be candidates for surgical myectomy or catheter-based alcohol septal ablation.[4] The patient in this case had a resting LVOT gradient of 131.8 mm Hg on echocardiography. The magnitude of this gradient placed the patient at a significantly higher risk of ventricular dysrhythmias and sudden cardiac death.[5]

Wall thickness also has prognostic implications.[6] Although any area of the myocardium can be affected, the septum is involved in about 90% cases. In their series of 48 patients followed over 6.5 years, Spirito et al found that the risk of sudden death in patients with HCM increased as wall thickness increased. For patients with a wall thickness of < 15 mm, the risk of death was 0 per 1000 person-years; however, this increased to 18.2 per 1000 person-years for patients with a wall thickness of > 30 mm.[7]

While many patients with HCM are asymptomatic, others may report dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea, chest pain, palpitations, presyncope/syncope, postural lightheadedness, fatigue, or edema. Symptomatology, however, is quite variable and does not necessarily correlate with the degree of outflow obstruction. Surprisingly, some patients with significant LVOT may have minimal symptoms, such as the patient in this case, while others with a lesser degree of LVOT obstruction may be very symptomatic.[3,4]

Physical examination of a patient with HCM may be normal or may reveal nonspecific findings such as a fourth heart sound or a systolic murmur. In general, physical examination abnormalities are related to LVOT obstruction. Those patients without significant outflow obstruction may have a normal cardiac examination. While patients with HCM may have a variety of systolic murmurs, the 2 most common are those related to outflow tract obstruction and mitral regurgitation caused by systolic anterior motion of the mitral valve.[4] The systolic murmur associated with significant LVOT obstruction has been described as a harsh, crescendo-decrescendo type that begins just after S1 and is heard best at the apex and lower left sternal border.[4] It may radiate to the axilla and base but not generally into the neck. The murmur usually increases with Valsalva maneuver and decreases with handgrip or going from a standing to a sitting/squatting position. The initial examination of the patient in this case was not suggestive of HOCM, as confirmed by 2 practitioners (a cardiologist and an internist), each with > 30 years of clinical expe-

rience. This may have been related to the patient's hydration status at the time, with Valsalva maneuver increasing obstruction to the point of reduced flow.

About 90% of patients with HCM will have abnormalities on ECG, most commonly LV hypertrophy with a strain pattern. Other ECG findings include: (1) prominent abnormal Q waves, particularly in the inferior (II, III, and aVF) and lateral leads (I, aVL, and V4-V6), reflecting depolarization of a hypertrophied septum; (2) left axis deviation; (3) deeply inverted T waves in leads V2 through V4; and (4) P wave abnormalities indicative of left atrial (LA) or biatrial enlargement.[8] It is notable that the patient in this case had a normal ECG, given that a minority of patients with HCM have been shown to have a normal ECG.[9]

Echocardiography plays an important role in diagnosing HCM. Diagnostic criteria include the presence of asymmetric hypertrophy (most commonly with anterior septal involvement), systolic anterior motion of the mitral valve, a nondilated LV cavity, septal immobility, and premature closure of the aortic valve. LV thickness is measured at both the septum and free wall; values ≥ 15 mm, with a septal-to-free wall thickness ratio of ≥ 1.3, are suggestive of HCM. Asymmetric LV hypertrophy can also be seen in other segments besides the septum, such as the apex.[10]

HCM/HOCM is the most common cause of sudden cardiac death in young people. The condition also contributes to significant functional morbidity due to heart failure and increases the risk of atrial fibrillation and subsequent stroke. Treatments tend to focus on symptom relief and slowing disease progression and include the use of medications such as β-blockers, nondihydropyridine calcium channel blockers, and the myosin inhibitor mavacamten.[11] Select patients, such as those with severe LVOT obstruction and symptoms despite treatment with β-blockers or nondihydropyridine calcium channel blockers, may be offered septal myectomy or catheter-based alcohol septal ablation, coupled with insertion of an implantable cardiac defibrillator to prevent sudden cardiac death in patients at high arrhythmic risk.[1,12]

Patients with HCM, particularly those with LVOT obstruction, pose distinct challenges to the anesthesiologist because they are highly sensitive to decreases in preload and afterload. These patients frequently experience adverse perioperative events such as myocardial ischemia, systemic hypotension, and supraventricular or ventricular arrhythmias. Acute congestive heart failure may also occur, presumably due to concomitant diastolic dysfunction. Patients with previously unrecognized HCM are of particular concern, as they may manifest unexpected and sudden hypotension with the induction of anesthesia. There may then be a paradoxical response to vasoactive drugs and anesthetic agents, which accentuate LVOT obstruction. In these circumstances, undiagnosed HCM should be considered, and intraoperative rescue transesophageal echocardiography be performed.[13] Once the diagnosis is confirmed, efforts should be made to reduce myocardial contractility and sympathetic discharge (eg, with β-blockers), increase afterload (eg, with α1 agonists), and improve preload with adequate hydration. Proper resuscitation of hypotensive patients with HCM requires a thorough understanding of disease pathology, as effective interventions may seem to be counterintuitive. Inotropic agents such as epinephrine are contraindicated in HCM because increased inotropy and chronotropy worsen LVOT obstruction. Volume status is often tenuous; while adequate preload is important, overly aggressive fluid resuscitation may promote heart failure. It is important to keep in mind that even patients without resting LVOT obstruction may develop dynamic obstruction with anesthesia induction due to sudden reductions in preload and afterload. It is also important to note that the degree of LV hypertrophy is directly correlated with arrhythmic sudden death. Those patients with LV wall thickness ≥ 30 mm are at increased risk for potentially lethal tachyarrhythmias in the operating room.[14]

These considerations reinforce the need for proper preoperative identification of patients with HCM. Heightened awareness is key, given the fact that HCM is relatively common and tends to be underdiagnosed in the general population. These patients are generally young, otherwise healthy, and often undergo minor operative procedures

in outpatient settings. It is incumbent upon the preoperative evaluator to take a thorough medical history and perform a careful physical examination. Clues to the diagnosis include exertional dyspnea, fatigue, angina, syncope/presyncope, or a family history of sudden cardiac death or HCM. A systolic ejection murmur, particularly one that increases with standing or Valsalva maneuver, and decreases with squatting or handgrip may also raise clinical suspicion. These patients should undergo a full cardiac evaluation, including echocardiography.

CONCLUSIONS

HCM is a common condition that is important to diagnose in the preoperative clinic. Failure to do so can lead to catastrophic complications during induction of anesthesia due to the sudden reduction in preload and afterload, which may cause a significant increase in LVOT obstruction. A high index of suspicion is essential, as clinical diagnosis can be challenging. The physical examination may be deceiving and symptoms are often subtle and nonspecific. It is imperative to alert the anesthesiologist before surgery so the complex hemodynamic management of patients with HOCM can be appropriately managed.

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Ethics and consent

This case report is exempt from Institutional Review Board approval. Written informed consent was obtained from the patient.

References

1. Cheng Z, Fang T, Huang J, Guo Y, Alam M, Qian H. Hypertrophic cardiomyopathy: from phenotype and pathogenesis to treatment. *Front Cardiovasc Med*. 2021;8:722340. doi:10.3389/fcvm.2021.722340
2. Semsarian C, Ingles J, Maron MS, Maron BJ. New perspectives on the prevalence of hypertrophic cardiomyopathy. *J Am Coll Cardiol*. 2015;65(12):1249-1254. doi:10.1016/j.jacc.2015.01.019
3. Hensley N, Dietrich J, Nyhan D, Mitter N, Yee MS, Brady M. Hypertrophic cardiomyopathy: a review. *Anesth Analg*. 2015;120(3):554-569. doi:10.1213/ANE.0000000000000538
4. Maron BJ, Desai MY, Nishimura RA, et al. Diagnosis and evaluation of hypertrophic cardiomyopathy: JACC state-of-the-art review. *J Am Coll Cardiol.* 2022;79(4):372–389. doi:10.1016/j.jacc.2021.12.002
5. Jorda P, Garcia-Alvarez A. Hypertrophic cardiomyopathy: sudden cardiac death risk stratification in adults. *Glob Cardiol Sci Pract.* 2018;3(25). doi:10.21542/gcsp.2018.25
6. Wigle ED, Sasson Z, Henderson MA, et al. Hypertrophic cardiomyopathy. The importance of the site and the extent of hypertrophy. A review. *Prog Cardiovasc Dis*. 1985;28(1):1-83. doi:10.1016/0033-0620(85)90024-6
7. Spirito P, Bellone P, Harris KM, Bernabo P, Bruzzi P, Maron BJ. Magnitude of left ventricular hypertrophy and risk of sudden death in hypertrophic cardiomyopathy. *N Engl J Med.* 2000;342(24):1778–1785. doi:10.1056/NEJM200006153422403
8. Veselka J, Anavekar NS, Charron P. Hypertrophic obstructive cardiomyopathy *Lancet*. 2017;389(10075):1253-1267. doi:10.1016/S0140-6736(16)31321-6
9. Rowin EJ, Maron BJ, Appelbaum E, et al. Significance of false negative electrocardiograms in preparticipation screening of athletes for hypertrophic cardiomyopathy. *Am J Cardiol*. 2012;110(7):1027-1032. doi:10.1016/j.amjcard.2012.05.035
10. Losi MA, Nistri S, Galderisi M et al. Echocardiography in patients with hypertrophic cardiomyopathy: usefulness of old and new techniques in the diagnosis and pathophysiological assessment. *Cardiovasc Ultrasound.* 2010;8(7). doi:10.1186/1476-7120-8-7
11. Tian Z, Li L, Li X, et al. Effect of mavacamten on chinese patients with symptomatic obstructive hypertrophic cardiomyopathy: the EXPLORER-CN randomized clinical trial. *JAMA Cardiol*. 2023;8(10):957-965. doi:10.1001/jamacardio.2023.3030
12. Fang J, Liu Y, Zhu Y, et al. First-in-human transapical beating-heart septal myectomy in patients with hypertrophic obstructive cardiomyopathy. *J Am Coll Cardiol*. 2023;82(7):575-586. doi:10.1016/j.jacc.2023.05.052
13. Jain P, Patel PA, Fabbro M 2nd. Hypertrophic cardiomyopathy and left ventricular outflow tract obstruction: expecting the unexpected. *J Cardiothorac Vasc Anesth*. 2018;32(1):467-477. doi:10.1053/j.jvca.2017.04.054
14. Poliac LC, Barron ME, Maron BJ. Hypertrophic cardiomyopathy. *Anesthesiology*. 2006;104(1):183-192. doi:10.1097/00000542-200601000-00025