Hemoptysis as a rare presenting symptom of hypertrophic obstructive cardiomyopathy

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Abstract

A 36-year-old female presented with two episodes of hemoptysis induced by exertion and acute dyspnea. She was diagnosed as a case of hypertrophic obstructive cardiomyopathy (HOCM) with systolic anterior motion (SAM)-induced severe mitral regurgitation (MR). She underwent extended septal myectomy with mitral valve replacement under cardiopulmonary bypass and recovered successfully. This is a unique and rare combination of HOCM with hemoptysis.

Keywords Hypertrophic cardiomyopathy · Hemoptysis · Alveolar hemorrhage · Septal myectomy

Introduction

Hemoptysis is an uncommon presentation in a number of structural heart diseases, the most common being mitral stenosis. Raised pulmonary capillary pressure leading to alveolar hemorrhage, similar to that seen in mitral stenosis, explains the pathophysiology of hemoptysis in hypertrophic obstructive cardiomyopathy. This report presents an uncommon case of hypertrophic obstructive cardiomyopathy who presented with hemoptysis.

Case report

A 36-years-old female came to the outpatient department with complaint of 2 episodes of hemoptysis within 15 days and chest pain. She also complained of dyspnea which had gradually increased over last 4 years. Patient was hemodynamically stable except tachycardia. There were no signs of heart failure like engorged neck veins and pedal edema. Cardiac auscultation revealed normal first and second heart sounds

and 2/6 systolic murmur over the apex suggestive of mitral regurgitation (MR). Her chest examination revealed bilateral coarse crepitations in mid zones. X-ray demonstrated bilateral diffuse infiltrates. Contrast enhanced computed tomogram of the chest was done which shows bilateral diffuse patchy ground glass opacities (Fig. 1). In view of the above findings suggestive of a possible infective etiology, sputum was sent for culture, gram-staining, fungus, and acid-fast bacillus examination, and the analysis was negative. A 2D transthoracic echocardiogram showed asymmetric septal hypertrophy, systolic anterior motion (SAM)-induced severe MR, and peak gradient across left ventricular outflow tract (LVOT) of 53 mmHg in resting phase. Her blood coagulation profile was normal. Patient was treated medically to improve the pulmonary vascular congestion with beta blocker (metoprolol), diuretic (furosemide), and chest physiotherapy and led to improvement. This was confirmed with repeat computed tomogram scan after 3 weeks which confirmed marked improvement in previous opacities (Fig. 2). Surgical correction was planned. Transaortic extended septal myectomy (classical septal myectomy plus resection of abnormal papillary muscles attaching directly from septum to mitral leaflet and extending the resection towards apical septum) was carried out. Mitral valve was inspected by left atrium approach, and anterior mitral leaflet was thinned out, enlarged, and distorted and not suitable for mitral valve repair. Therefore, mitral valve replacement (St. Jude Michael (SJM) bi-leaflet mechanical valve prosthesis—size 27) was performed after septectomy. Post-operative period was uneventful. She was discharged on the 7th postoperative day.

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Fig. 1 Contrast enhanced computed tomogram of the chest

Fig. 2 Repeat computed tomogram scan after 3 weeks

Discussion

Hypertrophic cardiomyopathy (HCM) is considered the most prevalent genetically transmitted cardiomyopathy [1]. Many patients of HCM present with primary functional and structural abnormalities of mitral valve apparatus like pathological elongation and thickening of mitral valve, anterior displacement and abnormal insertion of papillary muscles, and abnormal attachment of chordae tendineae and elongated leaflets resulting in SAM and malcoaptation of mitral leaflets [2]. SAM of mitral valve resulting in dynamic LVOT obstruction is an important pathophysiological hallmark in HCM.

The disease does not usually manifest until puberty, and most of the patients remain asymptomatic. The onset of symptoms develops with the gradual development of LVOT obstruction. Most of the patients have mild symptoms only. Dyspnea is the most common presenting symptom followed by angina and, less frequently, fatigue, palpitations, pre-syncope, syncope, and congestive heart failure symptoms [1]. Complete heart block is an uncommon complication [3]. A few cases were also reported mimicking myocardial infarction as their presenting symptom [4]. Reduced functional capacity is common in HCM. Orthopnea and paroxysmal nocturnal dyspnea are rare, but bendopnea and postprandial breathlessness are common symptoms [5]. LVOT obstruction is a common cause of these clinical manifestations, but exertional dyspnea is also caused by diastolic and systolic left ventricular impairment and atrial arrhythmia.

Presentation of HCM as hemoptysis has been reported in the literature [6]. Cardiovascular conditions that increase the mechanical pressure overload of the capillaries result in alveolar hemorrhage. The most common cardiovascular causes of hemoptysis include mitral stenosis, pulmonary edema, pulmonary embolism with pulmonary infarction, rupture of aortic aneurysm into bronchopulmonary tree, rupture of bronchopulmonary collaterals, and pulmonary artery aneurysm. Common pulmonary parenchymal causes for hemoptysis in India are tuberculosis, bronchiectasis, lung abscess, aspergilloma, and pneumonia. All of these cardiac and pulmonary causes were excluded on differential diagnosis. Obstructive HCM as in this case has been reported to be associated with abnormal spontaneous bleeding in some cases. HCM causes blood to be exposed to high shear stress. This impairs Von Willebrand Factor and promotes abnormal bleeding. It has been reported that Von Willebrand Factor impairment is independently related to the magnitude of outflow obstruction [7]. But as earlier mentioned, this patient had normal blood coagulation profile. MR can also result in diffuse alveolar hemorrhage and hemoptysis owing to increased pulmonary venous hypertension [8].

This patient had 2 episodes of hemoptysis which was preceded by acute dyspnea. This case demonstrates an unusual presentation of HCM, in which alveolar hemorrhage presented hemoptysis as a main symptom along with dyspnea. HCM

with severe LVOT obstruction and severe MR may present with episodic pulmonary edema leading to acute pulmonary hypertension resulting in alveolar hemorrhage. This patient had a peak LVOT gradient of 53 mmHg in resting phase and this gradient was increasing more with exertion, as we hypothesize, leading to sudden rise in pulmonary capillary pressure. Hemoptysis secondary to diffuse alveolar hemorrhage is a rare sign of left ventricular heart failure and should be considered if there is evidence of elevated left-sided filling pressures or valve disease [6, 7]. This pathophysiology is similar to that of mitral stenosis [7] and MR [8] as it relates to elevated filling pressure causing increased pulmonary capillary pressure.

Pharmacological therapy with beta-blockade, disopyramide, verapamil, and their combinations is recommended as a first-line modality for HCM. In high-risk HCM patients, intra-cardiac defibrillator remains the primary mode of prevention of sudden death. For those patients who are refractory to pharmacological management or with a systolic gradient of > 50 mmHg or who have side effects of pharmacological therapy, septal reduction therapies including percutaneous alcohol septal ablation and surgical myectomy are recommended [9]. Patients of HCM with mitral valve abnormalities have a different LVOT obstruction physiology and should not be treated by isolated septal reduction therapy. Surgical myectomy with mitral valve repair or replacement is the preferred choice [1, 10].

Conclusion

Hemoptysis is a rare presentation of HCM. This case suggests that MR and increased pulmonary capillary pressure in HCM can lead to alveolar hemorrhage resulting in hemoptysis. High index of suspicion and a thorough cardiac evaluation is needed to diagnose the underlying structural heart disease in absence of other non-cardiac etiologies. If indicated, an early intervention can produce excellent results and faster recovery.

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Compliance with ethical standards

Conflict of interests The authors declare that they have no conflict of interest.

Ethical approval Ethical approval not required for case reports as per institutional policy.

Informed consent Informed consent was obtained.

References

1. Raj MA, Bansal P, Goyal A. Hypertrophic obstructive cardiomyopathy. [Updated 2020 Feb 18]. In: StatPearls [Internet]. Treasure

- Island (FL): StatPearls Publishing; 2020 Jan-. Available from: <https://www.ncbi.nlm.nih.gov/books/NBK430820/>
2. Maron MS, Olivetto I, Harrigan C, et al. Mitral valve abnormalities identified by cardiovascular magnetic resonance represent a primary phenotypic expression of hypertrophic cardiomyopathy. *Circulation*. 2011;124:40–7.
 3. Animashaun IJB, Shepherd FM, Chinnadurai TV, et al. Atypical presentation and management of Hypertrophic Cardiomyopathy with Atrioventricular Conduction Abnormality. *Med Sci Case Rep*. 2016;3:82–85.
 4. Daralammouri Y, El Garhy M, Same K, Lauer B. Hypertrophic cardiomyopathy mimicking acute anterior myocardial infarction associated with sudden cardiac death. *Case Rep Med*. 2012;2012: 236154. <https://doi.org/10.1155/2012/236154>.
 5. Díez-López C, Salazar-Mendiguchía J. Clinical presentations of hypertrophic cardiomyopathy and implications for therapy. *Glob Cardiol Sci Pract*. 2018;2018:19. <https://doi.org/10.21542/gcsp.2018.19>.
 6. Mock V, Singh J, Scardella A. 1072: Hypertrophic obstructive cardiomyopathy as a rare cause of hemoptysis. *Crit Care Med*. 2018;46:520. <https://doi.org/10.1097/01.ccm.0000529078.03461.59>.
 7. Le Tourneau T, Susen S, Caron C, et al. Functional impairment of von Willebrand factor in hypertrophic cardiomyopathy: relation to rest and exercise obstruction. *Circulation*. 2008;118:1550–1557. <https://doi.org/10.1161/CIRCULATIONAHA.108.786681>.
 8. Izarnotegui WV, Luna M. Diffuse alveolar hemorrhage due to severe mitral valve regurgitation. *J Am Coll Cardiol*. 2015;65:A721.
 9. Hong JH, Schaff HV, Nishimura RA, et al. Mitral regurgitation in patients with hypertrophic obstructive cardiomyopathy: implications for concomitant valve procedures. *J Am Coll Cardiol*. 2016;68:1497–504.
 10. Gersh BJ, Maron BJ, Bonow RO, et al. 2011 ACCF/AHA guideline for the diagnosis and treatment of hypertrophic cardiomyopathy: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines. *Circulation*. 2011;124:2761–96.

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